Surgical technique for the resection of tumors in relation with the V3 and V4 segments of the vertebral artery

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The technique for the resection of pathologies related to the V3 and V4 segments of the vertebral artery (VA) is discussed together because it is related to lesions located at the craniocervical junction (CCJ) level. The CCJ is defined by the occipital bone, the atlas, and axis. These bone structures fix the limits of the foramen magnum (FM). The limits of the FM area previously defined are the lower third of the clivus and upper edge of the body of C2, anteriorly; the jugular tubercles and upper aspect of C2 laminae, laterally; and the anterior edge of the squamous occipital bone and C2 spinous process, posteriorly (1–4). Control of the VA V3 segment is necessary for treating not only bones and extracanalar lesions of the CCJ but also in some dumbbell-shaped tumors and FM pathologies more specifically in relation with the VA V4 segment.

Tumors in relation with the V3 and V4 segments of the VA

Our surgical experience of tumors located at the CCJ and the FM level encompasses 329 cases (Table 1). These tumors have been classified, and we differentiate among them three groups according to their compartment of development: intradural, intra-extradural, and extradural. This criterion is crucial for defining the appropriate surgical approach and is the first used in our classification system.

The first group of 143 intradural tumors essentially includes FM meningiomas (FMMs). Besides meningiomas, 31 nerve sheath tumors (NST) were also observed, including one of the spinal component of the cranial nerve (CN) XI, and 6 and 24 of the C1 and C2 roots, respectively. Eleven other types of tumors such as hemangioblastoma, melanoma, ependymoma, and different types of cysts also belong to this group of intradural tumors.

The second group of 22 intra- extradural tumors essentially consists in dumbbell-shaped NSTs that were encountered in 19 cases. Although the vast majority of FMMs were entirely intradural, three were intradural with an extradural component.

The third group of 164 extradural lesions had chordomas as the most commonly observed tumors, with 53 cases. Twenty-five purely extradural NSTs, 23 sarcomas, and 20 metastases were also encountered. Finally, different types of primary bone tumors were also observed: osteoid osteomas and osteochondromas were the most frequent ones, in seven and five cases, respectively. Some other types of tumors account for 20 cases, including tumors such as aneurismal cysts, histiocytosis, fibrous dysplasias, or histiofibromas. It must be noticed that three cases of entirely extradural meningiomas also belong to this group. Pseudotumors, which raise the problem of differential diagnosis with actual tumors, were also found in this group.

This tumoral classification according to the compartment of development helps for defining the appropriate surgical approach. Intradural tumors of the group I can all be adequately treated through a posterolateral approach. Intra-extradural tumors of group II and extradural tumors of group III can be treated either by a posterolateral approach, if the tumor extends extradurally posteriorly to the VA, or by an anterolateral approach, if its extension is anterior to the VA. The difference between group II and III is linked to the trans-

Table 1 – Summary of our series of lesions in relation with the VA V3 and V4 segments.

Groupe	I	II	III
	Intradural	Intra- Extradural	Extradural
FMM	101	3	3
Neurinomas	31	19	25
Chordomas			53
Sarcomas			23
Metastases			20
Osteoid osteomas			7
Osteochondromas			5
Pseudotumors			8
Others	11		20
	143	22	164

dural extension in group II, which is associated with a higher surgical complexity and the need for a complex dural reconstruction, especially when the tumor extends extradurally anteriorly.

Preoperative considerations

Preoperative workup

Clinical examination is crucial for detecting lower cranial nerves as well as long tract palsy. MRI, CT scan, and, on rare occasions, angiography constitute the classical preoperative workup. Gadolinium-enhanced MRI sequences help to define precisely the dural attachment zone, the tumor itself, and its relation to neural and vascular structures. T2-weighted images allow sometimes for identifying the presence of an arachnoid plane between the neuraxis and the tumor.

Bone windows CT scan is especially interesting when the lesion invades the extradural compartment. It also provides important anatomical details if a fusion has to be scheduled preoperatively.

Conventional angiography is generally useless. There are only two indications for preoperative angiography:

1) if a highly vascularized tumor is suspected and embolization is contemplated,

2) to perform a balloon occlusion test in case of VA encasement (extradural or recurrent meningioma and meningioma inserted around the VA). In our experience, it has never been necessary to occlude the VA preoperatively.

Specific preoperative recommendations for the VA management

These recommendations are identical to those presented in the previous chapter.

1. Evaluate VA dominance.

In approximately 40% of the cases, a size difference exists between both VAs. Analysis of VA dominance is very important preoperatively to determine potential risks in case of inadvertent injury and temporary or definitive occlusion. VA dominance can be determined not only directly on angiography but also after a careful analysis of the preoperative MRI and enhanced CT scan. The size of the transverse foramen can also provide some indirect information.

2. Evaluate the risk of intraoperative VA injury.

Several criteria can help for determining this risk (Table 2).

Table 2 – Criteria for evaluating the risk of intraoperative injury.

Severe VA compression
VA encasement
Malignant tumor possibly resulting in arterial wall infiltration
Prior radiation therapy
Prior surgery

• *Importance of the VA compression*

This criterion is only relevant when operating on extradural tumors through an anterolateral approach. The resection of large extradural tumors, such as C1–C2 peripheral nerve sheath tumors (PNST), by a posterolateral approach does not carry this increased risk because the tumor is largely resected when approaching the VA, which is therefore no more compressed at this step of the surgery.

During an anterolateral approach, opening the transverse foramen can be more dangerous due to the VA compression induced by the tumor. For the same reason, separating the posterior aspect of the VA sheath from the VA is also delicate since the plane of demarcation in between is tight.

• *Encasement of the VA*

Encasement of the VA V3 segment can be present in case of bone tumors of the CCJ or soft tissues sarcomas. At this level, benefit can be taken from the periosteal sheath that acts as a strong physical barrier protecting the VA adventitia from tumoral infiltration. At the V4 level, no such protection exists. Meningiomas may develop on both sides of the level of entrance of the VA within the dura and be also responsible for total VA encasement. In this situation, the dissection of the VA from the tumor represents a higher surgical complexity. In order to work confidently, it is advised to look for the VA in a tumor-free area and progress from this controlled segment to the encased segment in order to identify and then to follow the demarcation plane. Careful analysis of preparative exams is of prime importance to anticipate the exact VA location during the dissection.

• *Arterial wall infiltration by malignant tumors*

Malignant tumors have a high propensity for infiltrating the VA sheath and the arterial wall. This condition precludes any safe separation between the tumor and the VA itself. Opening and resecting the VA sheath as well as peeling the arterial adventitia is possible, but this represents a major surgical difficulty and risk. A radical resection may need the VA occlusion and resection at the tumoral level. A balloon occlusion test is a safe prerequisite.

• *Prior radiation therapy and prior surgery*

Both factors are responsible for fibrosis and thereby for increasing adherences with the VA.

3. Evaluate the risks associated with VA sacrifice (Table 3).

VA sacrifice has evidently to be avoided but can nevertheless be performed without consequences in some conditions. Main factors determining whether VA sacrifice can be performed are the VA size, the collateral circulation, and the territory of the arterial supply. VA sacrifice may be performed securely in case of involvement of the minor VA, except if the VA ends at the posterior inferior cerebellar artery (PICA). On the other hand, if the tumor involves an equivalent or a dominant VA, the risk of VA injury must be considered. If the risk is low, we advise not to perform an occlusion test because the risk of the test is not counterbalanced by the risk of injury. However, if the risk of VA injury is high, meaning that one of the risk factors previously detailed is present, then an occlusion test must be performed for evaluating the collateral flow and clinical tolerance. In some conditions such as in case of malignant tumor, the VA has to be sacrificed for oncological purposes in order to achieve complete resection if the arterial wall is infiltrated. If the test occlusion is tolerated, then VA sacrifice can be performed without any hazard. If it is not, then distal VA revascularization with a saphenous vein carotidovertebral bypass is mandatory.

4. Check the VA location and exclude anatomical variations.

Any displacement of the VA by a tumor must be analyzed carefully to anticipate the accurate VA location. Whatever this displacement, the VA V3 segment has 3 fixed points that can be found at the levels of the C2 and C1 transverse foramina as well as its entry point into the dura mater of the FM. The VA displacement is always opposite to the growth center of the tumor.

The trajectory of the VA V3 segment between the C2 transverse process and the dura mater at the FM level must be checked to rule out the persistence of the first intersegmental artery. In this situation, the VA turns medially and runs intradurally after exiting the C2 transverse foramen, without passing through the C1 one. The incidence of this anomaly is 0.6% in the absence of associated bone anomalies at the CCJ level but increases up to 19–36.4% if present (5–7).

An abnormal extradural origin of the PICA is another highly relevant variation. Found in 5 to 20% of the cases, this anomaly is not so rare (8). The PICA may originate on the horizontal portion of the VA V3 segment penetrating the dura mater close to the VA itself or on the vertical portion, entering the dura mater between C2 and C1 (8).

5. Check bone modifications.

Bone anomalies may increase surgical difficulty. A typical example is the posterior arch of atlas turns into a tunnel after calcification or ossification of the occipitoatlantal membrane (9, 10). This condition induces some difficulties for exposing the horizontal portion of the VA V3 segment. We deal with this problem by pursuing the dissection slowly from a normal area.

Another important variation is the incomplete closure of the C1 posterior arch. Bone landmarks are modified during the dissection, and failure to detect this anomaly may result in VA damage during the subperiosteal dissection or neural structures injury. Finally, slow-growing and aggressive tumors may also be responsible for bone erosion or destruction.

All bone modifications can be detected on CT with bone windows. This preoperative careful analysis is especially important because bone structures are crucial landmarks during the procedure and failure to recognize any variation will expose to troublesome complications as a result of technical mistakes.

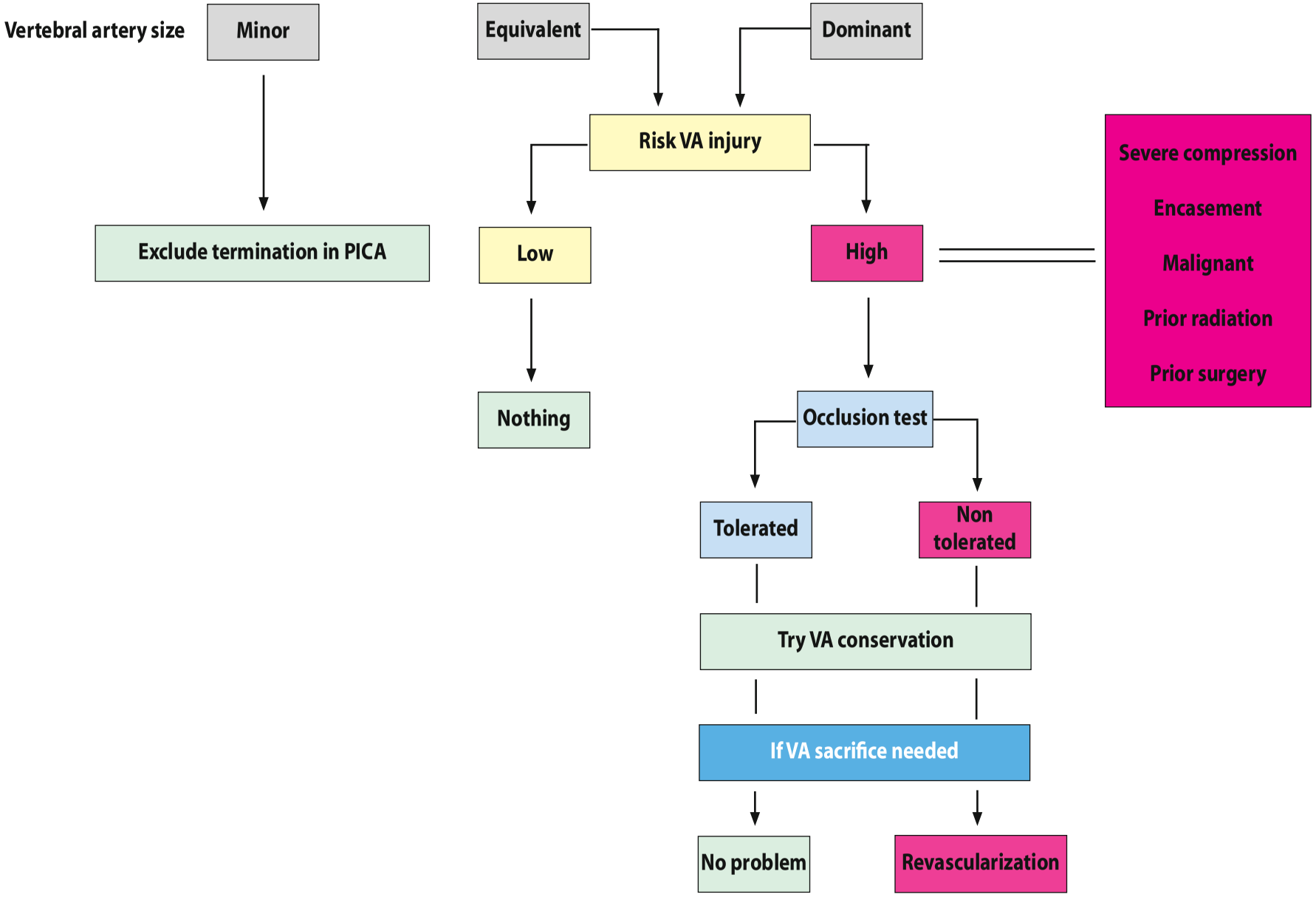
Contrary to the recommendation given at the V2 segment, the level and side of the anterior spinal artery (ASA) never needs to be determined when approaching lesions in relation with the VA V3 segment because the ASA always originates below the C3 level.

Intraoperative recommendations

Obtain early VA control

The main principle when operating on a tumor in close relation with the VA is that the VA must be controlled as soon as possible. Whatever happens afterwards, if the artery is controlled on both sides, the surgeon will at any time be able to control bleedings, even by temporary occlusion of the artery. Bleedings can be extremely difficult to control in case of inadvertent damage. Nevertheless, proximal and distal controls can be difficult in some circumstances. Indeed, proximal control of the VA V3 segment at the C2 level is very difficult through a posterolateral approach. It is also the case for obtaining distal V4 segment control in presence of a large intradural tumor that prevents further distal access until

Table 3 – Risks associated with vertebral artery sacrifice. Algorithm for occlusion test and revascularization.



tumor debulking has been performed. Great care must be taken in such circumstances because VA tearing could become troublesome.

Tumor specificities

Foramen magnum meningiomas

Meningiomas are common neoplasms that represent 14.3–19% of all intracranial tumors (11). At the FM level, meningiomas are the most commonly observed tumors, representing 70% of all benign tumors (2–4, 12–15). On the other hand, among all intracranial meningiomas, only 1.5–3.2% develops at the FM level (15, 16). Several reasons explain why the lesion is often large when discovered: their growing rate is slow, their development is indolent, the diagnosis is difficult leading to a long interval since the first symptom, and the subarachnoid space is wide at this level (17). The vicinity of the brain stem, medulla oblongata, lower cranial nerves, and the VA renders undoubtedly FMMs challenging tumors. Several approaches have been advocated. The choice of the most appropriate must be determined according to the tumor development with the definite goals to achieve the largest tumor removal with the lowest morbidity rate as possible. For these reasons, the approach must allow adequate controls of all important neurovascular structures, without exposing to unnecessary risks.

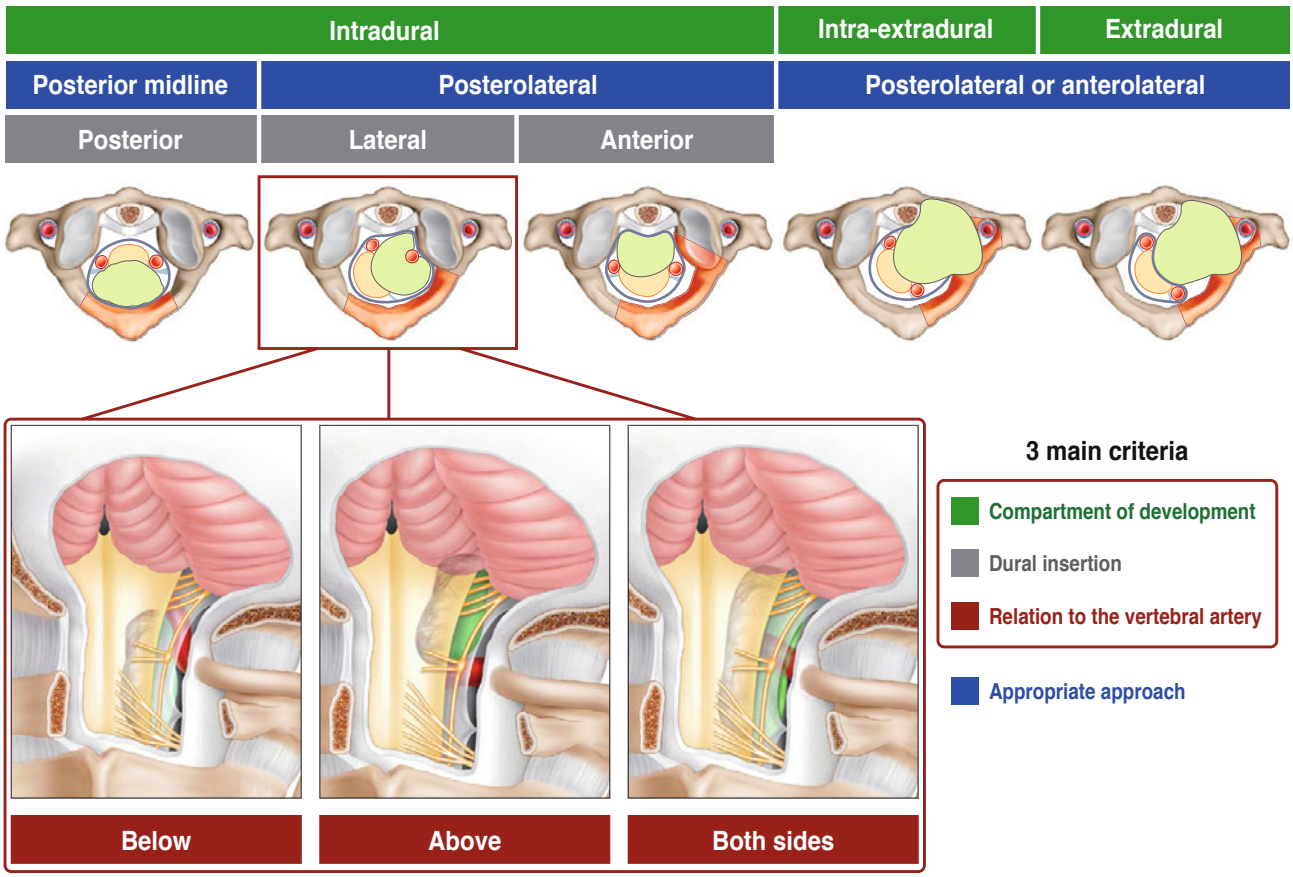
Classification system (Illustration 1)

The definitive goal of this classification system is to allow for determining preoperatively the adequate surgical strategy on the basis of the preoperative imaging characteristics of the lesion. This surgical strategy serves not only for determining the appropriate surgical approach but also for anticipating the modified position of vital neurovascular structures, with the definite goal of reducing the postoperative morbidity.

By definition, meningiomas of the FM have their primary base of insertion within the FM limits. This rule excludes tumors invading the FM region secondarily.

The first criterion for classifying FMMs is the compartment of development; therefore, it is impor-

Illustration 1 – Classification system of foramen magnum meningiomas.



tant to revisit the classification within three groups of all tumors of the CCJ. Group I is composed of purely intradural tumors (Figs. 1–12), group II of intra-extradural ones (Fig. 13), and group III of purely extradural lesions. This classification determines the adequate surgical approach.

The group I of purely intradural meningiomas represent the main group of tumors. Ninety-four per cent (101 out of 107 tumors) belongs to this group.

Intradural meningiomas must be subdivided according to two criteria: their base of insertion and their relation with the VA, determining, respectively, the tumor position in the horizontal and vertical plane.

According to their base of insertion, FMMs can be classified along the horizontal plane into anterior, lateral, and posterior. An intradural FMM is defined as anterior if its base of insertion is observed on both sides of the midline (Figs. 1–6), as lateral if it takes its origin between the midline and the dentate ligament (Figs. 7–10), or as posterior if found behind this ligament (Figs. 11–12). Out of 104 intradural tumors (101 of group I and 3 of group II), the percentage of tumor in each group is 39.4%, 54.8%, and 5.8%, respectively.

The dural insertion is in fact the area where the meningioma starts its growth; therefore, the neuraxis is displaced opposite to it and the surgical field opened accordingly. The anatomy of the bone structures at the FM level is also crucial for tailoring the surgical approach to minimize bone resection and maintain joint stability. In fact, at the FM level, the C0–C1 and C1–C2 joints are located anteriorly. Therefore, the surgical access is already enlarged and the need for bone resection reduced. Lateral meningiomas induce a posterolateral displacement of the spinal cord. In this situation, the surgical access required during a posterolateral approach is enlarged and drilling of the lateral mass of the atlas or the occipital condyle is never requested. Anterior meningiomas displace the neuraxis posteriorly. So, the surgical corridor between the neuraxis and the FM lateral wall is narrower than in lateral meningiomas; therefore, the bone resection must be extended to the medial part of the FM lateral wall to improve the access. Nevertheless, in almost every case no drilling of the lateral mass of atlas and occipital condyle is necessary because the size of the tumor has enlarged the access (Fig. 1). The rare occasion in which there may be a need for more bone resection is in case of small anterior meningiomas. In this situation, some bone resection of the lateral wall may be needed but always limited to less than one-fifth of the FM lateral wall. In that way, instability is never observed postoperatively.

The main reference structure in the vertical plane is the VA, which allows for determining the vertical position of FMMs. The vertical plane contains also the lower cranial nerves whose position can afterwards be anticipated according to the position of the meningioma related to the VA. In fact, FMMs are able to develop below, above, or on both sides of the VA. In

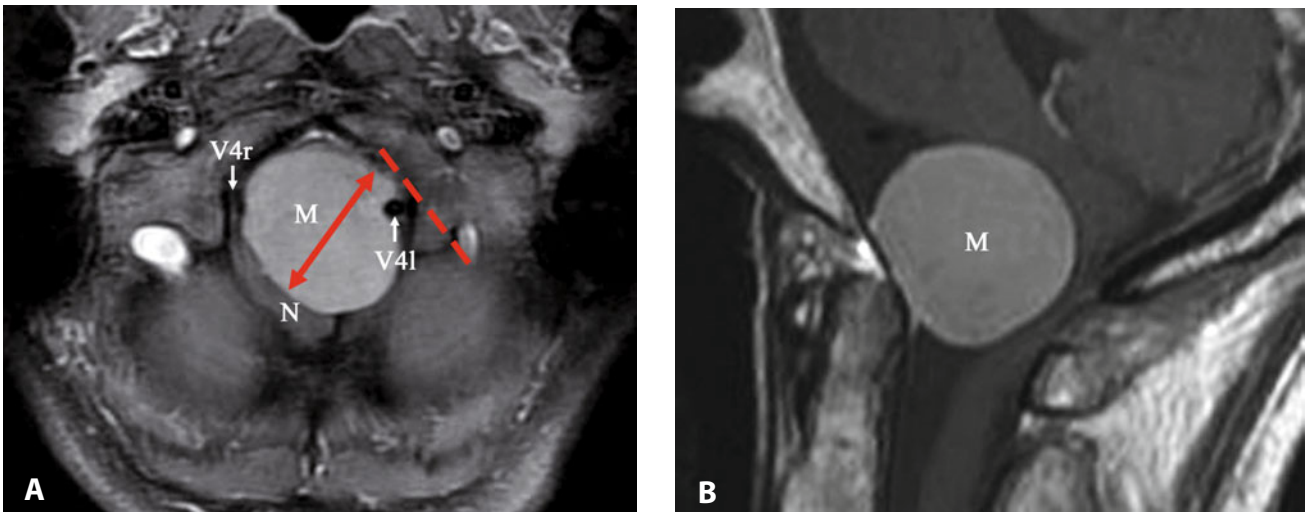
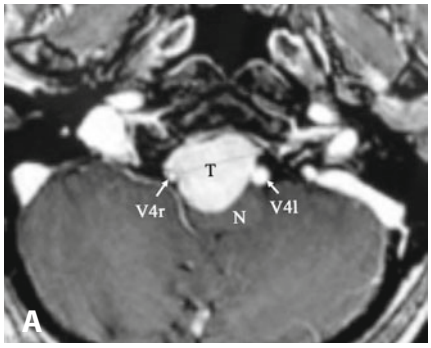


Fig. 1 – Anterior foramen magnum meningioma. **A**. Axial T1-weighted MRI with gadolinium. This meningioma inserts on both sides of the midline. Notice large lesions create by itself an extended surgical corridor *(double arrow)*. Consequently, bone resection of the foramen magnum lateral wall can be unnecessary or at maximum limited to one-fifth *(dotted line)*. The larger the lesion, the more important is the compression of the neuraxis, which is displaced controlaterally to the base of insertion. **B**. Sagittal view. M: meningioma. N: neuraxis. V4l: left vertebral artery V4 segment. V4r: right vertebral artery V4 segment.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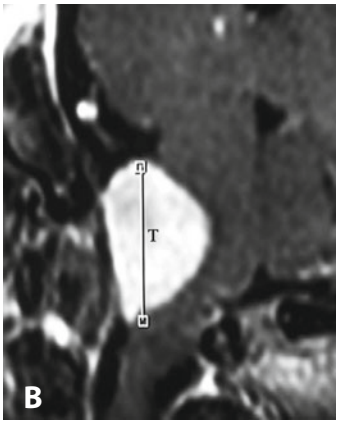
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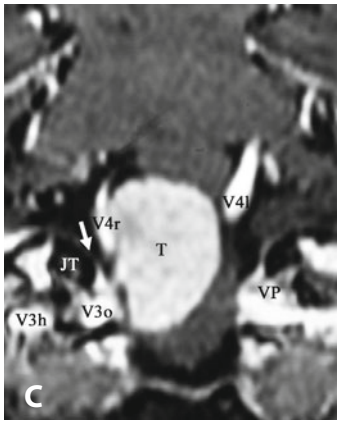
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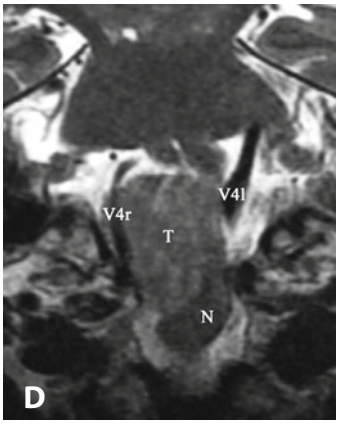
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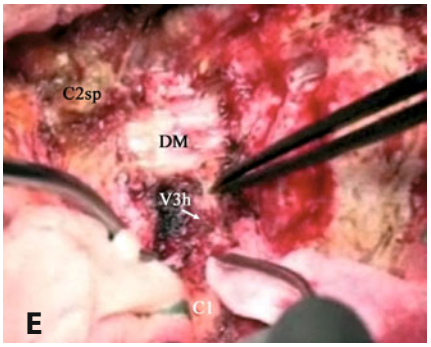
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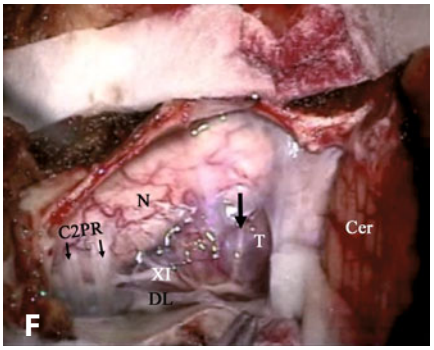
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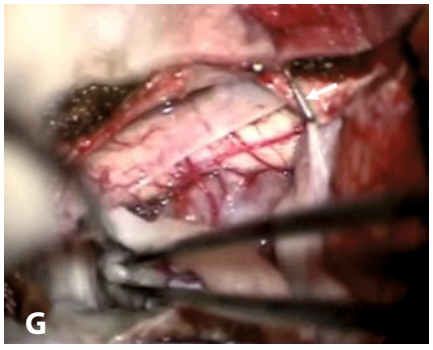
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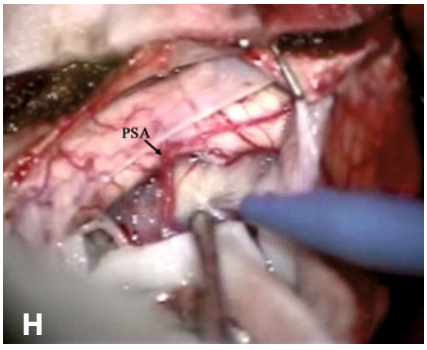
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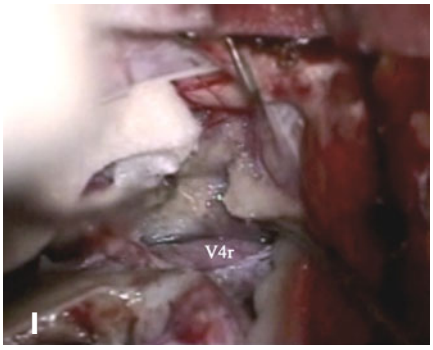
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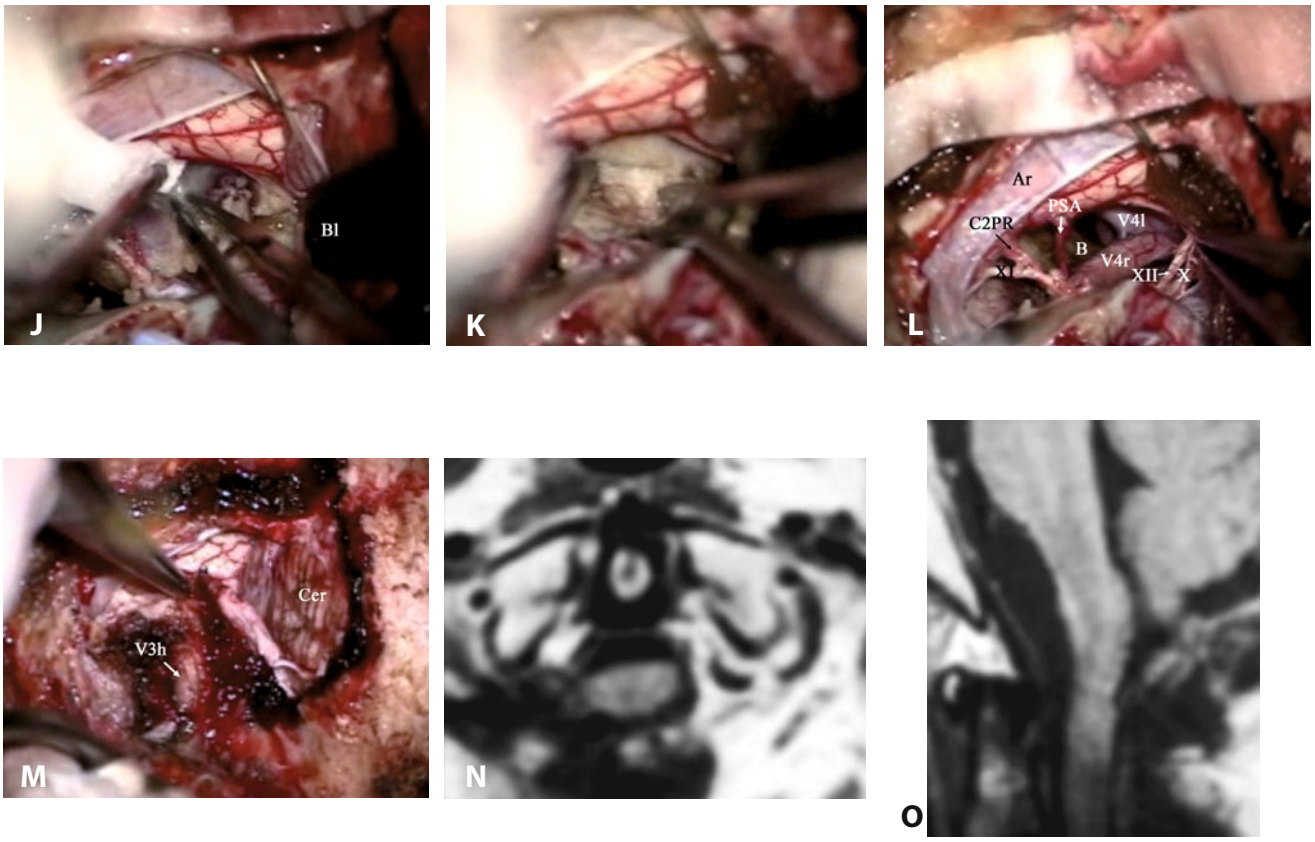


Fig. 2 – Anterior meningioma developed below the VA. **A–D**. Preoperative workup. **A**. Axial T1-weighted MRI with gadolinium. Relations between the tumor, neuraxis, and both VAs are well observed. **B**. Sagittal view. **C**. Coronal view. Clearly the right VA V4 segment runs above the meningioma. In this condition, the lower cranial nerves will be found at the superior pole of the tumor at surgery. Notice the dura mater *(arrow)* of the foramen magnum lateral wall that forms at the level of the occipital condyle the dural ring at the junction between the V3 and V4 segments of the VA. At this level, a venous plexus is observed surrounding the artery. **D**. Same coronal view on T2-weighted MRI. **E–M**. Operative views. **E**. Posterolateral approach with lateral suboccipital craniotomy and C1 laminectomy extended on the pathological side to the lateral mass after control of the horizontal portion of the VA V3 segment (located between the tip of the bipolar forceps and the sucker) and controlaterally apart from the midline. **F**. The dura mater has been incised exposing the neuraxis, the right cerebellar hemisphere and the tumor. Notice the C2 posterior rootlets, the accessory nerve, and a C1 posterior rootlet branch joining it *(black arrow)*. Cutting the dentate ligament at this step is important to release the spinal cord and improve the access to the tumor. **G**. Because the meningioma grows below the VA and lower cranial nerves are displaced upward, the resection has to start at the inferior pole. It allows also to divide the blood coming from a dural branch of the third interspace. The arachnoid has been incised and fixed to the dura mater with a vascular clip *(arrow)*. **H**. The lesion is then incised and debulked. Notice the posterior spinal artery crossing the posterior aspect of the meningioma. **I**. The left VA V4 segment is identified by mobilizing the tumor medially. This maneuver can only be done safely after tumor debulking in order to avoid an increased pressure on the neuraxis. **J**. The tumor is progressively separated from the neuraxis. Retraction is never apply on the neuraxis itself. A blade is now used to prevent the fall of the cerebellum hemisphere and improve the access to the superior aspect of the lesion. **K**. Tumor debulking progresses. **L**. View after complete resection of the lesion. Both VA V4 segments are visible. The lower cranial nerves are well visible. Notice the base of insertion of the tumor at the anterior aspect of the foramen magnum dura mater. **M**. View before dural closure when the dura mater is turned down. **N–O**. Postoperative MRI after complete tumor resection. Ar: arachnoid. B: meningioma base of insertion. Bl: blade. C1: lateral mass of atlas. C2PR: C2 posterior rootlets. C2sp: spinous process of the axis. Cer: cerebellum. DL: dentate ligament. DM: dura mater. JT: jugular tubercle. N: neuraxis. T: tumor. PSA: posterior spinal artery. X: vagal nerve. XI: accessory nerve. XII: hypoglossal nerve. V3h: horizontal portion of the vertebral artery V3 segment. V3o: oblique portion of the vertebral artery V3 segment. V4l: left vertebral artery V4 segment. V4r: right vertebral artery V4 segment. VP: venous plexus.

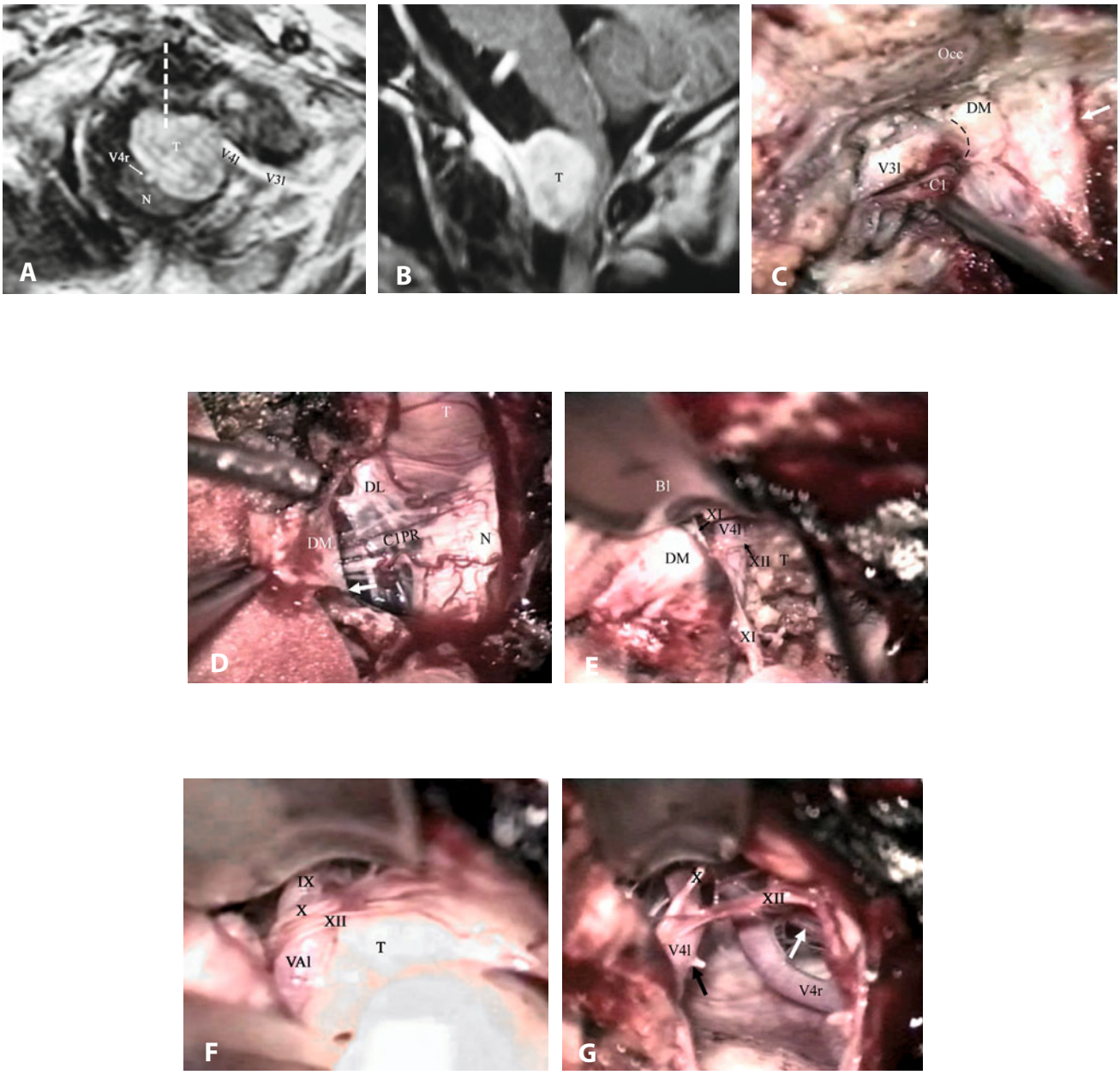


Fig. 3 – Anterior foramen magnum meningioma growing below the VA. **A**. Axial T1-weighted MRI with gadolinium at the junction level between the left VA V3 and V4 segments. This intradural meningioma is anterior in the horizontal plane since inserting on both sides of the midline *(dotted line)*. **B**. Sagittal view. **C–G**. Operative views. **C**. A C1 laminectomy has been performed with control of the left VA V3 segment. Bone resection must be extended over the midline *(arrow)*. Dotted line indicates the distal dural ring where the VA passes through the dura. **D**. View after dural opening. Contraincisions are performed at the level of the inferior aspect of the distal dural ring *(arrow)*. C1 posterior rootlets are as well observed as the dentate ligament. The tumor displaces the neuraxis. **E**. The inferior pole of the lesion has been resected. The left VA V4 segment has been dissected, as well as the accessory nerve along the lateral aspect of the tumor. The hypoglossal nerve is visible. A blade retracts the cerebellum upward. **F**. The lower cranial nerves are found as expected at the superior aspect of the tumor. **G**. View after complete resection of the lesion. Both vertebral arteries are identified. The right PICA is visible *(white arrow)*. A feeding vessel arising from the left VA has been divided *(black arrow)*. All lower cranial nerves have been respected. Bl: blade. C1: atlas posterior arch. C1PR: C1 posterior rootlets. DL: dentate ligament. DM: dura mater. N: neuraxis. Occ: occiput. T: tumor. IX: glossopharyngeal nerve. X: vagal nerve. XI: accessory nerve. XII: hypoglossal nerve. V3l: left vertebral artery V3 segment. V4l: left vertebral artery V4 segment. V4r: right vertebral artery V4 segment.

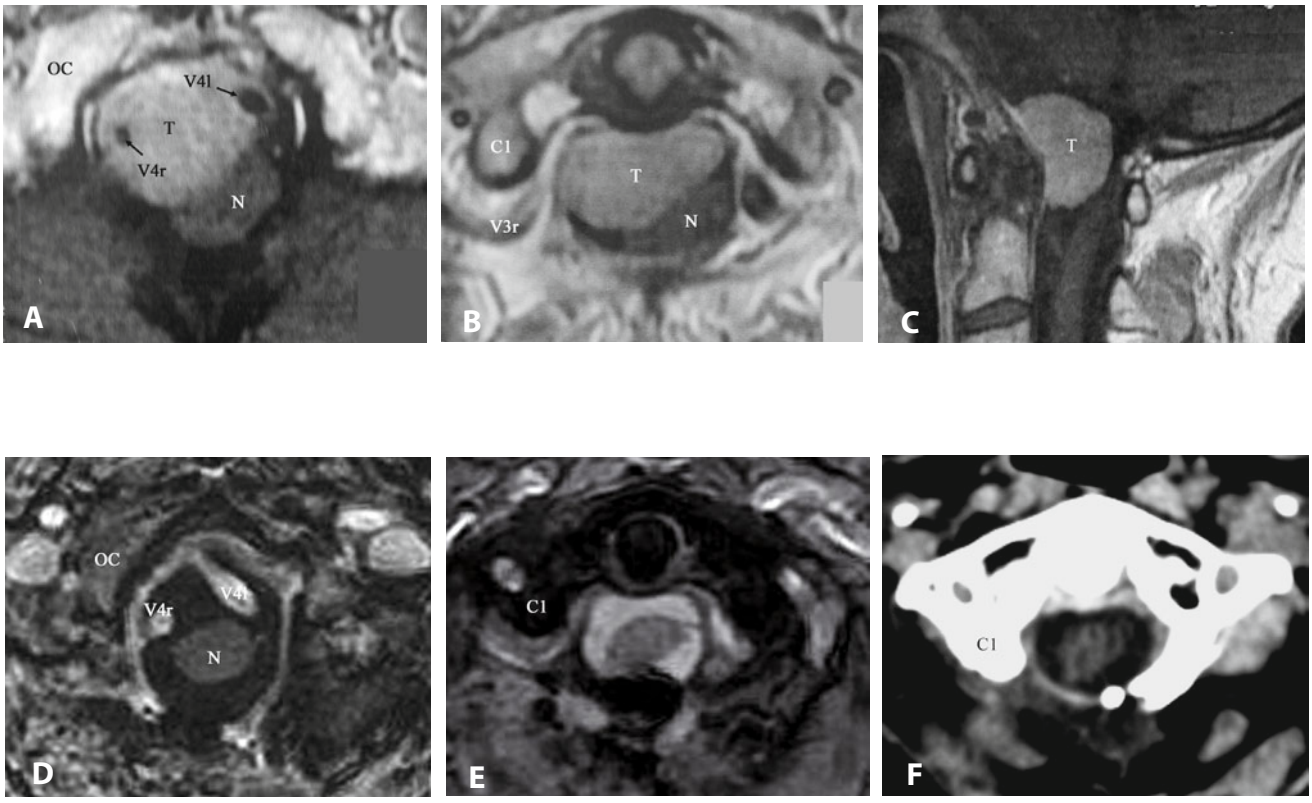


Fig. 4 – Anterior foramen magnum meningioma totally encasing the right VA. **A–C**. preoperative views. **A**. Axial T1-weighted MRI with gadolinium at the level of the occipital condyle. The right VA is completed embedded within the tumor. This characteristic worsens the surgical difficulty. The VAs are observed above the tumor. **B**. Axial view at the C1 level/VA V3 segment. **C**. Sagittal view showing the meningioma insertion. **D–F**. postoperative control. **D**. Same view as A after complete resection of the tumor. The neuraxis is well decompressed. **E**. Same view as **B**. **F**. Axial CT scan at the C1 level showing the extent of bone resection. C1: atlas. N: neuraxis. OC: occipital condyle. T: tumor. V3r: right vertebral artery V3 segment. V4l: left vertebral artery V4 segment. V4r: right vertebral artery V4 segment.

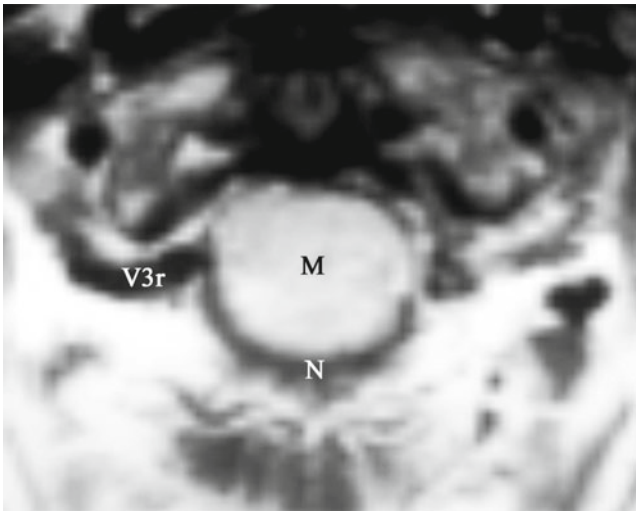


Fig. 5 – Huge anterior foramen magnum meningioma. The lesion is so large that the neuraxis is almost no more visible. N: neuraxis. T: tumor.

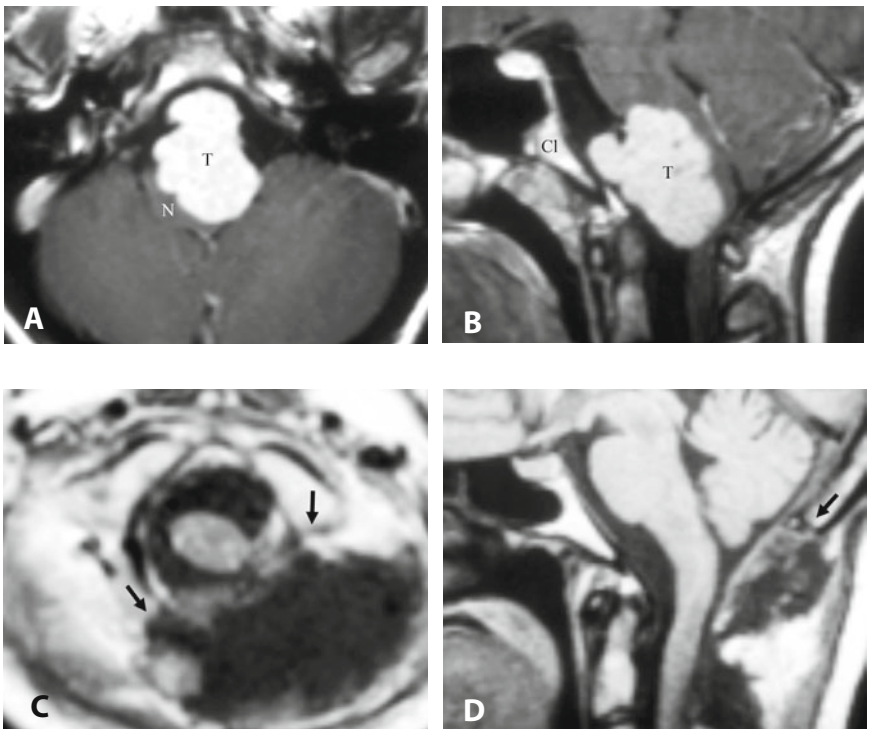


Fig. 6– Anterior foramen magnum meningioma with a base of insertion at the inferior aspect of the clivus. **A–B**. Preoperative workup. **C–D**. Postoperative control after complete tumor resection. The limits of the bone opening are indicated with the black arrows. Cl: clivus. N: neuraxis. T: tumor.

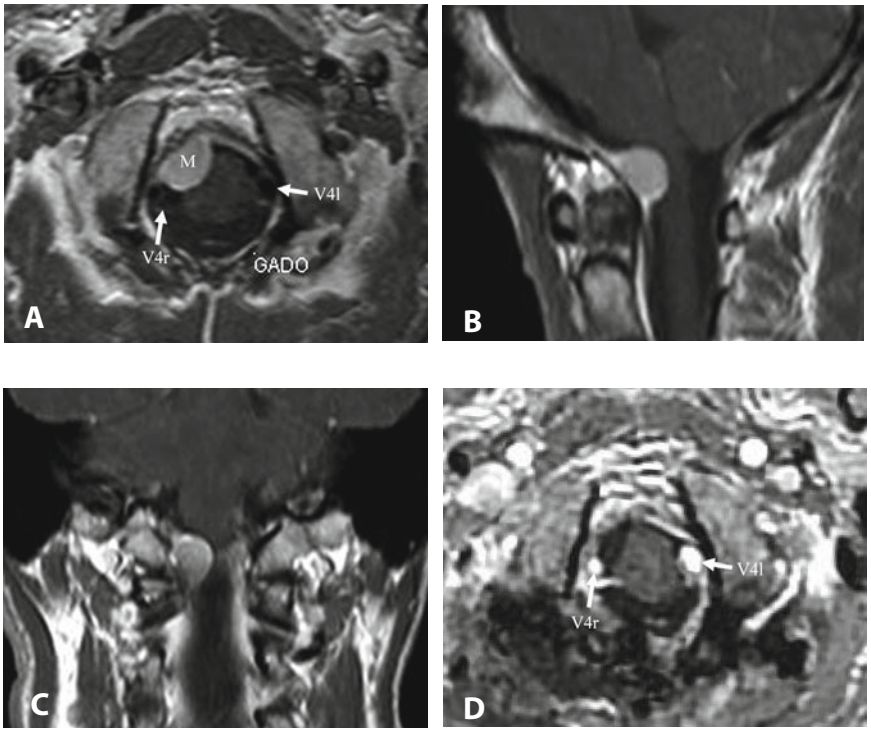


Fig. 7 – Lateral foramen magnum meningioma. **A–C**. Preoperative MRI with gadolinium in axial *(A)*, sagittal *(B)*, and coronal *(C)* views. **D**. Postoperative control after complete tumor resection. M: meningioma. V4l: left vertebral artery V4 segment. V4r: right vertebral artery V4 segment.

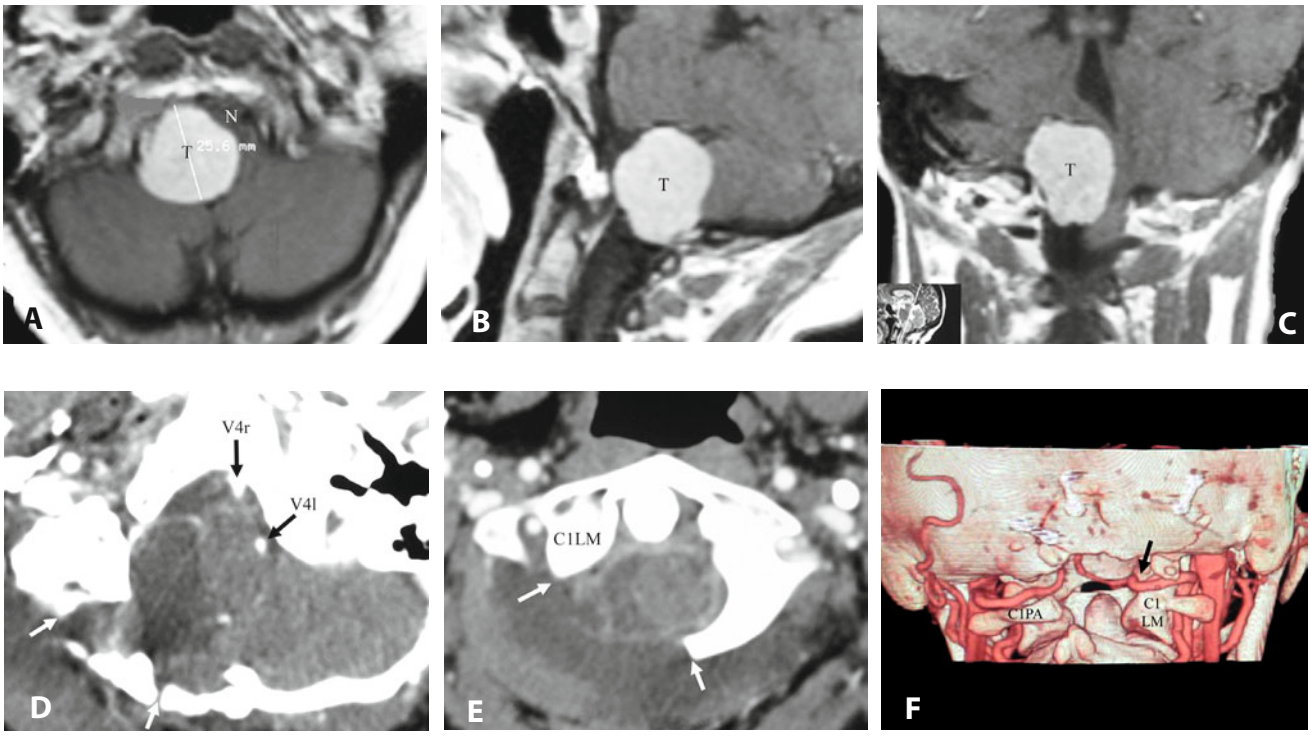


Fig. 8 – Large lateral foramen magnum meningioma. **A–C**. Preoperative pictures in axial *(A)*, sagittal *(B)*, and coronal *(C)* views. In A, notice the anterolateral displacement of the neuraxis because the lesion grows behind it, contrarily to anterior foramen magnum meningiomas. **D–F**. Postoperative control. **D.** Postoperative axial CT scans at the posterior fossa *(D)* and C1 *(E)* levels showing the extension of bone opening for the approach. One-fifth of the posterior aspect of the right C1 lateral mass has been removed to obtain a tangential access along the lateral dura mater. **F**. 3D angio-CT reconstruction showing the view that can be obtained by this approach if the neuraxis is displaced. The arrow indicates the level of the distal dural ring where the VA V3 segment enters the dura mater of the foramen magnum. C1 LM: lateral mass of atlas. C1PA: posterior arch of atlas. N: neuraxis. T: tumor.

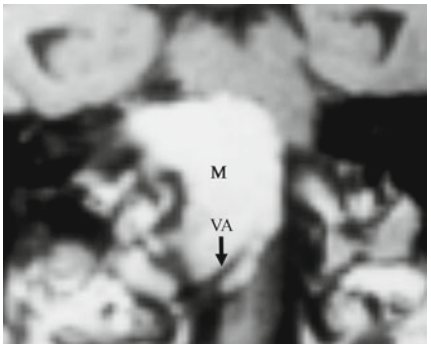


Fig. 9 – Huge lateral foramen magnum meningioma growing above the VA on coronal MRI. In this case, the position of the lower cranial nerves can not be anticipated. M: meningioma. VA: vertebral artery.

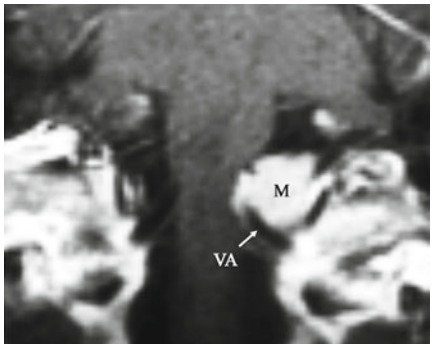


Fig. 10 – Lateral meningioma inserted above the VA on coronal T1-weighted MRI with gadolinium. The lesion is developed above the VA. M: meningioma. VA: vertebral artery.

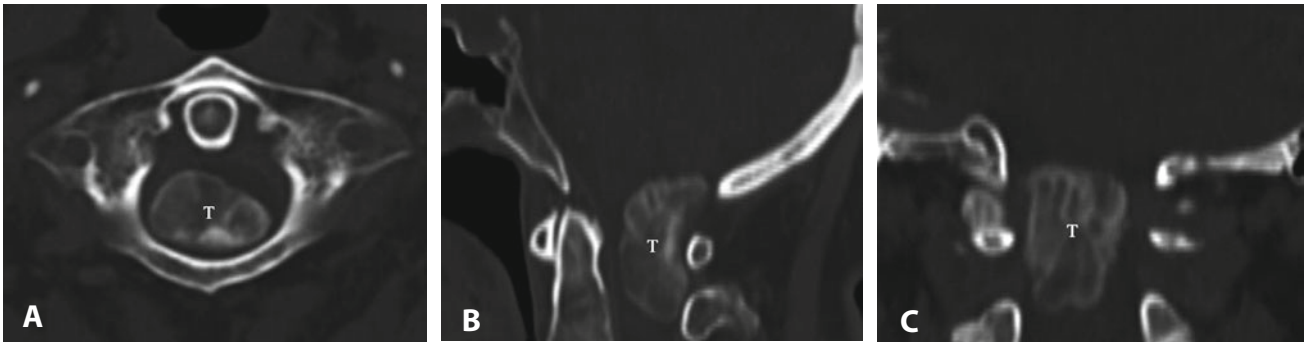


Fig. 11 – Posterior calcified meningioma. Axial (**A**), sagittal (**B**), and coronal (**C**) CT scans in bone windows. *Abbreviation :* T: tumor.

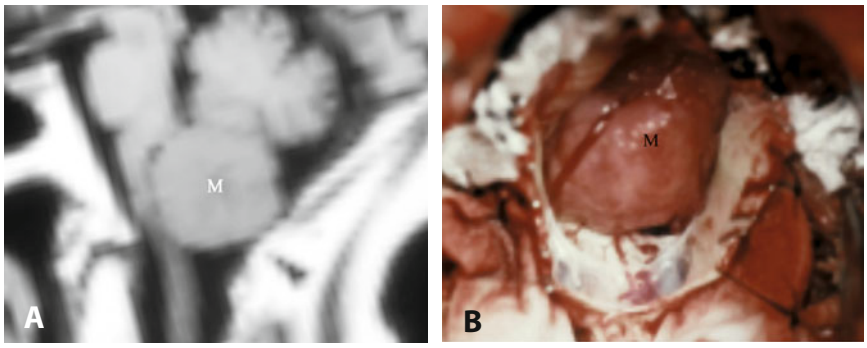


Fig. 12 – Posterior foramen magnum meningioma without dural attachment. **A**. Preoperative sagittal T1-weighted MRI. **B**. Operative view. M: meningioma.

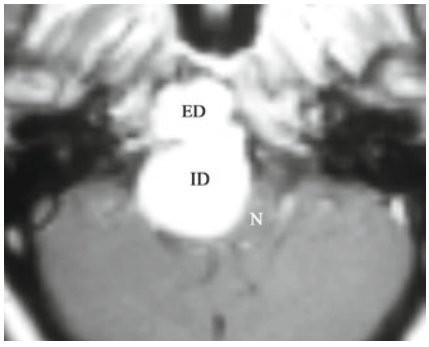


Fig. 13 – Intra-extradural meningioma invading the clivus. **A**. Axial T1-weighted MRI with gadolinium. N: neuraxis.

the cases of our series, these meningiomas developed in 78.8%, 16.3%, and 4.8% in the respective positions; thus, these are significantly more often located below the VA (Figs. 2–3). In this circumstance, the lower cranial nerves are always displaced cranially and posteriorly. This is an advantageous condition because as long as the tumor is resected from its caudal aspect, there is no need to look for the lower cranial nerves. The nerves will only come into view on reaching the superior tumoral part at the end of the procedure when tumor debulking has facilitated the dissection. The surgical condition is entirely different if the lesion grows above the VA because the position of the lower cranial nerves cannot be anticipated (Figs. 9–10). The nerves can be displaced separately in any direction and one has to look for them after partial debulking. Perioperative monitoring can be helpful in this condition. The same problem exists in presence of tumoral development on both sides of the VA. One supplementary problem exists in this latter condition: the dura around the VA penetration may be infiltrated by the tumor. Complete tumor resection then becomes hazardous because of the tumor involvement in an area where adherences between the VA adventitia and the dura are already present anatomically. In this case, rarely observed, we advise to leave a cuff of infiltrated dura around the VA and only to coagulate this zone.

Intraoperative monitoring

Intraoperative neurophysiological monitoring has been used by several surgeons (16, 17). This includes somatosensory evoked potentials, brain stem auditory evoked potentials, and electromyographic monitoring of lower cranial nerves, by recordings through endotracheal tube (CN X), and with needle in the sternomastoid (18) muscle (CN XI) and the tongue (CN XII).

Surgical approach

The simplest FMM is intradural with a posterior insertion. In this lesion, the best surgical approach is undoubtedly the posterior approach that is well known by all neurosurgeons and is associated with a low morbidity rate.

The choice of the appropriate surgical approach is discussed more often for lateral and especially anterior intradural meningiomas.

The transoral approach was sporadically reported 20 years ago (19–21). Although it provides access to the anterior part of the CCJ, this approach is associated with several important drawbacks in the presence of intradural lesions that have limited its use: the infectious risk, the cerebrospinal fluid (CSF) fistula risk, the rate of postoperative instability and velopalatine insufficiency are increased, and the access is poor to lateral extensions of the tumor (19, 21).

In the literature, the two main surgical approaches reported are the far lateral approach (also called posterolateral approach or lateral suboccipital approach) and the extreme lateral approach (also named anterolateral approach). As described by Rhoton (22), the far lateral approach is a lateral suboccipital approach directed behind the sternocleidomastoid muscle and the VA and just medial to the occipital and atlantal condyles and the atlanto-occipital joint. The extreme lateral approach is a direct lateral approach deep to the anterior part of the sternocleidomastoid muscle and behind the internal jugular vein along the front of the VA. In fact, both approaches permit drilling of the occipital condyle but provide different angles of attack.

Sen and Sekhar (23) were the first to report the extreme lateral transcondylar approach for FMMs in 1990, and several subsequent reports refined the technique (24–26). Salas et al. (24) reported in 1999 several variations of the extreme lateral approach based on a series of 69 patients, including 24 meningiomas. A partial transcondylar approach was used most of the time, while the transfacetal, retrocondylar, and extreme transjugular approaches were used more rarely. During the partial transcondylar approach, the posterior one third of occipital condyle and superior facet of C1 were drilled away. A partial mastoidectomy was also performed during the procedure (23, 24, 26). In 2000, Arnautovic et al. (16) published their experience on the basis of a series of 18 ventral FMMs treated through an extreme lateral transcondylar approach. The condyle resection ranged approximately from one-third to one-half, without causing any craniocervical instability. This approach requires VA transposition (16, 23, 24, 26–28).

As other surgeons (13, 28–37), we advocate rather the posterolateral approach, also called far lateral approach, even for anterior intradural FMMs. During this approach, the VA has to be controlled in the horizontal portion of the V3 segment, above the C1 posterior arch but VA transposition has not to be performed. It has only been reported in two series (38, 39).

The extent of FM lateral wall drilling is variable, in fact directly proportional to the tumor extension to

the contralateral side. In the literature, the occipital condyle resection varies between 0% and 66% (13, 28, 31, 32, 38, 39). Recently, Bassiouni et al. (37) classified judiciously surgical approaches in two groups: transcondylar or retrocondylar if the occipital condyle is not drilled. In our review of the literature (Table 4), we found 10 series in which the approaches to reach anterior or anterolateral meningiomas were retrocondylar (13, 17, 30, 32–34, 37, 38, 40, 41). In five of these series, a retrocondylar approach was not used in all cases but in 46.7–89.2% (13, 33, 34, 38, 40). The five other series only included patients treated through a retrocondylar approach (17, 30, 32, 37, 41). Of these, complete resection in 100% of the patients was reported in three studies (30, 32, 41). In the two others, complete resection was noted in 90% and 96% of the cases and the remaining lesions were resected subtotally (17, 37). Surgical results were excellent and surgical morbidity and mortality rates low (17, 32, 37, 41).

Group I: intradural FM meningiomas

Posterior meningiomas

The midline posterior approach is the approach of choice for posterior meningiomas, irrespective of their intra- and extradural extensions, if the tumor remains posterior to the plane of the dentate ligament and medial to the VA (1, 15). After posterior dura opening, the tumor will come into view hiding the neuraxis, which is displaced ventrally by the lesion.

The midline posterior approach

The patient may be placed in the sitting, ventral, or lateral position. We prefer the sitting position to decrease venous bleedings as far as there is no contraindication such as a patent foramen ovale. Air embolism has to be prevented by hypervolemia, G-suit, esophageal echodoppler, and venous hyperpressure tests during the procedure.

The skin is incised on the midline from the occipital protuberance down to the upper neck. The midline avascular plane is incised between the posterior muscles, up to the occipital protuberance and down to the spinous process of C2.

For this surgery, the VA is rarely concerned. Only in case of lateral extension, the oblique and the lateral aspect of the horizontal portions of the VA V3 segment have to be elevated subperiosteally before bone resection. This is carried out using drill and Kerrison rongeurs; it is always limited to the lower part of the occipital bone and the posterior arch of the atlas. The dura is then incised in a T- or Y- shaped fashion and retracted with stitches.

The VA V4 segment is not at risk because it is pushed anteriorly at some distance from the meningioma, with the neuraxis in between, except laterally after the exit of the VA from the dura, where the meningioma can be in contact with the posterior aspect of the VA for few millimeters. The VA location has to be anticipated in this condition by pursuing the course of the VA V3 segment through the dura mater.

Lateral and anterior intradural FM meningiomas

We mainly use the far lateral retrocondylar approach (posterolateral approach) for any intradural process located laterally and/or anteriorly to the neuraxis and thus especially for intradural FMMs (1, 3, 4). In some cases, the drilling of the FM lateral wall has to be performed for intradural anterior meningiomas but remains limited at the utmost to the medial 20% of the FM lateral wall. Whatever, this extent of drilling has to be tailored according to the tumor characteristics. We consider nevertheless that more bone resection is never necessary due to the anatomical anterior position of the lateral mass of the atlas and the occipital condyle. Our attitude has been reinforced by cadaveric study that has demonstrated that increasing the bone drilling is not associated with a significant widening of the surgical corridor (32). In fact, resecting one-third and one-half of the occipital condyle increases the visibility by 15.9° and 19.9°, respectively. Two anatomic reports have demonstrated that condyle resection allows a wider angle of exposure to gain the anterior foramen area (42, 43). However, these studies did not consider the fact that in surgical approaches to anterior lesions, space-occupying lesions enlarge the surgical corridor. We consider that the extreme lateral approach could be associated with a higher morbidity rate than the far lateral approach. The exposure allowed by the far lateral retrocondylar or partial transcondylar approach is adequate for resecting even anterior intradural FMMs. The supposed benefit in term of exposure provided by the extreme transcondylar approach is counterbalanced by the risks associated with the CN XI dissection, the VA transposition, and the condyle drilling.

The posterolateral approach has also the advantage to allow a bilateral approach in the same stage for tumors extending far beyond the anterior midline.

Posterolateral approach (Illustration 2)

The surgical technique for the resection of lateral and anterior FMMs through a posterolateral approach is described in detail in the chapter 28. For this reason, we have limited our description to the main principles.

The patient is in the sitting, supine, or even lateral decubitus position. Incision is along the midline from the C4 level to the occipital protuberance, then curved laterally along the superior occipital crest toward the mastoid process. Occipital bone and posterior arch of the atlas are resected on both sides but much more on the tumor side. Contralateral bone resection is achieved in order to avoid compression of the neuraxis against the bone during tumor manipulation. The VA groove is exposed up to the occipital condyle and C1 lateral mass by subperiosteal dissection so that the VA and perivertebral venous plexus are kept in their periosteal sheath. Venous bleeding must not be a problem around the VA, while it can be troublesome from the C1C2 intervertebral space around the C2 nerve root.

Then, the dura is opened in a curvilinear fashion with the vertical segment a little away from the midline so that the neuraxis is kept covered and protected by the dura. Then it is safe to start by the section of the two first arches of the dentate ligament (around the VA and the second cervical nerve root). This releases some tension on the neuraxis. Similarly, and for the same reason, it is wise to cut the first cervical nerve root lateral to the spinal root of the accessory nerve. Doing so maximizes access to the tumor. The tumor is debulked starting at the inferior pole, from where the main vascular supply comes and progressing upward.

Groupe II: intra-extradural FM meningiomas

Few meningiomas belong to this subgroup. Only three cases out of 107 tumors in our series were treated, which represents an incidence of 2.3%.

Among these intra-extradural meningiomas, one was posteriorly located with extension into the cervical muscles; one was anterior, extending into the clivus; and one was lateral along the VA at the level of its dural penetration. All cases were treated by a posterolateral approach.

The anterolateral approach could be justified in this group if the extradural component of the tumor grows anteriorly to the VA V3 segment, such as that discussed in the next section. Posterior extension to the VA and anterior midline invasion into the clivus remains ideally treated by a posterolateral approach.

Groupe III: extradural FM meningiomas

Extradural lesions can be treated by either the posterolateral or the anterolateral approach depending of the tumor relation with the VA V3 segment. The anterolateral approach with VA transposition was merely performed in selected cases of FMMs with extradural extension and never for intradural lesions.

The extent of drilling of the FM lateral wall during this procedure can be larger but is only dictated by the tumoral invasion. Nevertheless, it must be only limited to the destroyed or invaded bone. In such a way, the question of instability is only a preoperative concern. On the basis of the experience of tumors located at the CCJ, instability was never observed when less than half of the C0–C1 and C1–C2 joints were resected (44).

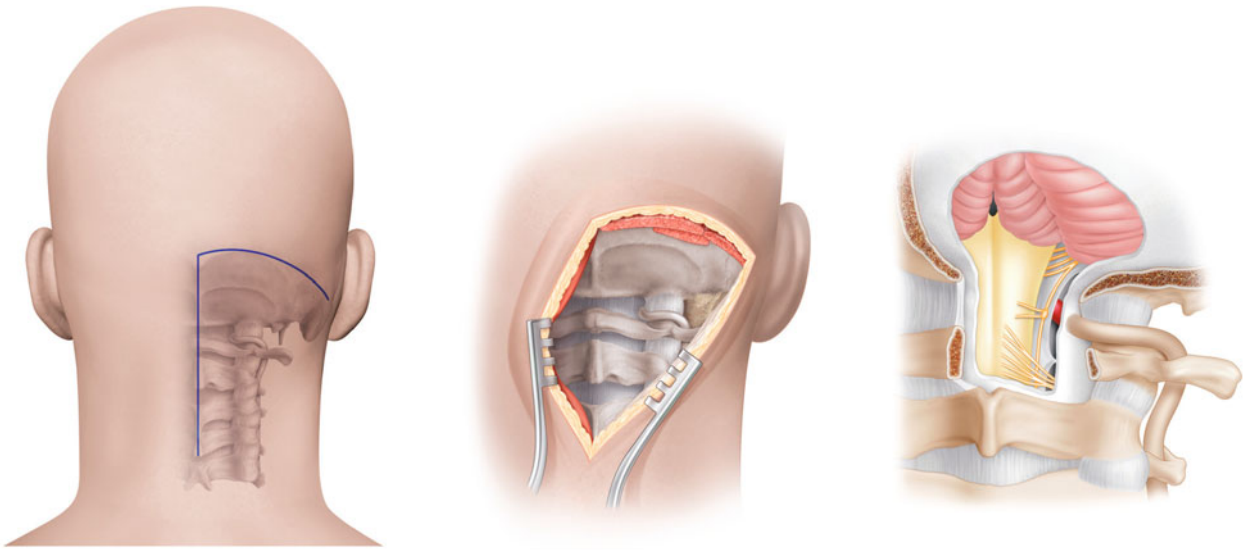


Illustration 2 – The postero-lateral approach.

Clinical results

Our experience was published in 1997 after the treatment of 40 FMMs operated during the period 1980–1993 (3). Thirty-four lesions were intradural, the other were extradural or intra-extradural. Of these, 18 were considered anterior, 21 lateral, and 1 posterior. The tumor was above the VA in four cases, below in 20 cases, and on both sides in 16 cases. The posterolateral approach was used in 31 cases, the anterolateral one in five, and the posterior midline in four. The rates of complete resection for intradural and extradural lesions were 94% and 50%, respectively. Postoperatively, the clinical condition improved in 90% of patients, remained stable in 2.5%, and worsened in 7.5%.

Our present series of intradural meningiomas consists of 101 tumors. To the series were added three cases of intra-extradural lesions and three purely extradural ones. Most of these meningiomas were referred at first presentation, but six patients were recurrent cases previously operated on elsewhere. There were two operative deaths in the first years of this series due to pulmonary and air embolisms respectively. Worsening of the preoperative condition was noticed in three patients, all of them related to swallowing difficulties in meningiomas located above the VA. Except the six recurrent and two primary cases, resection was total and confirmed by postoperative MRI. Follow-up includes for most patients MRI scans at 1, 3, 6, and 10 years. In the group of primary cases, we are aware of only one recurrence at nine years after surgery. At the first surgery, the meningioma had a usual (mushroom) type; at recurrence, it was *en plaque* type, involving half of the circumference of the FM dura and bone.

To the 104 intradural meningiomas must be added three cases entirely extradural operated on through an anterolateral approach.

Morbidity – mortality – prognosis factors

Series reported in the literature from 1924 to 1976 were reviewed by Yasargil et al. who noted, based on 114 cases, an overall mortality rate of approximately 13% but which could become as high as 45% in some series (45). The overall mortality reported over the last 20 years has lowered to 6.2%, and the mortality rate is comprised between 0% and 25% (Table 4). Mortality rates higher than 10% were mainly observed in small series (23, 26, 28, 30, 39, 40).

In the Yasargil's review, a good outcome was noted in 69%, and a fair and poor outcome in 8% and 10%, respectively. In series larger than 10 patients published over the last 20 years (Table 4), improvement, stability, and worsening of the neurological status were reported in 70–100%, 2.5–20 %, and 7.5–10% of the cases, respectively. The permanent morbidity rate was comprised between 0% and 60%. The rate of permanent morbidity rate after far lateral approach (either transcondylar or retrocondylar) of 0–17% was lower than the rate of 21–56% through an extreme lateral transcondylar approach (considering series without recurrent tumor). The most frequent preoperative deficit affects the lower cranial nerves. Hopefully, these deficits were prone to recover even completely postoperatively (16, 26, 37), except in cases of en plaque meningiomas or recurrent tumor (13).

The morbidity rate may be adversely influenced by several factors which make the surgical procedure still more difficult: the anterior tumor location (3, 13), the tumor size (smaller lesions are more difficult to resect because of a small surgical corridor), the tumor invasiveness, the extradural extension (3), the VA encasement (46), the absence of arachnoidal sheath (12, 37, 47), and adherences in recurrent lesions (13, 23, 26, 37).

Rate of tumoral resection

A multicentric study from 21 hospitals published by George et al. (2) reported complete, subtotal, and partial removals in 77%, 16%, and 7% of the cases, respectively. Over the last 22 years, most of the studies reported complete or subtotal removal of the tumor (2, 3, 17, 21, 23–28, 30–41, 46). Adherences of the lesion to vital structures, VA encasement, and invasiveness of the lesion are factors limiting the resection completeness. Adherences are encountered during repeated surgery and explained the lower rate of complete tumor resection (60–75% of Simpson grade 1) in surgical series in which a high proportion of recurrent tumors are included (13, 16, 23). In recurrent tumors, it may be advised to leave a small tumor along the medulla and lower cranial nerves to preserve a low morbidity rate (3, 37), while Arnautovic et al. (16) favors radical removal of recurrent tumors with the goal of providing a relatively long and stable postoperative course, even at the price of frequent but transient morbidity induced by lower cranial nerves dysfunction. We have also experienced like Arnautovic et al. (16) that the rate of complete removal is higher at first surgery than when treating recurrence. For this reason, it is justified to be aggressive at the first surgical presentation. VA encasement was noted in 38–59% in some series (3, 13, 31, 34, 37). This factor was recognized as an independent factor of incomplete removal (13). The location of the meningioma, either intra-extradural or extradural, reflects tumoral invasiveness. For this reason, these tumors are less favorable for complete

Table 4 – Review of the literature of foramen magnum meningiomas series.

Author	Year	Nb pt	FMM location			Recurrence	VA encasement	Approach	VA transp	Res. JT	Partial mast	Nb CR	Extent CR	Instability	Outcome			Resection		Transient morbidity	Permanent mobidity	Mortality	FU Months	Recurr
			Ant	Lat	Post										Impr	Unch	Wors	Total	Subtotal					
Gilsbach	1987	5		100%			-	FL		+			1/3					100%	0%		20%	0%		
Guidetti	1988	17	82.4%															100%			12%	11%		
Sen, Sekhar	1990	5	80%	20%		80%	-	EL	+	+	+	100%	1/3-1/2	0%	20%	20%	60%	60%	40%		60%	20%		
Crockard	1991	3	100%			33%	33%	TO	-			0%	0	33%			100%	0%	66%	100%	100%	66%	20.6	33%
Kratimenos	1993	8	100%			12.5%	-	FL	+				1/3	0%				87.5%	12.5%		0	25%	–	–
Babu	1994	9		100%				EL	+	+	+	100%	1/3-1/2	0%				88.8%	11.2%	78%	56%	11.1%	9.4	0%
Akalan	1994	8	12.5%	87.5%				PM				0%	0		88%	12%		100%			0%	0%		
Bertalanffy	1996	19		100%			-	FL SO TC		+		100%	1/3	0%				100%	0%		0%	0%	–	–
Samii	1996	38		95%	5%	5%	40%	PM, LSO				17.5%	1/3	0%				63%	30%	37%	5%	6%	21	5.01%
George	1997	40	45%	52.5%	2.5%		38%					5%	Partial	0%	90%	2.5%	7.5%	87.5%	10%		0%	7.5%	57.6	0%
Pirotte	1998	6		100%			-		+			100%	1/2-1/3	0%				100%	0%		17%	17%		
Sharma	1999	10	50%		50%			PM, FL				0%	0	Yes				100%				15%		
Salas	1999	24		100%			-	TC / ELTJ	+		+	100%	1/3	0%				66%	33%		–	0%	14.8	–
Arnautovic	2000	18	100%			11.1%	-	TC EL TC	+			100%	1/2-1/3	0%	89%	11%		75%	12.5%	55%	11.1	16.6%	40	5.5%
Roberti	2001	21																76%	24%		21.5%	9.5%		
Goel	2001	17		100%			59%	SO				11.8%	1/3-1/4	0%	100%			82%	18%		6%	0%	43	0%
Nanda	2002	6	100%				-	FL				0%	0		100%			100%	0%		0%	0%		
Marin Sanabria	2002	7		72.5%	28.5%		-	TO, SO, TC	+			29%	1/3-1/2 <1/2	0% 0%	80%			100%	0%	72.5%	5%	14%		
Parlato	2003	7																86%	14%			0%	24	0%
Boulton	2003	10	60%	10%	30%							0%	0		70%	20%		90%	10%	40%	10%	0%		
Pamir	2004	22		91%	9%		40%	FL				95%	1/3	0%				95.5%	4.5%	27%	4.5%	0%	40	0%
Margalit	2005	18		100%				Lat				50%	partial (9/18)									0%		
Bassiouni	2006	25	32%	57%	11%	4%	43%	FL				0%	0					96%	4%	40%	8%	4%	73.2	0%
Borba	2008	15	53.4%	46.6%		0%		FL	33%	+		53.4%	1/3-2/3		100%			80%	13.3%	13.3%	0	0%	23.6	0%

CR: condyle resection; EL: extreme-lateral; FMM: foramen magnum meningioma; FL: far lateral / postero-lateral; FU: follow-up; JT: jugular tubercle; mast: mastoidectomy; Lat: lateral; Nb: number; pt: patient; Recurr: recurrence; SO: suboccipital; TC: transcondylar; TO: transoral; transp: transpositiion; VA: vertebral artery; +: yes; -: no

resection than pure intradural lesions (19), as demonstrated in the French cooperative study (2) in which the rate of complete removal of intradural, extradural, and intra-extradural meningiomas was 83%, 50%, and 45%, respectively.

Nerve sheath tumors

General features

Peripheral NSTs encompass schwannomas, neurofibromas, neufibrosarcomas, hemangioblastomas, and rare malignant variants. Their development at the levels of the first two cervical nerve roots is rare. These lesions represent approximately 5–10% of all spinal NSTs and 18–19% of the cervical localizations in the few published series (48–51). C1- and C2-NSTs are most often included in series of spinal NSTs or FM tumors (46, 49, 50, 52–58). Despite exhibiting specific features such as multiplicity, hourglass formation, and relationship with the VA, rare studies were specifically dedicated to this subject (48, 59). Multiplicity is a particular characteristic of NSTs (4–17%) and is related to the incidence of neurofibromatosis (48). The literature also has two cases of neurofibrosarcomas with evidence of von Recklinghausen's disease (58, 60).

Location on the C-2 nerve root is much more common than on C-1; C-2 and C-1 were both noted by Yasuoka et al. (61) in 18 and 1 of their cases, by Guidetti and Spallone (46) in six and three patients, and by George and Lot (48) in the French cooperative study in 44 and 6 tumors, with seven patients suffering from multiple tumors (six with bilateral C-2 neurinomas and one with a C-1 tumor plus bilateral C-2 neurinomas). Of note, in several cases, intradural schwannomas are diagnosed, but association with any root is not identified (48, 62). Indeed, a melanotic intradural schwannoma was found adherent to the surface of the spinal cord but no rootlet of origin was observed.

Our series of NSTs of the CCJ is composed of 75 cases, divided into three groups according to their compartment of development. Group I is constituted of 31 purely intradural lesions, group II of 19 dumbbell-shaped tumors, and group III of 25 purely extradural NSTs. The histological diagnoses were mainly schwannoma and neurofibroma. One of the tumors in our series was probably radiation-induced and was identified as a sarcoma when it recurred 1 year after resection. This occurrence is rare, and only two cases of radiation-induced neurinoma of C-2 were reported in the literature by Rubinstein et al. (63). The rare diagnosis of melanotic schwannoma was encountered in one case from our series. At this level, only few cases have been published (64, 65). Melanotic schwannomas are a rare variant of schwannoma composed of melanin-producing cells with ultrastructural features of Schwann cells (64). These lesions have to be distinguished from malignant melanoma for their management (64). In a recent large review of the literature, these tumors were demonstrated to be prone to local recurrence, malignant progression, leptomeningeal spread, and metastasis (66). In this condition, as in others (64, 66), we consider that complete resection must be attempted and strict follow-up is mandatory.

All tumors were resected completely except the melanotic schwannoma located at the level of the C-1 nerve root. Subtotal resection was only possible in this case because of invasion of the pial covering of the spinal cord and medulla oblongata. Histological examination showed no sign of malignancy and therefore no adjuvant treatment was proposed.

All over the series, no recurrence was noted during the follow-up period, except in the case previously described, even of the subtotally resected sarcoma after 2 years of follow-up monitoring.

The relationship with the VA could be demonstrated in 23 cases by CT or MR imaging and was confirmed by angiography, which showed displacement of the VA in 16 cases and stenosis in three cases. With a likely diagnosis of neurinoma, angiography is certainly not necessary. A complete and precise MR imaging with an angiographic MR study is sufficient to give the information necessary to appreciate the relationship with the VA and to choose the appropriate surgical technique.

Surgery

Group I: intradural NSTs

In our series of NSTs, most lesions (31/75, 41%) were purely intradural. Almost the same proportion (46%) of purely intradural tumors has been noted on a large series of spinal schwannomas (50). This high proportion has not been always the rule; in the French cooperative study (48), the rate of purely intradural lesions was 16%, and in the Jinnai et al. (49) publication, it was only 7%. Intradural NSTs at this level can be subdivided according to their origin from the C-1, C-2, or spinal accessory rootlet. In few cases, no origin is observed despite careful analysis.

Symptoms related to intradural FM NSTs are related to high cervical cord compression (48). Two reports in the literature described presentation by a subarachnoid hemorrhage from a C-2 intradural NST and a spinal accessory NST (67, 68). One case of acute quadriparesis was also due to intratumoral hemorrhage (51). Patient

age at diagnosis of a C-1 or C-2 neurinoma is similar to that of other locations in the spine (48). Diagnosis in patients under 20 years old is very often associated with von Recklinghausen's disease (48).

Purely intradural NSTs may grow laterally, anterolaterally, or posterolaterally to the spinal cord according to their size and their rootlet of origin. Most often, the lesion originates from the sensory root of the spinal nerve and occasionally from its motor root (18, 69). In some cases, the tumor comes from both roots (70).

The approach of choice for the resection of FM NST is the posterolateral approach. Using this, all of these tumors can be resected efficiently. Contrary to intradural anterior meningiomas, anterior extension of NSTs is never problematic because the lesion is not adherent to the dura. Therefore, after resection of the lateral part of the tumor, the lesion can be gently tracted laterally. Using a step-by-step lateral retraction technique, complete resection can be obtained without difficulty.

C2-NST

Patient positioning

Most of the time, the patient is placed in the prone position for this surgery. The back end of the table has to be elevated of 20° to decrease the venous pressure. The head is fixed with a Mayfield headholder and is slightly flexed. Overflexion must be absolutely avoided in tumors extending anteriorly to the spinal cord to avoid increased spinal cord compression. Long-tract monitoring can be relevant at this moment to prevent neurological worsening.

Approach

A standard midline approach more extended on the tumoral side is used. A paramedian dura mater incision is performed on the side of the tumor.

Tumor resection

Large C2-NSTs have to be debulked to become mobile, thereby facilitating the dissection from the spinal cord and the VA. The tumor surface has to be coagulated and incised inferolaterally. Afterwards, tumor reduction is obtained by piecemeal resection or with an ultrasonic aspirator. Any medial traction or movement must be avoided during this step before the spinal cord has been decompressed.

The tumor has to be finally separated from the spinal cord gently. Great care must be taken at this point, especially to avoid any damage to vessel supplying the spinal cord. Exceptionally, tumor resection has to be interrupted after subtotal resection and a tumor remnant left in place in case of subpial invasion of a malignant tumor. This situation was encountered in only one case.

Separation from the VA V4 segment is never problematic because, except in very large tumors, the VA is at some distance from the tumor. Because of important distal anastomoses, division of the proximal segment of the C-2 nerve root generally produces only a very mild sensory deficit, if any, and then in a very limited area. Moreover, distal branches were always divided in the posterior muscles with the posterolateral approach. This leads to hypesthesia or dysesthesia in the corresponding territory, which is the posterior region of the head.

C1-NST

Bone opening

A C1 laminectomy with resection of the inferior part of the occipital bone has to be performed. This step can only be carried out after subperiosteal control of the horizontal portion of the VA V3 segment above the C1 posterior arch as explained in Chapter 25. The laminectomy has to be extended laterally to permit lateral dura retraction after its incision. A C2 laminectomy is normally not useful since the tumor grows cranial to the C2 level.

Dura opening

Under microscopic magnification, a paramedian incision of the dura is performed vertically straight above the tumor.

The relation between the tumor and the VA must have been carefully analyzed on preoperative injected-CT scan or MRI. The close relationship between both requires a special consideration.

The principle of tumor resection is the same as the one explained above for C2 NSTs. Large tumors have to be debulked after coagulation and incision of the tumor lateral aspect.

When tumor reduction has been achieved, the dissection can be pursued on its outside aspect for separating it from surrounding structures, the VA, most of the time superoanteriorly, the CN XI nerve, and adjacent rootlets.

Complete resection is obtained after division of the C-1 nerve root. This is never problematic, as it does not produce any appreciable sensory deficit. Nevertheless, when the C-1 nerve root is cut intradurally, care must be taken to preserve the medullary root of the accessory nerve.

CN XI NST

Accessory nerve (CN XI) NSTs are developed along the intradural spinal segment of the nerve and therefore grow posterolaterally behind the dentate ligament and lateral to the neuraxis. The lower cranial nerves

are displaced superiorly and are found on top of the tumor. Consequently, the surgical technique is similar to the one used in case of FMMs of intradural, lateral and below the VA type. A posterolateral approach not much extended laterally is used except at the level of the jugular foramen.

CN XII NST

We have no experience of intradural CN XII NST, but the surgical technique is similar to CN XI tumors because these nerves are posterior to the VA and lateral to the neuraxis.

Hemangioblastomas

Excluding intra-axial lesions, they originate from the first or second cervical nerve root and are managed like neurinomas. However, they need generally an en bloc resection; a good control of the extradural VA is useful to divide the vascular supply before tumor resection.

Other types

Different tumors such as ependymomas or epidermoid cysts are usually developed posteriorly in the cisterna magna and consequently only necessitate a standard midline opening.

Group II: intra-extradural tumors (Figs. 14–16)

Among all dumbbell-shaped tumors we have treated at the CCJ level, NSTs were the commonest with 19 tumors out of 22 (86.3%). This group of patient represents 25.3% (19/75) of all patients treated for a CCJ NST.

The extradural component of the tumor can be responsible for displacement and even stenosis of the VA V3 segment. The displacement has to be very carefully analyzed on preoperative exams. Bone erosion induced by chronic compression has also to be studied because their protective effect during the dissection can no more be present when the bone is eroded (Figs. 14 G and I).

When the tumor is not too large, lateral access to normal portion of the root is possible. The C-2 nerve root is easily exposed as it separates into anterior and posterior branches while curving around the VA V3 segment, and it has a long course between the dural sac and the VA. Therefore, it is almost always possible to control the C-2 nerve root distal to the NST. The C-2 nerve root was always entirely involved in cases of NST with an extradural component, so it must be divided at the level of its main trunk or sometimes more distally at the level of its two branches. On the other hand, identification of the extradural and extraspinal portions of the C1 nerve root is always difficult between the VA and the groove of the atlas and is not essential. The C1 nerve penetrates the dura with the VA, through a funnel-shaped foramen, coursing at this level along the posteroinferior aspect of the artery (71). While passing intradurally, the ventral root of C1 is located anteriorly to the dentate ligament (71). The dorsal root of C1, when present, passes posterior to the ligament (71).

The surgical strategy is always to start working on the extradural component after VA control through a posterolateral approach. According to the lateral extension, the VA is exposed as far as the C1 transverse foramen and even sometimes in its C1-C2 segment.

C2-NST

As soon as possible, the distal part of the C2 nerve root is divided, also suppressing the main vascular supply to the tumor from the radicular artery branches accompanying the root. Then, resecting the medial extradural aspect of a dumbbell-shaped NST allows for already decompressing the neuraxis at the beginning of the procedure. The dura mater can then be later opened and retracted laterally in the empty space created, before removing step-by-step the intradural component, which can then be progressively tracted laterally. This step is otherwise hampered by the mass effect of the extradural component.

The tumor is progressively resected toward the dura. In C2-NSTs, it is often useful to remove more or less completely the posterior arch of the atlas at the VA groove level. This allows for a safe control of the superior part of the tumor.

The extradural part of the tumor often passes throught the dura but, and it even appears that some intra-extradural NSTs are in fact completely extradural.

The debulking of the extradural part always induces some bleeding from the C1C2 intervertebral venous plexus. This is sometimes very troublesome. It is important to control these bleedings by resorbable cellulose and bipolar coagulation before opening the dura.

The intradural part is then pulled out, but before that, the dura has to be cut all around the tumor at the level of its dural penetration. There is always a tight adherence between the tumor capsule and the dura because this is where the tumor originates. Gently pulling out the intradural part of the tumor raises no problem since there is always an excess of length of the proximal rootlets, which can be brought into view and cut. Moreover, there are never any adherences between tumor and spinal cord except in recurrent cases. The only problem may be the large size of the intradural component, which may require some debulking to permit its extraction.

So doing, there is a small defect of the dura that must be carefully repaired with a piece of aponeurosis sutured and sealed with fibrin glue.

Lateral tumoral extension beyond the vertical C1C2 segment of the VA is exceptional. No case of intra-extradural NST harbored this feature in our series. Only one case with strictly extradural extension has been observed. The extradural extraspinal extension was so large toward the cervical area reaching the carotid artery that it was removed in two stages; a posterolateral approach followed by an anterolateral approach a few days later allowed for a complete removal.

CN XII NST

CN XII NSTs are rare but often exhibit an hourglass form with intracranial, foraminal, and cervical extension. In this case, an anterolateral approach with some condylar drilling combined with a limited retrosigmoid approach has to be realized. The posterolateral approach is not suitable because it would need an extensive drilling of the condyle to reach the condylar foramen; moreover, the cervical part is out of reach. We have no experience of such neurinomas. Some cases have been reported in the literature including one in which they could even reconstruct the nerve with a nerve graft led through the enlarged foramen and sutured on both sides (72).

Group III: purely extradural tumors

Among 75 NSTs of our series, one-third was purely extradural. The technique applied for their resection is the same as that explained for the resection of the extradural component of dumbbell-shaped lesions. There is usually no risk for direct spinal cord injury in this circumstance and no risk of CSF leak. It must be underlined that often an extradural NST looks intra-extradural, as it extends sometimes beyond the midline; in fact, the dura is still covering the tumor and is pushed inside the spinal canal with severe spinal cord compression.

Bone tumors

Types

Bone tumors located at the CCJ level are rare (2, 73). They may be either primary or secondary. As already stated in the previous chapter, primary bone tumors may be subdivided in three subgroups: benign, benign locally aggressive, and malignant (74). From our series of 105 cases, chordomas, classified as locally aggressive tumors, were by far with 53 cases the most common type of tumors that we encountered. With 20 cases, metastases were second in frequency.

The most usual benign primary tumors were osteoid osteomas and osteochondromas, with seven and five cases, respectively. Some other types of tumors accounted for 20 cases, including other benign tumors such as aneurysmal cysts, histiocytosis, fibrous dysplasias, histiofibromas, or benign locally aggressive tumors like giant cell tumors, or malignant ones such as plasmocytomas.

Bone tumors of the OCJ raise several problems. First, their preoperative diagnosis remains difficult despite neuroradiological advances. Some apparently very destructive or invasive tumors are in fact actual benign tumors. Moreover, some lesions are rather pseudotumors requiring a special management. Second, their surgical resection is challenging and must be as complete as possible for oncological reasons. Third, extensive tumor infiltration and large resection may raise the problem of the spinal stability, which can be compromised. The adequate strategy may therefore be difficult to decide. Misdiagnosis of malignant tumors may lead to aggressive resection instead of simply decompressing a pseudotumor. Biopsy may be helpful to the decision but is not always reliable.

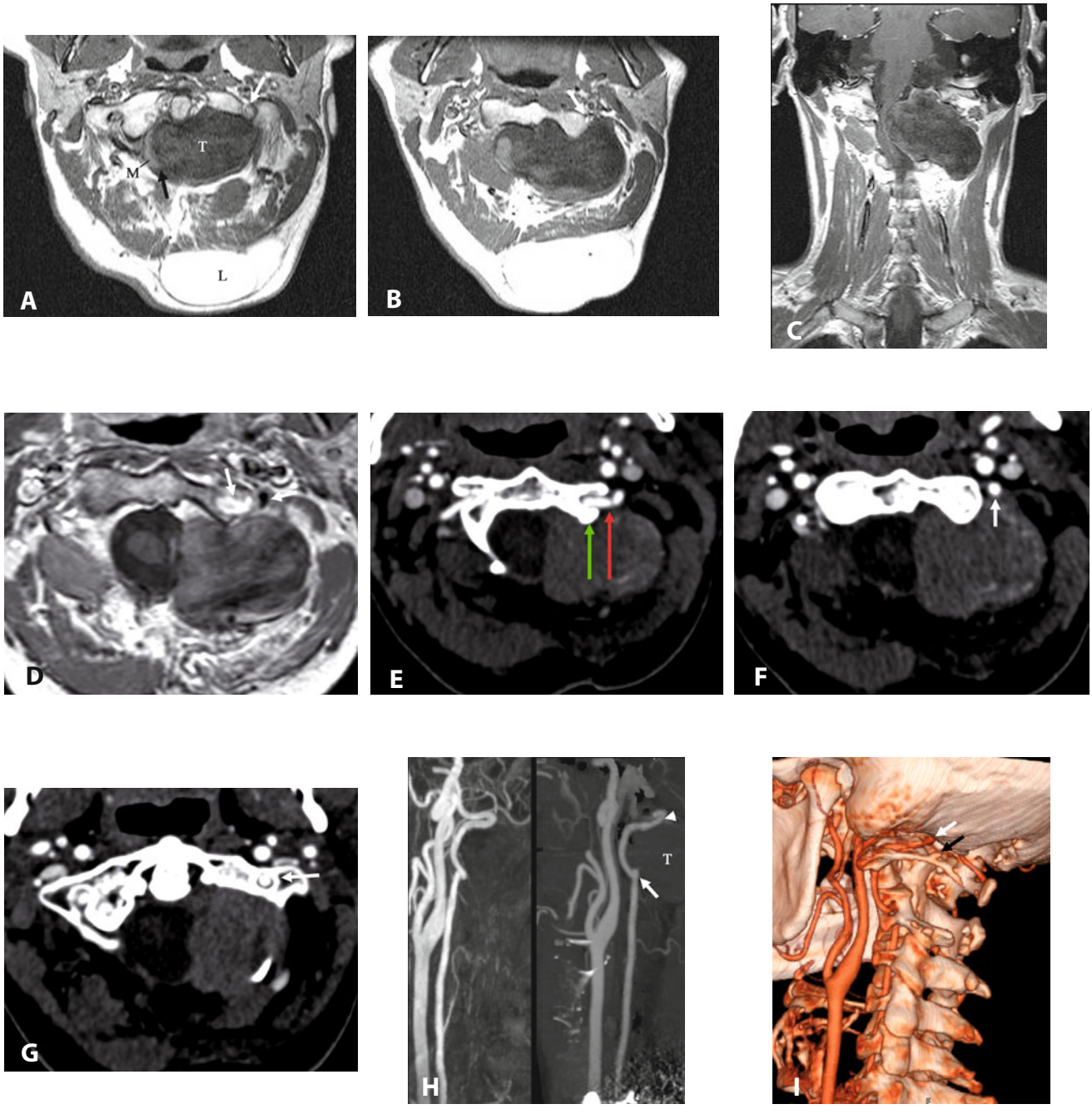
Preoperative imaging

CT scan has a crucial role in analyzing all extradural lesions of group III, especially bone tumors. It reveals bone modifications induced by the tumors such as local destruction evoking a malignant process as well as erosion induced by a benign tumor. The radiological characteristics of some tumors are very suggestive for instance in case of fibrous dysplasia, osteochondromas, or osteoid osteomas that appear as a nidus encircled by a radiolucent area. Nevertheless, in some cases, the diagnosis remains uncertain.

The amount of bone and joint destructions and the involvement of adjacent structures, therefore, have also to be determined giving valuable information about the spinal stability and the need for concomitant spinal stabilization. Comprehension of the tumor location and extension is obtained by analysis not only of axial slides but also of coronal and sagittal reconstructions. In selected cases, 3D-CT reconstruction improves this understanding.

Careful analysis of the bone landmarks used during the surgical approach to the VA is one of the most critical points. Tumors in the surrounding of the C1 posterior arch or the C1-C2 transverse processes can erode or destroy them. Therefore, the surgical technique must be adapted accordingly.

MRI by its high soft-tissue resolution is valuable for analyzing extraosseous extension especially toward the neuraxis.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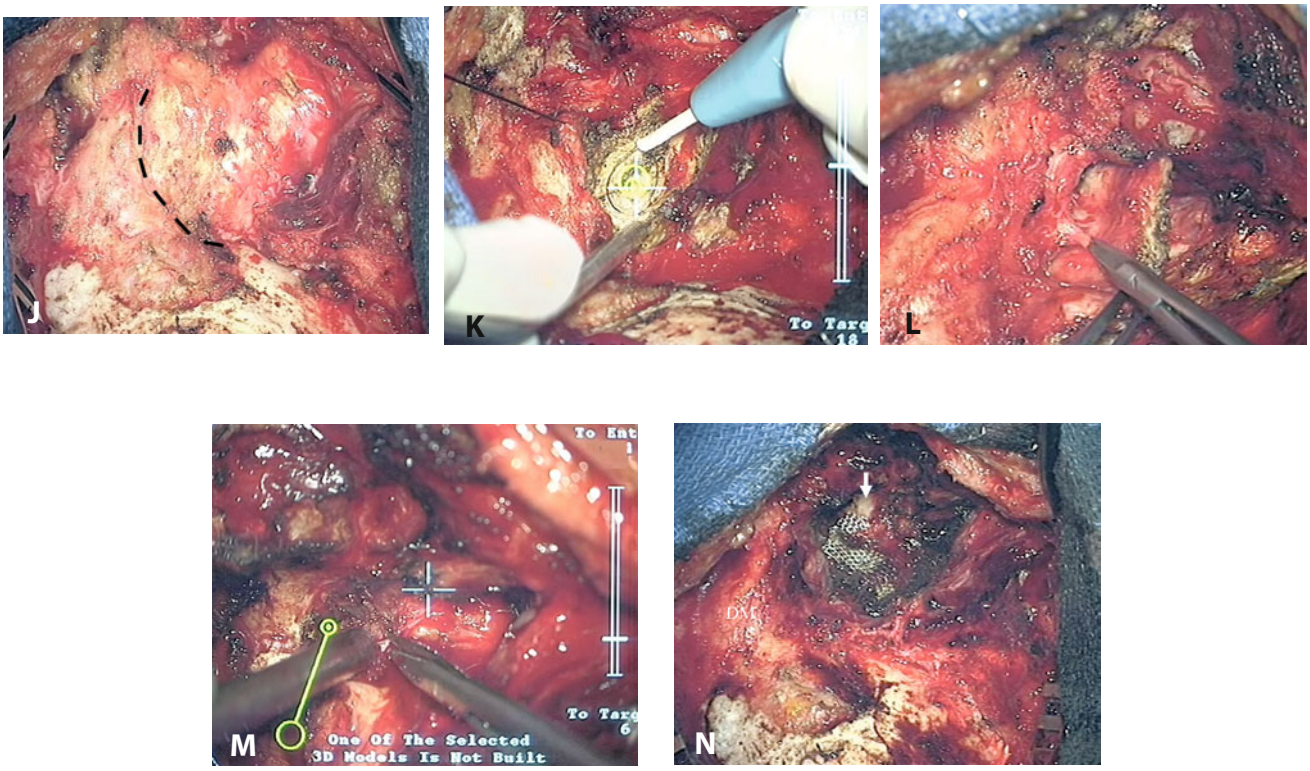
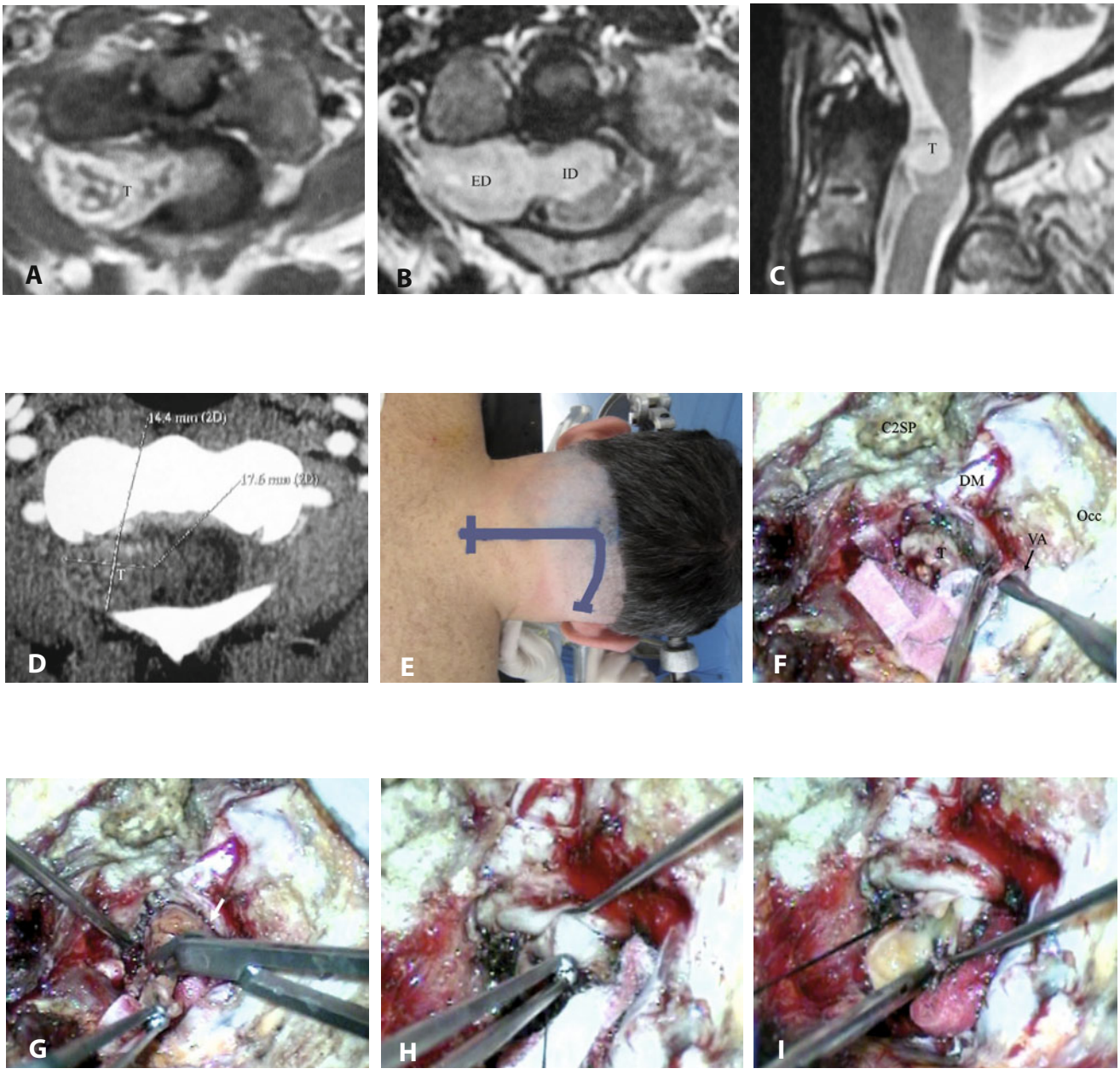


Fig. 14 – C1–C2 intra-extradural calcified neurofibroma. A 45-year-old man who suffered from cervicalgia since his twenties. Before removing a posterior neck lipoma, the plastic surgeon sent him for an MRI, which reveals the lesion. Neurological examination was normal. The patient was initially operated for the resection of the intradural component of the tumor. During the next 3 years, the extradural portion demonstrates a progressive growth. A second procedure was indicated with VA control due to the close relation of the tumor with the artery. **A–C**. MRI performed before the first surgery, in axial and coronal planes. The medulla is extremely compressed by a C1–C2 intra-extradural neurofibroma. *Black arrow:* the plane between the tumor and the neuraxis. *White arrow:* vertebral artery. **D–H**. Workup before the second surgery. **D**. Axial T1-weighted MRI. **E–G**. Injected CT scan at the C2 *(E)*, C1–C2 *(F)*, and C1 *(G)* levels showing the VA position. The main difficulties in this procedure, performed through a posterolateral approach, are that the VA is identified at the end of the tumor removal and vascular control cannot be obtained proximally. In this case, the tumor was particularly firm and could only be debulked with aggressive instruments such as a coagulation ansa. The crucial point is thus to avoid VA injury. In order to work more confidently, a navigation system was used to target osseous structures close to the artery that provide protection. Figure **E** illustrates this technical point. The goal was to follow the green trajectory targeting the bone. Conversely, the red trajectory could have been a wrong debulking plane exposing the artery to an inadvertent injury. **H**. VA displacement depicted on reconstructed angio-CT *(left)* and MRI *(right)*. *Arrow:* proximal extremity of the VA V3 segment. *Arrowhead:* VA distal extremity. **I**. 3D-angio CT reconstruction illustrating thinning of the C1 posterior arch *(black arrow)* due to the tumor. **J–N**. Operative views during the posterolateral approach. **J.** Dotted line indicates the interface between the tumor (on the right side) and the dura mater (on the left side). **K.** Debulking of the tumor. **L.** The intracanalar extradural portion of the tumor has been removed completely. **M.** Working laterally to the trajectory implies that the VA is very close. **N.** View at the completion of the tumor resection. Arrow indicates the VA level. DM: dura mater. M: medulla. T: tumor.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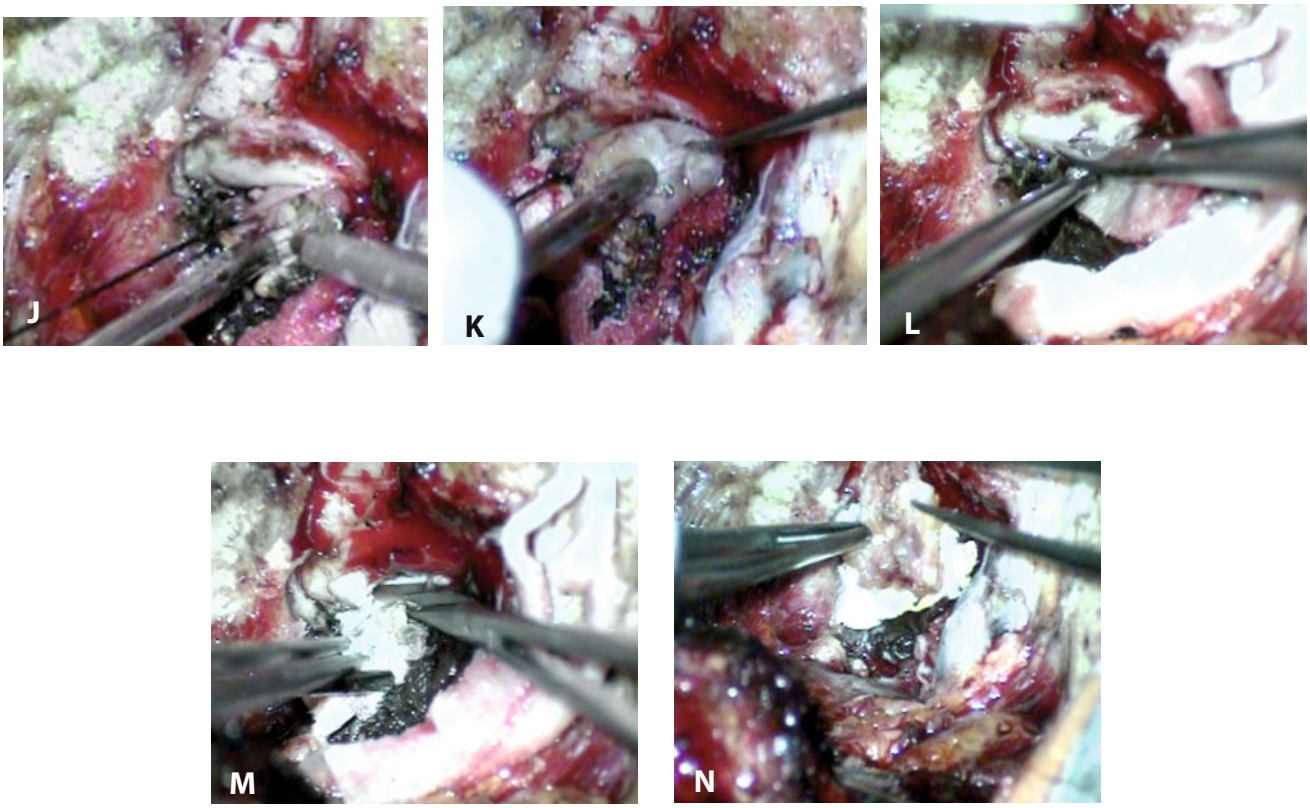

Fig. 15 – C1–C2 intra-extradural neurofibroma. **A–D**. Preoperative workup. **A**. Axial T1-weighted MRI with gadolinium. The tumor compresses the anterolateral aspect of the spinal cord. **B**. Axial T2-weighted MRI depicting the extradural and intradural portions of the tumor. **C**. Sagittal T2-weighted MRI. **D**. Axial injected CT scan. **E–N**. Operative views. **E**. Patient positioning and skin incision for a right-sided posterolateral approach. **F**. Subperiosteal dissection on the superior aspect of the C1 posterior arch. The VA V3 segment is visible above. **G**. The extradural portion of the tumor is resected piecemeal. *Arrow:* the interface between the tumor and the dura mater. **H**. The tumor is separated from the dura mater cautiously. **I**. The tumor capsule, corresponding to the nerve root sheath, is incised along its axis allowing resecting the tumor proximally. **J**. Use of ultrasonic aspirator is helpful for debulking the lesion especially in restricted areas. **K**. The tumor can then be tracted progressively laterally within the surgical field. **L**. The dura orifice is slightly enlarged to access the intradural compartment more easily. **M**. Tumoral rootlets are divided. **N**. Watertight closure is obtained by straightening the suture line with a muscular or fat patch. C2SP: Spinous process of axis. DM: dura mater. ED: extradural portion. ID: intradural portion. Occ: occipital bone. T: tumor. VA: vertebral artery.

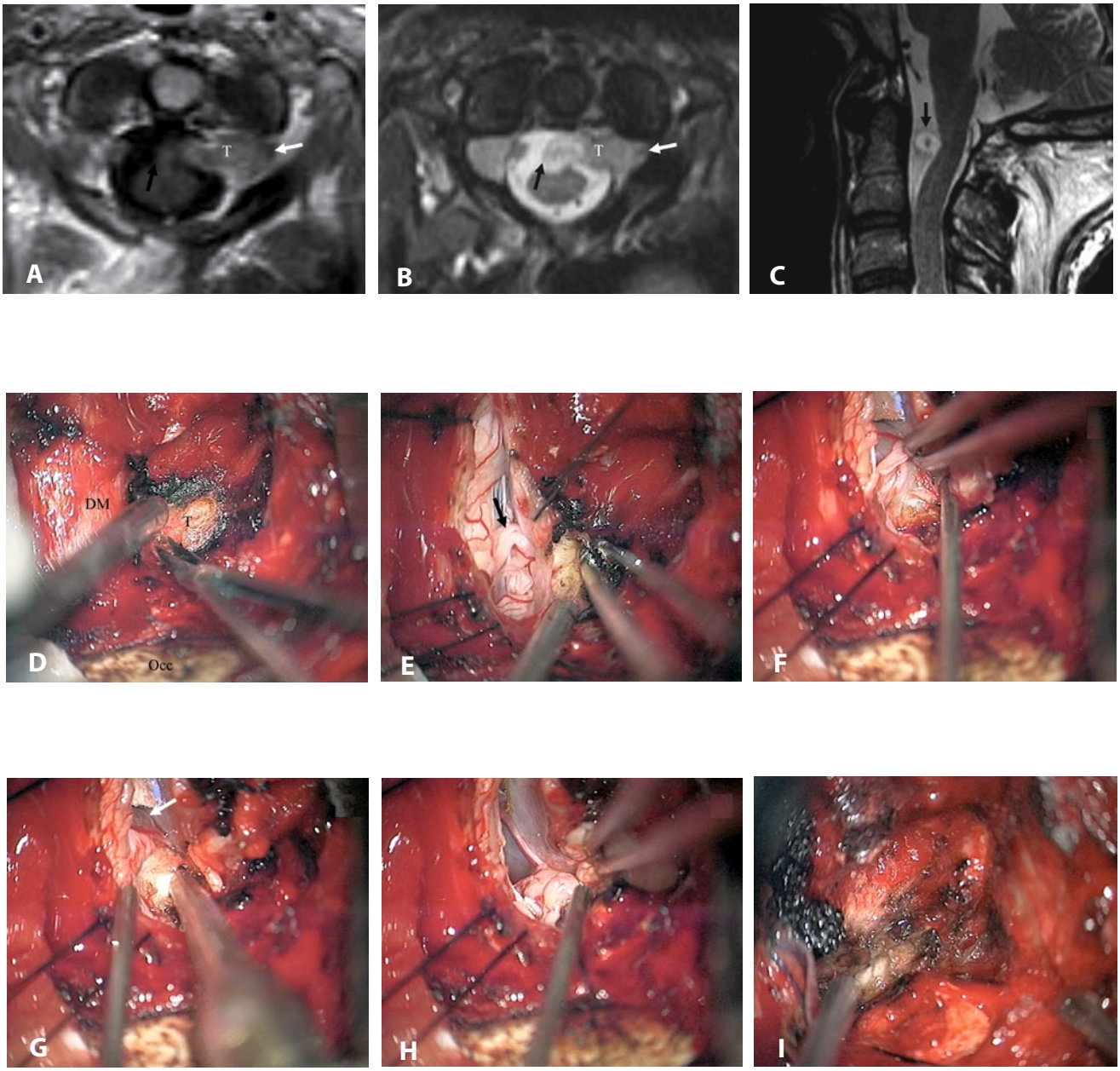

Fig. 16 – C1–C2 intra-extradural neurofibroma. This patient is known for a neurofibromatosis type I. He suffered from a progressive myelopathy due to a C1–C2 neurofibroma. Symptoms resolved after complete tumor removal through a posterolateral approach. **A–C**. Preoperative workup. **A**. Axial T1-weighted MRI with gadolinium. The tumor extends from the extradural compartment *(white arrow)* to the intradural one in front of the spinal cord *(black arrow)*.
B. Axial T2-weighted MRI. **C**. Sagittal T2-weighted MRI. The spinal cord is displaced backwards by the tumor *(arrow)*.
D–I. Peroperative views. **D**. The C2 nerve sheath is dissected extradurally. **E**. The dura mater has been incised longitudinally with contraincision to reach the level of the nerve root exit. A gentle traction is applied on the extradural tumor remnant. Arrow indicates a C2 posterior rootlet. **F**. Pathological rootlet are coagulated and will be divided. **G**. The tumor is progressively debulked using a ultrasonic aspirator. Arrow indicates the anterior extension of the tumor, in front of the spinal cord. **H**. The lesion can then be progressively tracted laterally. **I**. Final view of the C1–C2 interspace at the completion of the tumor resection.
DM: dura mater. Occ: occipital bone. T: tumor.

Angio-CT or angio-MRI provides pivotal information about the location of the VA V3 segment when tumors grow in its vicinity. Benign tumors can just displace the artery opposite to their growing center but aggressive tumor may completely encircle the artery. This last situation increases the surgical complexity and questioned to the need for a VA occlusion test and VA sacrifice.

Embolization

Embolization has to be considered preoperatively for highly vascularized tumors such as plamocytomas, thyroid, and renal cancer metastases. In other cases, even in vascular tumors such as aneurysmal bone cysts, preoperative embolization was not undertaken since the vascular feeders can be interrupted early in the surgical exposure.

Choice of the technique: biopsy

Biopsy is indicated in patients when two conditions are filled out: the diagnostic is unclear after preoperative radiological workup and the lesion did not necessarily require a surgical resection for decompression. Five of our patients were biopsied. Histopathological results were inflammatory processes, especially due to tuberculosis in three.

The main problem with CT-guided biopsy is misdiagnosis since samples are small. In our series, two diagnoses were reconsidered after complete tumor removal.

Choice of the approach

The surgical approach has to be decided according to the location of the lesion, with the objective of minimizing bone resection to preserve stability. The posterolateral approach, described earlier, is advocated for bone tumors developed in the posterior elements of the atlas and the axis. The anterolateral approach is advocated for lesions located in the occipital condyle, the anterolateral aspect of the C1 lateral mass, the anterior arch of the atlas, the vertebral body, and the dens of the axis. Using this approach, the VA must be controlled to allow confident work and improve surgical accessibility.

The anterolateral approach

This approach has been fully described in Chapter 25. Some points that require special consideration are listed here.

Head positioning

For reaching lesions located anteriorly to the VA, the head does not have to be rotated to the opposite side of more than 30°; otherwise, anterior structures will be projected too anteriorly. The relation between the VA V3 segment above and below the C1 posterior arch has to be anticipated accordingly.

Vasculonervous control

Bone tumors at the suboccipital level can develop in close relation with the vasculonervous structures passing anteriorly when gaining the cranial base. An adequate control of these structures in the neck must be obtained. By this way, pursuing the cephalad dissection of the ICA and of the lower cranial nerves may anticipate their positions.

VA transposition

VA transposition is a prerequisite to anterior tumor targeting. It implies opening of the C1 transverse foramen and unroofing of the VA. In some conditions, the VA can be totally embedded in the tumor (see Chapter 25, fig. 16). This condition represents a major surgical difficulty since the tumor prevents VA transposition. The VA must then be dissected away from the tumor at the beginning of the procedure. In this situation, proximal and distal controls must be obtained for two reasons. First, in case of VA injury during the dissection, the artery can be clamped on its both extremities therefore allowing for stopping profuse bleedings. Second, dissection of the VA from the tumor is less difficult by following the artery from normal areas found on both sides.

Bone resection

The basic principle is to resect as less normal bone as possible to reach the tumor to preserve a preoperative stability whenever it is still present.

When the VA is completely liberated and transposed, the FM lateral wall is brought into view: the occipital condyle, the anterior, and posterior arches of the atlas, the lateral mass of the atlas, and the C1–C2 joint come within reach. The exposure must now be adapted to each case, depending on the location and extension of the tumor. The odontoid process can be reached by passing over or through the C1–C2 joint.

Image-guidance

In our series, tumors were only targeted intraoperatively using anatomic landmarks based on a careful analysis of preoperative CT, with coronal, sagittal, and 3D reconstructions, without any help provided by intraoperative guidance systems such as navigation or fluoroscopy. Intraoperative guidance cannot be based on preoperative exams because of modifications of the position of the different anatomical structures according to the head rotation, except at the C0 level when data acquisition is based on head scanning. Image-guided surgery at the C1 or C2 level must be recalibrated on intraoperative images.

Postoperative complications

The anterolateral approach could induce a transient dysfunction of the accessory nerve resulting from the

nerve manipulation, responsible for pain along the trapezius muscle and weakness of the trapezius muscle and/or the sternocleidomastoid muscle. During this approach, manipulation of the sympathetic chain could also be at the origin of a transient Horner's syndrome.

VA damage has never been observed. In case of tumoral encasement, a balloon occlusion test must be realized.

Preservation of the lower CNs can be hazardous. If CN IX and X are damaged, postoperative swallowing problems must be anticipated.

Tumor particularities

Osteoid osteomas and osteoblastomas (Fig. 17)

Osteoid osteomas (OO) and osteoblastomas (OB) are pathologically similar bone producing lesions of osteoblastic origin, distinguished by their size (75–78). A lesion is diagnosed as OO below 15 mm, while above this size an OB is evoked (79–81). OBs own a more aggressive comportment than OOs as demonstrated by several features. First, the tumor extends to extraskeletal soft tissues and therefore explaining the incidence as up to 50% of preoperative neurologic deficits and the increased surgical difficulty for obtaining a complete resection (75, 82, 83). Second, their aggressive behavior explains also the increased recurrence rate after surgery, sarcomatous changes, and even metastases (77, 83–87).

Clinical symptoms of OOs are often characteristics: the pain presents a nocturnal pattern and is specifically relieved by acetyl salicylic acid. On the other hand, the pain produced by OBs is less severe at night than during the day and does not have a propensity to be relieved by acetylsalicylic acid (75).

When these lesions develop in the spine, a majority of them occur in the lamina, pedicles, or in the transverse and spinous processes. Vertebral body involvement is uncommon, very limited, and secondary to anterior extension of the tumor (75, 80, 82, 83, 88–90). Their location at the CCJ level is exceptional. Most publi-

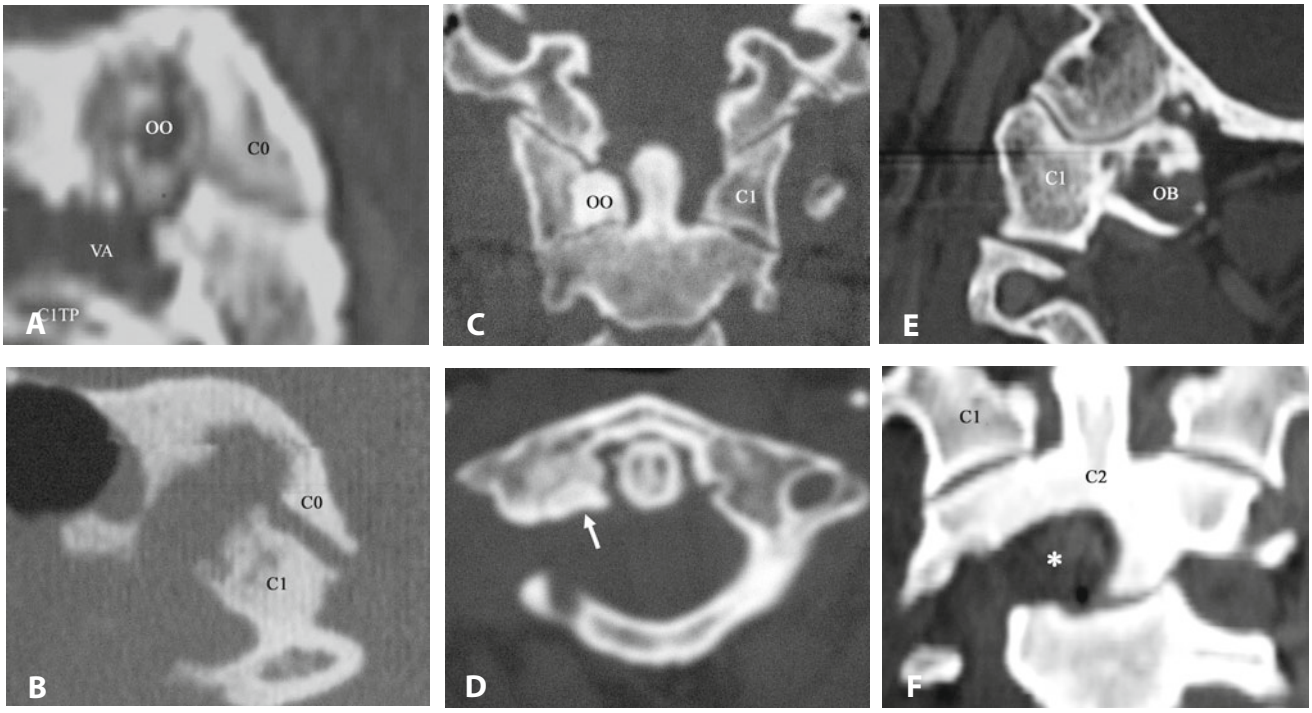

Fig. 17 – Osteoid osteomas and osteoblastomas at the C0–C2 level. **A**. Coronal bone CT reconstruction. The osteoid osteoma is developed just above the VA that has to be transposed for reaching it. **B**. Postoperative control. **C**. Coronal bone CT reconstruction. An osteoid osteoma is present at the medial aspect of the C1 lateral mass. **D**. Postoperative axial CT scan. A small remnant is seen *(arrow)* but was stable over time, while patient remains symptom free. **E**. Osteoblastoma of the C1 lateral mass, at its posterior aspect. **F**. Postoperative coronal CT reconstruction after complete resection of an osteoid osteoma *(star)* developed at the base of the C2 body. Limited access allows to preserve the stability without any need for fusion. C0: occipital condyle. C1TP: transverse process of atlas. OB: osteoblastoma. OO: osteoid osteoma. VA: vertebral artery.
Figures 17A, 17B, 17C, 17D, 17F modified from Bruneau M, Cornelius JF, George B (2005) Osteoid osteomas and osteoblastomas of the occipitocervical junction. Spine 30(19) : E567–71. Used with permission from Lippincott Williams & Wilkins.
Figure 17 E modified from Bruneau M, Polivka M, Cornelius JF, George B (2005) Progression of an osteoid osteoma to an ostteoblastoma. J Neurosurg Spine 3: 238–41. Used with permission from American Association of Neurosurgeons.

cations are case reports (76–78, 91–93), with one few series (78, 94). All tumors do not have to be removed surgically but only after failure of conservative treatments, if a lesion is suspected to be an aggressive OB, or if neural structures are stressed. Most of the time, surgery is indicated in OOs due to the persistent pain despite conservative treatments. OBs are removed due to pain, increasing size, and bone destruction (75).

Alternative techniques such as radiofrequency coagulation or percutaneous biopsy are efficient to treat OOs and control the pain but have never been applied at the CCJ level, probably due to the VA vicinity, which could be injured by a needle, even under CT control.

During surgery, a nidus was found in all cases of OOs. It appears as a small round ball within a cavity, possibly adherent to a part of its wall. In two cases, an unexplained inflammation surrounded the VA. This inflammatory reaction was responsible for adherences that render the dissection more difficult. Because one of the goals of the surgery is to maintain the preoperative stability, bone removal has to be very limited. In some cases, the bone resection can be restricted to the nidus, leaving in place partially the surrounding sclerotic bone. Conversely, OBs must be largely resected in relation to the tumor aggressiveness. In our series of OOs and OBs of the OCJ, arthrodesis and/or osteosynthesis was never required. In all patients, drilling was limited to less than one-third of the joints. As previously demonstrated, removal of less than half of the C0–C1 or C1–C2 joint is not associated with a postoperative instability (44).

The recurrence or remnant rate of OOs after surgical treatment reported in the literature is 4.5% and approximately 9.8–15% for OBs (75). Ozaki et al. (95) described two patients with an OO, who relapsed after an incomplete resection. In our series, one patient had to be reoperated: the OO was initially developed within the occipital condyle; the nidus was completely resected but the peripheral condensation was partially left in place. A symptomatic lesion recurred, suspected before the second surgery to be close to the previous surgical field. At surgery, another tiny osteoid osteoma was found and completely removed after complete drilling of the peripheral bony condensation.

Chordomas (Figs. 18–25)

Chordomas are midline locally aggressive tumors arising from notochordal remnants (74, 96). They account for 2–4% of primary malignant tumors and represent the most common primary malignant tumor of the mobile spine (96, 97). Three pathological subtypes of chordomas are: classic, chondroid, and dedifferentiated chordomas (98). Differential diagnosis from chondrosarcomas may at times be difficult but is important since the tumor behavior may be different (99). In this aggressive tumor, the role of the surgical resection is of paramount importance and the resection must be as radical as possible at first presentation while preserving quality of live (100–102).

Our series of CCJ chordomas encompasses 53 cases.

Tumor location

In all cases, the main part of the tumor involved the C2 body, odontoid process, or the C1 anterior arch. All these chordomas extended laterally toward the VA.

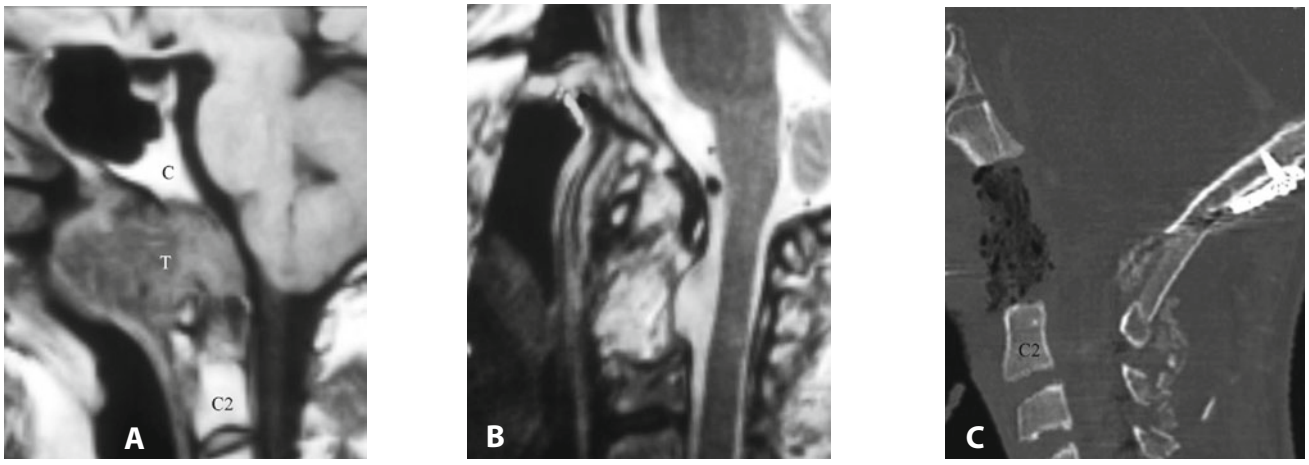

Fig. 18 – Chordoma. **A**. Preoperative sagittal T1-weighted MRI. **B**. Postoperative sagittal T2-weighted MRI after complete resection of the tumor through an anterolateral approach. **C**. CT scan sagittal reconstruction showing the bone resection and the posterior craniocervical bone grafting. C: clivus. C2: axis. T: tumor.

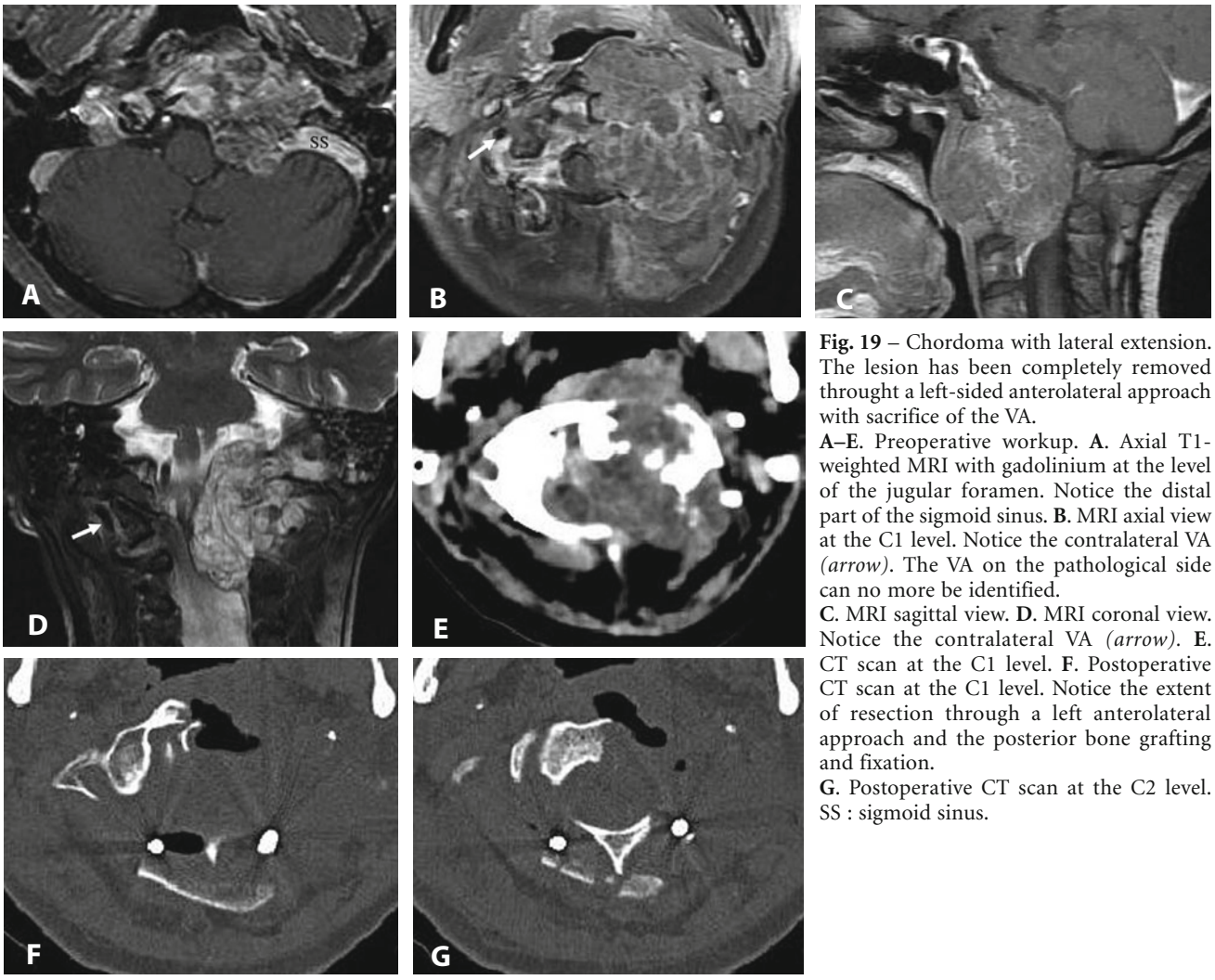

Fig. 19 – Chordoma with lateral extension. The lesion has been completely removed throught a left-sided anterolateral approach with sacrifice of the VA.
A–E. Preoperative workup. **A**. Axial T1-weighted MRI with gadolinium at the level of the jugular foramen. Notice the distal part of the sigmoid sinus. **B**. MRI axial view at the C1 level. Notice the contralateral VA *(arrow)*. The VA on the pathological side can no more be identified.
C. MRI sagittal view. **D**. MRI coronal view. Notice the contralateral VA *(arrow)*. **E**. CT scan at the C1 level. **F**. Postoperative CT scan at the C1 level. Notice the extent of resection through a left anterolateral approach and the posterior bone grafting and fixation.
G. Postoperative CT scan at the C2 level. SS : sigmoid sinus.

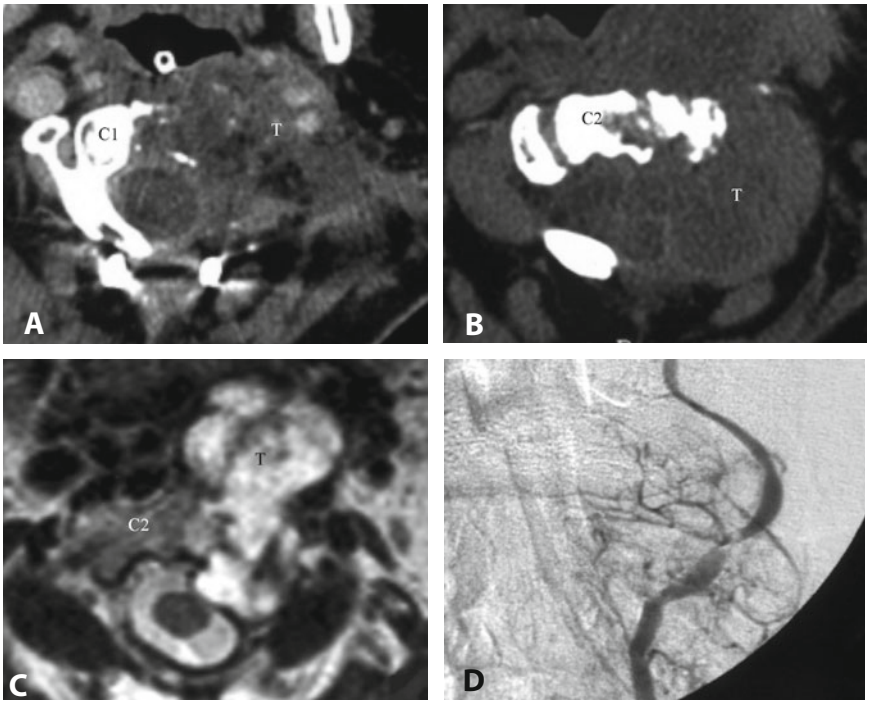

Fig. 20 – Chordoma with lateral extension. **A–B**. Axial CT scan. C. Axial T2-weighted MRI. D. Angiography showing severe tumoral induced stenosis suggesting also wall infiltration. C1: atlas. C2: axis. T: tumor.

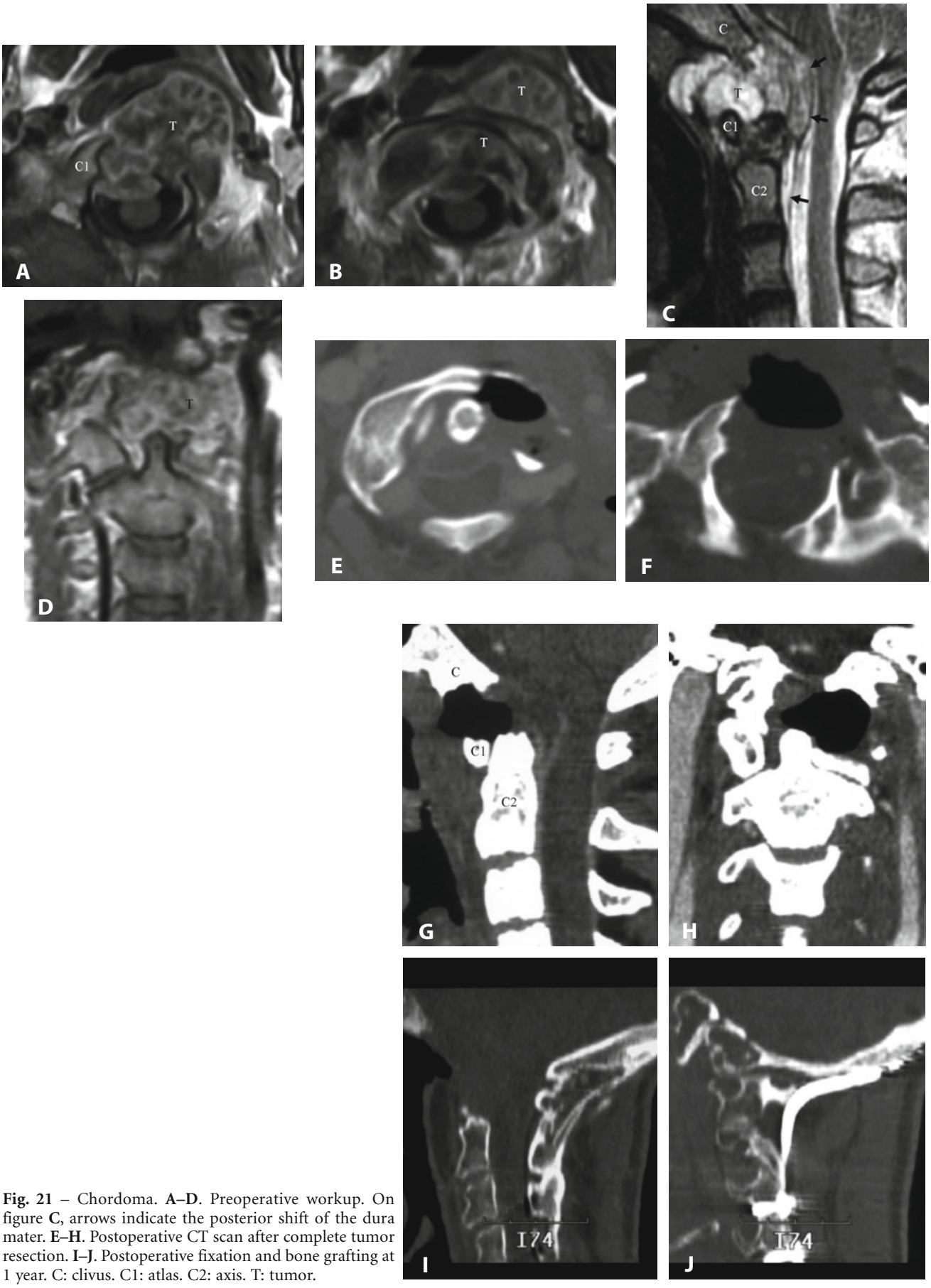

Fig. 21 – Chordoma. **A–D**. Preoperative workup. On figure **C**, arrows indicate the posterior shift of the dura mater. **E–H**. Postoperative CT scan after complete tumor resection. **I–J**. Postoperative fixation and bone grafting at 1 year. C: clivus. C1: atlas. C2: axis. T: tumor.

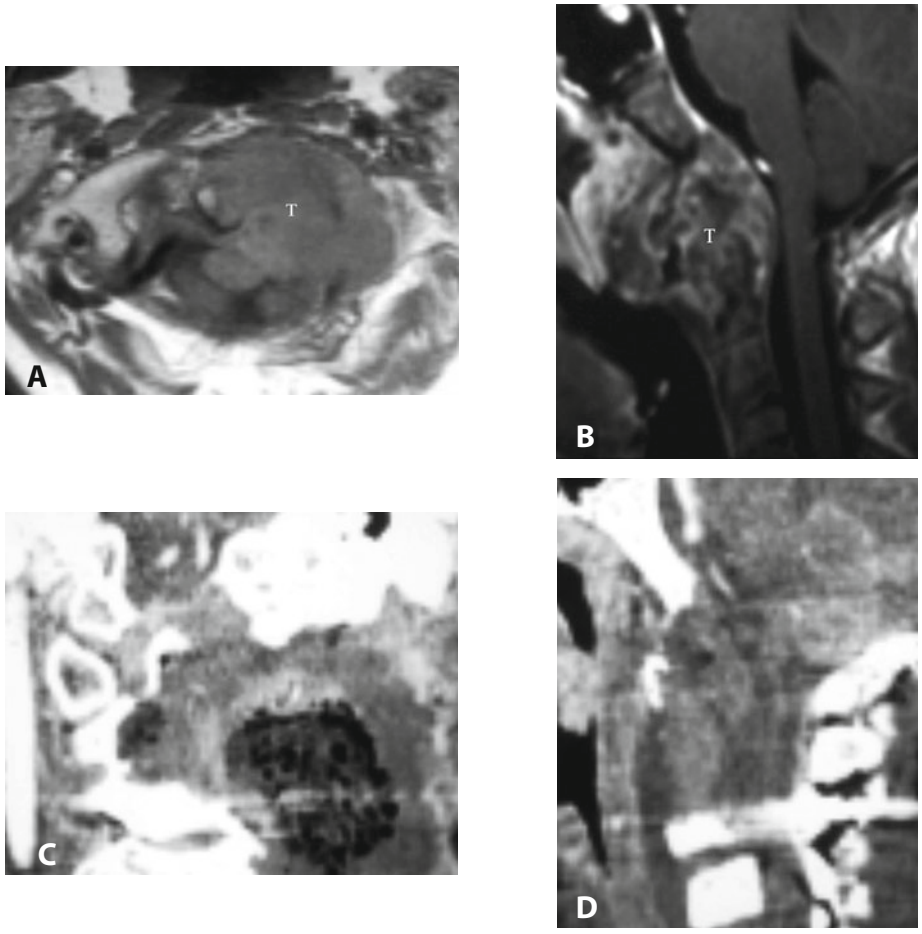

Fig. 22 – Chordoma. **A–B**. preoperative axial and sagittal MRI. **C–D**. Postoperative CT scan. Notice in **B** the extent of bone resection on the odontoid and up to the contralateral C1–C2 joint. T: tumor.

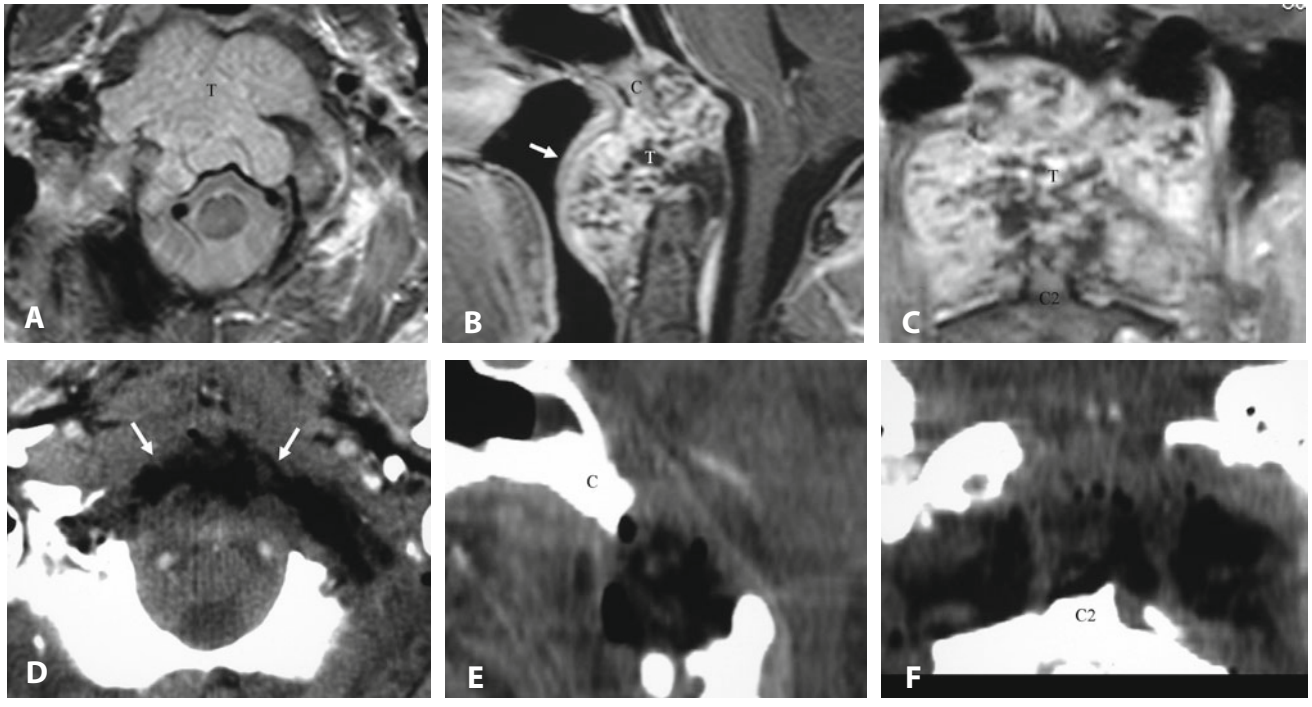

Fig. 23 – Chordoma with bilateral extension. **A–C**. Preoperative workup. **D–F**. Postoperative control corresponding to views **A–C**, after complete tumor resection through a bilateral anterolateral approach. *Arrow:* posterior pharyngeal wall. C: clivus. C2: axis. T: tumor.

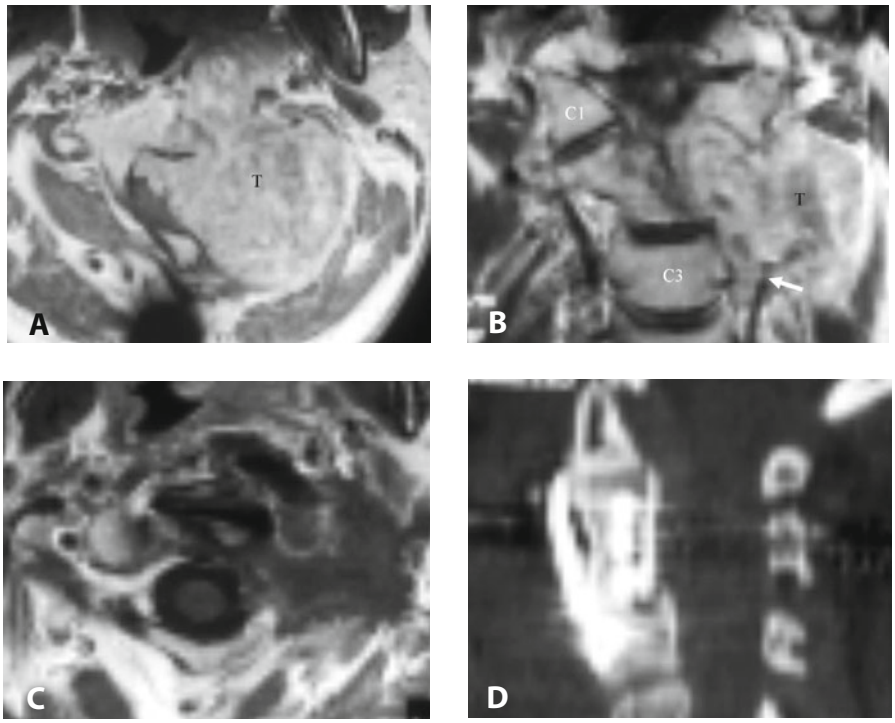

Fig. 24 – Recurrent chordoma with lateral extension. **A–B**. preoperative workup. In **B**, notice the VA encasement *(arrow)*. **C**. Postoperative axial MRI after complete tumor resection through a left-sided anterolateral approach. **D**. Sagittal CT reconstruction after anterior grafting and plating. C1: atlas. T: tumor.

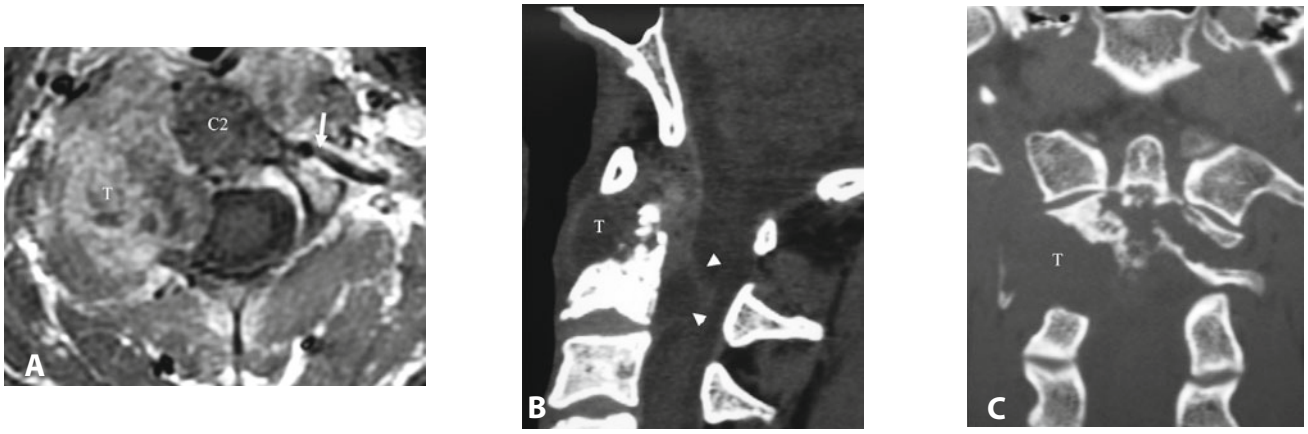

Fig. 25 – Chordoma with lateral extension. **A**. Axial T1-weighted MRI with gadolinium. Arrow indicates the controlateral VA. The ipsilateral VA is completely embedded within the tumor. **B**. Preoperative sagittal CT reconstruction. Arrowheads indicate a tumoral extension behind C2. **C**. Preoperative coronal CT reconstruction demonstrating C2 destruction. C2: axis. T: tumor.

Lateral extension of the tumor either reaches the lateral aspect of the VA, displacing it (23%), or usually completely encasing it (77%). This propensity for completely encasing the VA renders the complete surgical resection highly challenging at the CCJ level. This is especially true because the VA has to be freed early in the procedure to go further with VA transposition in order to access the anterior component of the tumor, which is otherwise out of reach. VA stenosis was observed in 20% of the patients, but no VA occlusion was detected.

Of note, 53% of the patients presented at first presentation but 47% were already operated on elsewhere previously. This point is very important because best results are achieved in patients with primary rather than with recurrent tumor (101, 102). This point justifies initial aggressive attempt of total tumor removal (101, 102).

A VA balloon-occlusion test was scheduled in some circumstances. Indications are summarized in Tables 2 and 3. Great care must be taken when facing a dominant VA especially in case of severe compression, encasement, prior surgery, and prior irradiation. In all our cases, the test was well tolerated and no revascularizing bypass procedure had to be anticipated.

Clinical symptoms at diagnosis consisted in neck pain and stiffness but long-tract palsy (23%) and lower cranial nerves dysfunction (30%) were also frequently observed.

Approaches

All patients were at least operated on through an anterolateral approach. However, most of them required a combined procedure with a contralateral anterolateral approach in the same or in a separate stage or with a posterolateral or a transoral approach. In average, one patient had 1.5 surgical approaches (from 1 to 4) for the tumor removal, excluding the approach used for the craniospinal fixation. Only four patients underwent only one surgical approach.

Tumor resection

Most of the time, chordomas are soft, gelatinous, slightly vascularized tumor, and can be rather easily resected with simple suction or ultrasonic aspiration. The problem at the CCJ is that tumors are generally intricated with tendons, muscles, and ligaments, so that not a single but multiple cavities are found. Very often, the tumoral limits seem to have been reached when a nice muscular or bony plane is obtained; in fact cutting the muscle or drilling the bone leads to discover more tumors. Extensive opening must therefore be realized and some landmarks (dura mater, odontoid tip, clivus, or contralateral joint) be defined preoperatively and reached intraoperatively.

The VA can often be freed from tumor but its separation generally needs a careful resection of the periosteal sheath surrounding the artery. This sheath is a very resistant barrier to the tumor extension. It is exceptional that at first presentation the actual wall of the VA is invaded asking for the sacrifice of the vessel (2 cases). This is more common in case of recurrence (6 cases). Whatever the condition (primary or recurrent) the VA has to be exposed and controlled inside its periosteal sheath proximal to the tumor; then the tumor is resected in a cranial direction along the VA. It is generally not possible to get a distal control, as this would need a dural opening, which is very rarely required by the tumor extension.

The resection must be as radical as possible but with preservation whenever possible of the lower cranial nerves. On the contrary, C1 and C2 nerve roots are generally sacrificed as this induces a limited sensory deficit. At the level of the CCJ, complete resection could be obtained in our series in 95% of the cases at first presentation. However, in some cases the tumors extend beyond the CCJ limits where the tumor cannot be followed in every part. Tumors may extend to the petrous bone with problem of internal carotid artery, lower cranial nerves, or even facial nerve preservation.

Craniospinal fixation

Osteosynthesis is often necessary to stabilize the CCJ. This is decided preoperatively from the tumor development. Two-thirds of our patients underwent a fixation procedure with plating and bone grafting but half of them were referred to us with a fixation previously done.

Radiotherapy

As already reported (29), complementary radiation therapy helps to delay the recurrence and is proposed to every patient after radical resection at first presentation. Recurrent cases have often already been treated by radiotherapy. Since high dose of radiation is only efficient, proton beam is the most appropriate form of radiation therapy. It permits to irradiate the tumoral region with much safety for the adjacent sensitive structures (100, 103). Gamma-knife is less adequate at first presentation as it can treat only a small volume. It may be useful in recurrent case with small focal remnants (104).

Outcome

In the group of patients treated at first presentation, recurrence was observed after a mean delay of 34 months. Actuarial survival rate was 80% and 65%, respectively,

at 5 and 10 years in the patients treated at first presentation. Conversely it was 50% and 0%, respectively, at 5 and 10 years in patients treated after recurrence.

Other tumors (Figs. 26–33)

Many different types of tumors can be observed at the CCJ level. Most often they are removed through an anterolateral approach. Surgical strategy is first to define where the proximal VA has to be primarily exposed before going to the tumor. This permits then a safe exposure of the tumor but also to divide most of the vascular supply in vascularized tumors (plasmocytoma, aneurysmal cyst, metastasis). Distal control of the VA is often difficult to obtain. Second is to estimate how much of the CCJ stability is compromised and whether or not a fixation procedure is necessary. Accordingly, the surgical resection is achieved with as less bone drilling as possible. In our experience, the surgical resection has never led to an instability not previously existing.

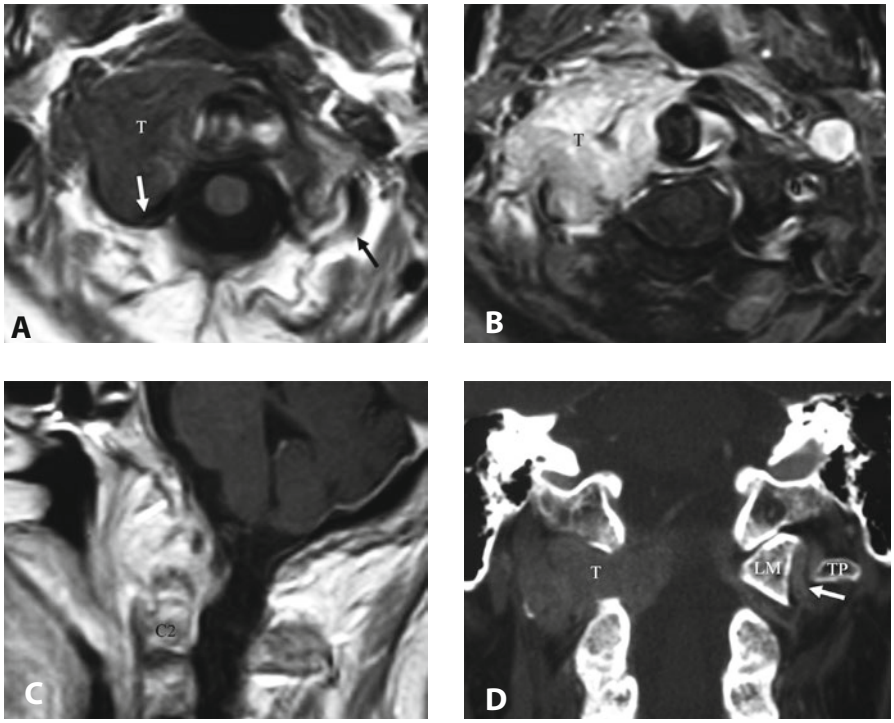

Fig. 26 – Thyroid metastasis. **A**. Axial T1-weighted MRI. White and black arrows show the right and left VAs. The right VA is involved within the lesion. **B**. Axial T1-weighted MRI with gadolinium showing clearly the tumor extension. **C**. Sagittal MRI with gadolinium. **D**. Coronal bone window CT reconstruction after injection. The left VA is indicated with the arrow. On the pathological side, the VA is no more distinguished since completely encased by the lesion. C2: axis. T: tumor. LM: C1 lateral mass. TP: C1 transverse process.

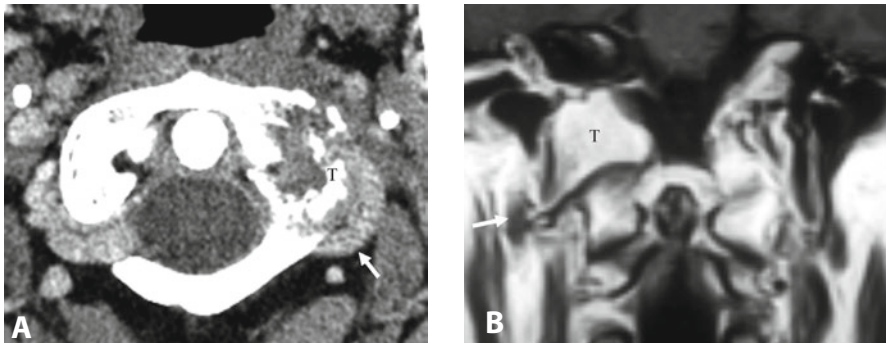

Fig. 27 – Two examples of lung metastases.
Lesions involved the VA V3 segment *(arrow)*. T: lung metastasis.

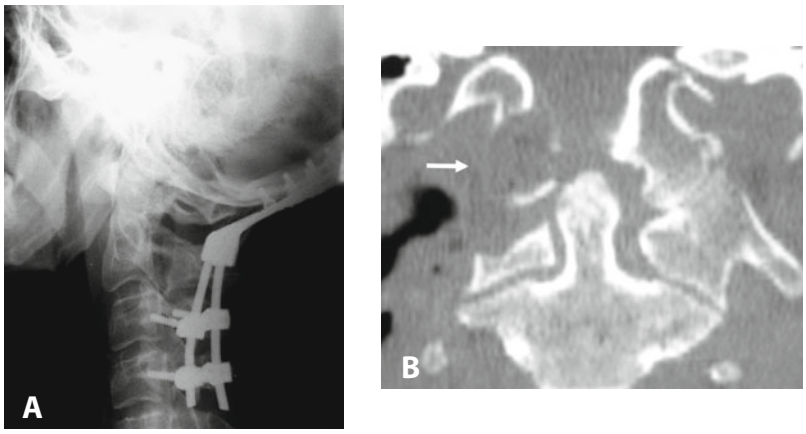

Fig. 28 – Metastasis involving C1–C2. **A**. Posterior craniocervical fixation. **B**. Postoperative CT scan, coronal reconstruction, demonstrating the complete tumoral.

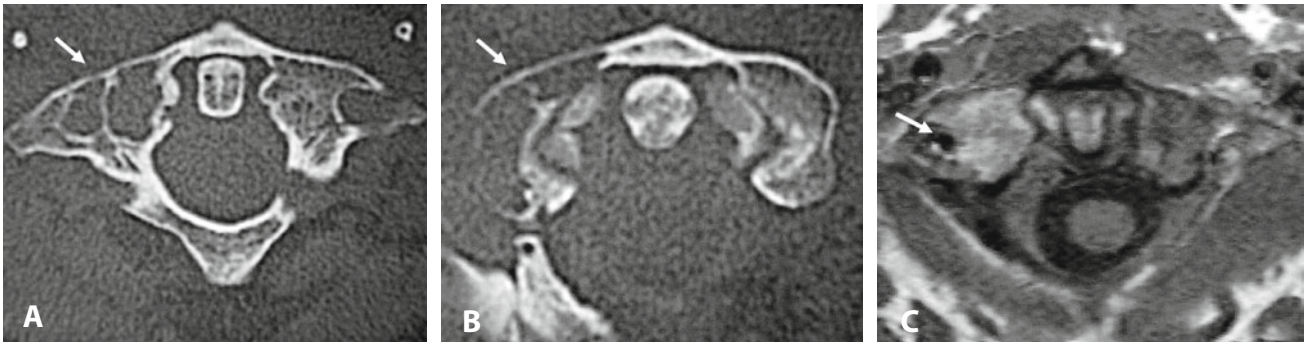

Fig. 29 – Histiocytosis involving the lateral mass of atlas and occipital condyle. **A–B**. Preoperative bone window CT scan depicting bone destruction (*arrow*). **C.** T1-weighted axial MRI shows the VA involvement *(arrow)*.

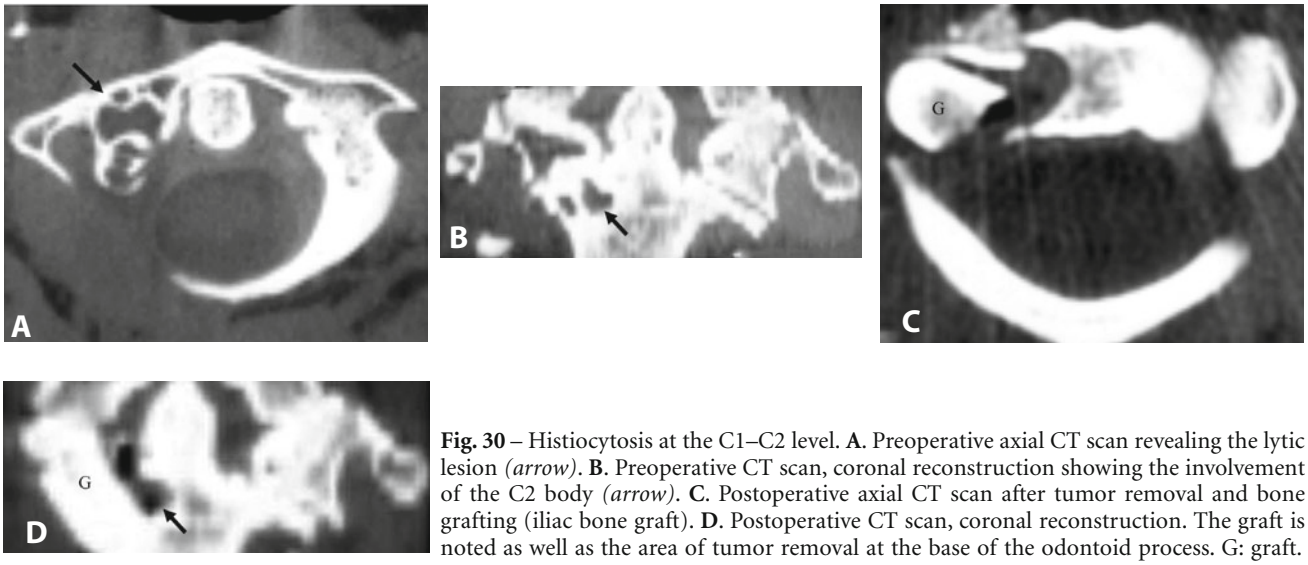

Fig. 30 – Histiocytosis at the C1–C2 level. **A**. Preoperative axial CT scan revealing the lytic lesion *(arrow)*. **B**. Preoperative CT scan, coronal reconstruction showing the involvement of the C2 body *(arrow)*. **C.** Postoperative axial CT scan after tumor removal and bone grafting (iliac bone graft). **D**. Postoperative CT scan, coronal reconstruction. The graft is noted as well as the area of tumor removal at the base of the odontoid process. G: graft.

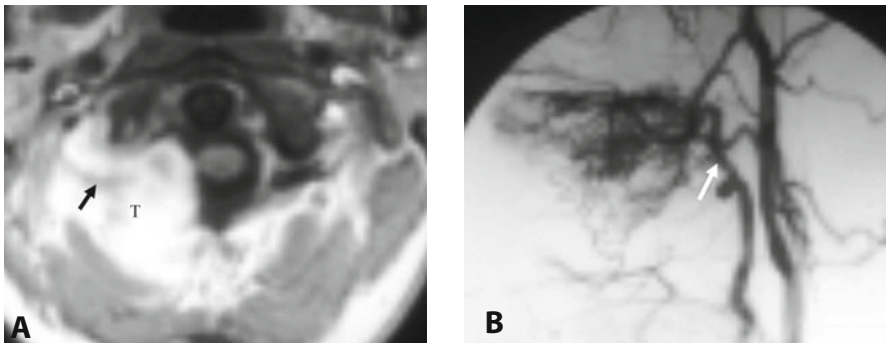

Fig. 31 – Histiofibroma.
A. Axial T1-weighted MRI with gadolinium. The lesion involves the right C1 lateral mass and encases the VA *(arrow)*. **B**. Angiogram showing the tumor hypervascularization from the VA *(arrow)*. T: tumor.

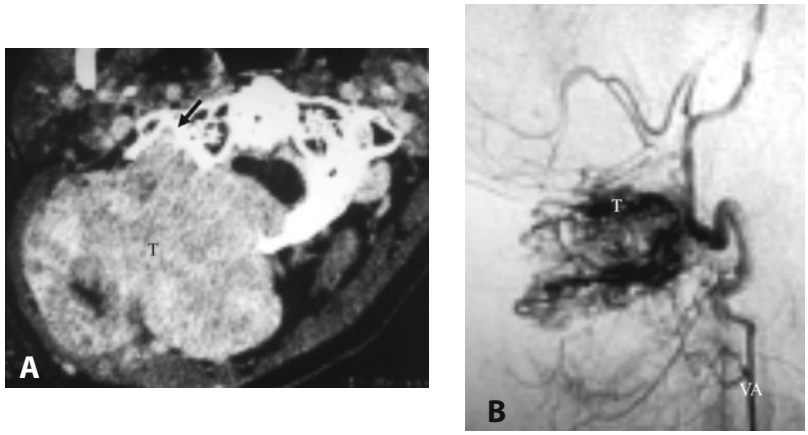

Fig. 32 – Angiomyolipoma.
A. Axial CT scan. The tumor extends into the C1 transverse foramen *(arrow)*. **B**. VA angiogram indicating the tumor hypervascularization. T: tumor. VA: vertebral artery.

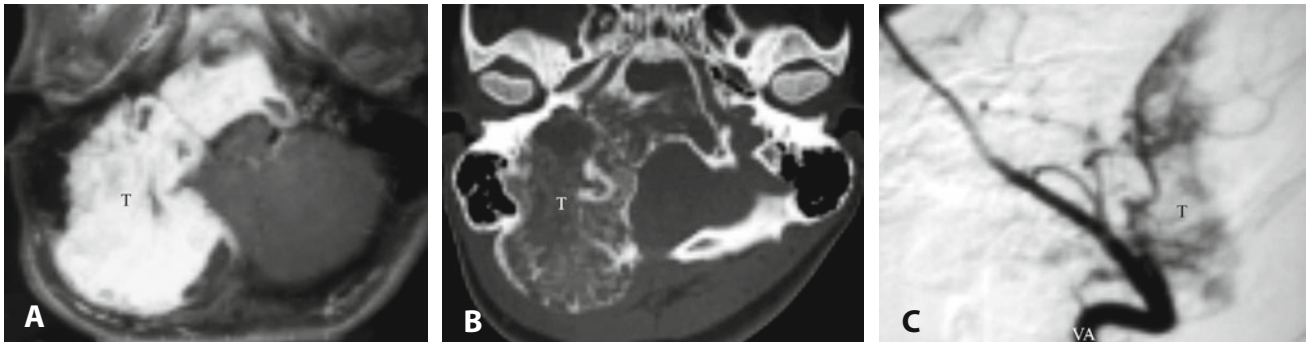

Fig. 33 – Fibrous dysplasia. **A**. Axial T1-weighted MRI. **B**. Axial bone CT scan. **C**. Angiography. T: tumor. VA: vertebral artery.

Pseudotumors (Figs. 34–35)

Pseudotumors are all included in group III that comprises purely extradural tumors. They raise the problem of differential diagnosis with actual tumors. In fact this may have important consequences, as most of these pseudotumors do not require as complete a resection as possible. In presence of infections especially tuberculosis, just a CT-guided biopsy has to be performed. In case of rheumatoid arthritis, synovial cyst, or bone malformations, a decompression is only needed.

Decompression is most frequently achieved using an anterolateral approach; however, some bony malformations are sometimes more easily reached through a posterolateral approach. This was the case of five posterolateral bony malformations, including one bilateral but also one synovial cyst and two inflammatory processes invaginating into the dura. Decompression has the advantage of preserving the stability. In fact it is in this group of pseudotumors that was observed the only case (rheumatoid arthritis compression) with a preoperative CCJ stability that had an extensive drilling of the C0–C1 joint leading to postoperative instability and requiring a stabilization procedure.

We include in this subgroup osteoarthritis of the atlantoaxial joints. With a prevalence between 5% and 18%, this entity is more common than generally suspected but only a minority of patients becomes symptomatic (105). Patients complain of severe suboccipital pain, irradiating into the occiput, vertex and sometimes as far as the eyes (105). The pain can be very debilitating. Conservative treatments with collar immobilization, anti-inflammatory drugs, and infiltrations can be helpful as first line therapy. If they failed, various surgical procedures have been reported in the literature including osteoarthrodesis with excellent long-term results after fusion (105–107).

The spondylothic formations resulting from degeneration of the C1–C2 joint can also stress the C2 nerve root. In our experience, we have encountered two patients suffering from osteoarthritis of the atlantoaxial joint and C2 radiculopathy. Our strategy was first to release radiculopathy by decompressing the C2 nerve root. Through a posterior midline approach, the vertical portion of the VA V3 segment is exposed on the pathological side. It is performed safely after a subperiosteal dissection of the C1 and C2 transverse processes. The C2 nerve root is exposed from medially to laterally up to its crossing on its anterior aspect with the VA. The conflicting zone between the hypertrophied joint and the nerve root can then be safely exposed and removed with pounches and a diamond drill. Subsequently, we perform a posterior osteoarthrodesis to decrease the stress on the pathological joint (Fig. 35). Postoperative outcome was excellent in all cases with immediate pain release, maintained over a long-term follow-up.

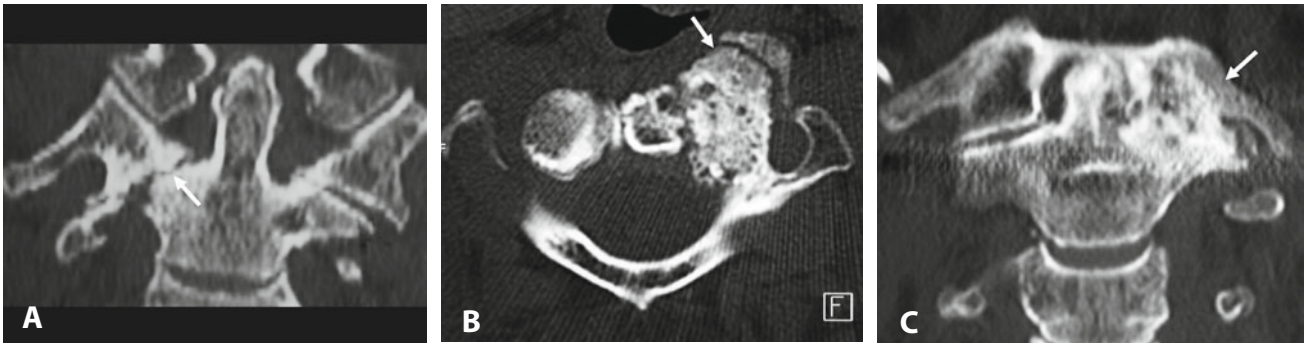

Fig. 34 – Examples of pseudotumor: spondylotic changes of the C1–C2 joint.

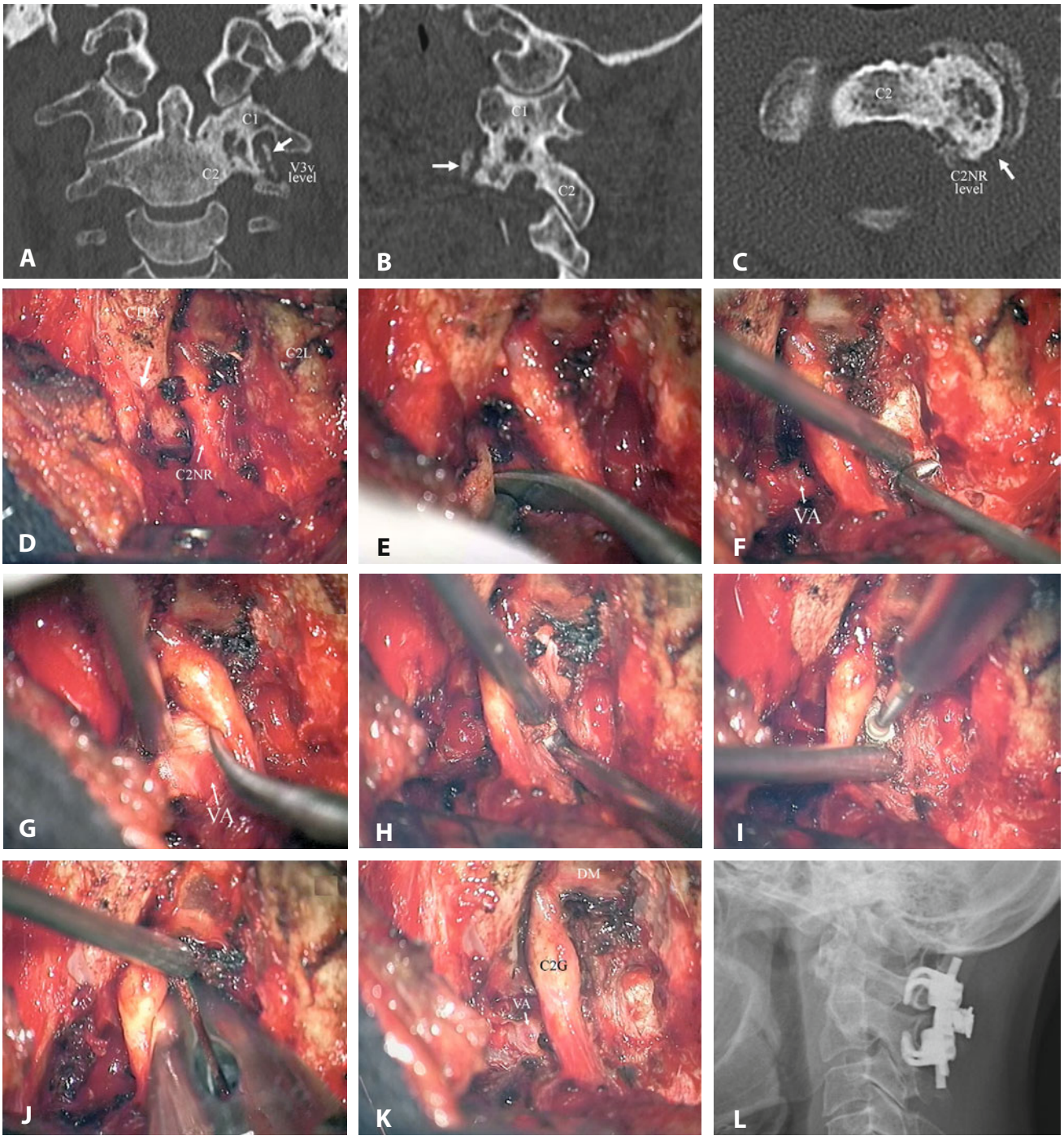

Fig. 35 – Pseudotumor at the C1–C2 level. The patient complained of severe irradiating cervical cephalalgia. Surgery consisted in decompression of the C2 nerve root and osteoarthrodesis through a posterior approach. Postoperatively the patient was pain-free. **A–C**. Preoperative bone window CT scans in coronal *(A)*, sagittal *(B)*, and axial *(C)* sections. Arrows indicate hypertrophic bone formation developed along the vertical portion of the VA V3 segment and the C2 nerve root. **D–K**. Operative views. **D.** The C2 nerve root is well observed. The horizontal portion of the VA V3 segment is located above the groove of the superior aspect of the C1 posterior arch *(arrow)*. **E**. Subperiosteal dissection at the inner aspect of the C1 transverse foramen. **F**. The vertical portion of the VA V3 segment has been exposed. The C2 transverse foramen is located using a blunt hook. The VA runs in front of the nerve root. **G**. The C2 nerve root is mobilized caudally to show the junction between the nerve root and the artery. The capsule joint and osteophyte are located between the succer and the dissector. **H**. The osteophyte is removed with a small curette along the VA. **I**. A diamond drill can also be used. Notice the role of the succer, allowing for retracting the nerve root and protecting the VA. **J**. Small bone remnant can be resected using a small gouge. **K**. Final view at the end of the decompression. **L**. Posteoperative lateral X-rays after C1-C2 osteosynthesis with sublaminar hooks. C1PA: posterior arch of atlas. C2G: C2 ganglion. C2L: lamina of C2. C2NR: C2 nerve root. DM: dura mater. V3v: vertical portion of the vertebral artery V3 segment.

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