27 Foramen Magnum Tumours: Posterior Approaches and Outcome

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Tumours at the foramen magnum are exceedingly rare. They are divided in *intradural tumours*, such as meningiomas, schwannomas, or neurofibromas, with foramen magnum meningiomas (FMMs) representing the majority among them. *Extradural tumours* include chordomas and chondrosarcomas, or metastases, with lower clival chordomas representing the most frequent entity of this group. Depending on size and extension a variety of surgical approaches to this region has been proposed, extending from endonasal endoscopic approaches to the anterior rim of the foramen magnum to far lateral trans-/paracondylar approaches with/without transient transposition of the vertebral artery (VA) to otherwise extended retrosigmoid approaches with additional opening of the foramen magnum—and to variations of the standard posterior midline approach [1–11].

FMMs originate from the dura of the foramen magnum proper, thereby accounting for less than 3% of intracranial meningiomas in large clinical series [12]. Epidemiologically they don't differ from meningiomas in other localizations, with a mean age of >50 years and a female predominance. They share with other intra- or extradural lesions of this particular region that they may reach a considerable size prior to diagnosis—despite their delicate localization close to critical neurovascular structures at the cranio-cervical junction. Most FMMs arise from the anterior or anterolateral aspect of the foramen magnum, that is anterior to the denticulate ligament. Likewise in FMMs and in other tumours, the relationship between the tumour and the V3 and V4 segments of the vertebral artery, with CN IX–XII, the posterior inferior cerebellar artery (PICA), and the brainstem are decisive for the surgical strategy. This anatomical complexity renders treatment of such tumours a challenge—despite all contemporary technology and techniques for diagnosis and for surgical treatment.

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27.1 Symptoms

Symptomatology includes spontaneous head and neck pain in the occipital and sub-occipital region, and painful limitation of head movements in flexion, extension and on rotation [2, 5, 7, 9–11]. Subtle neurological deficits such as mild to moderate lower cranial nerve palsies may go unnoticed. For example, especially elderly patients may present with long-standing dysphagia, hoarseness, or with deviation of their tongue, problems which they may have attributed to more generalized aspects of ageing. Depending on the site of dural attachment, precise localization, and size, other symptoms include unilateral or bilateral long-tract signs such as spasticity and gait ataxia [2, 5, 7, 9–11].

27.2 Diagnosis

Adequate contemporary diagnostic workup includes not only thorough clinical examination and standard imaging, but video oesophago- and tracheoscopy and X-rays or MRI of the swallowing act as well. MR imaging of the whole neuraxis is recommended to exclude multiplicity of lesions. That is, elderly patients in particular who are presenting with uni- or bilateral long-tract signs may have suffered from previous stroke, or they may suffer from additional cervical spondylotic myelopathy (CSM), the latter of which has to be taken into account when it comes to positioning for surgery. MRI should include not only native and injected standard sequences, but MR venography in addition, because, especially in large FMMs, there may be involvement of the sigmoid sinus. A persisting occipital venous sinus should be ruled out as well, as this may represent a major source for surgical complications during the early phases of craniotomy and dural opening. High-resolution CT of the cranio-cervical junction is equally recommended: this helps to assess the degree of calcification of the FMMs and its potentially hyperostotic root, to understand the bony relationship with the occipital condyles and the transverse processes of C1, with the jugular tubercle, and to prepare for the case that instability may ensue due to (partial) drilling of the C0-C1 joints. Should involvement or encasement of the vertebral artery (typically segments C3 and C4) be suspected from the MRI, then performance of CT angiography is recommended. This may influence the surgical strategy, i.e. by preparation for vascular repair.

27.3 Classification

There are several classifications for FMMs (Lit) [3, 9, 13]. A more recent one was proposed by Bernard George's group from Paris, which is based on three main criteria: the compartment which contains the majority of the tumour mass, the site of dural insertion, and the degree of vertebral artery involvement. The tumours can be localized strictly intradurally (anterior, lateral, or posterior), combined intra-extradurally, or extradurally [3]. The latter two types are localized antero- or posterolaterally. An additional criterion concerns the relationship between the tumour and the VA: the FMMs may be localized above or below, or combined above and below with subsequent encasement of the VA.

27.4 Presurgical Considerations

In this author's view, it doesn't make sense to be apodictic about the approach to these lesions: posteriorly located FMMs are approached by a posterior midline approach anyway. When it comes to the surgical resection of antero- or posterolaterally located FMMs, different neurosurgical schools apply different rules. This includes patient installation in semi-sitting, or in supine, or in park-bench positions, and the practice of far lateral, or trans- and paracondylar approaches with varying degrees of drilling of the C0-C1 joints and the jugular tubercle [1, 2, 12, 14–17]. The vast majority of FMMs may be accessed by a posterior midline approach, however [12, 15, 18, 19].

27.5 Technique of Dorsal Midline or Paramedian Approach

The dorsal midline approach to FMMs is performed in the following steps:

1. Surgeon should assure herself/himself that adequate haemostatic material is available on the scrub nurse's table (i.e. Gelfoam™, Tachosil™, Surgiflo™, aneurysm clips in case of involvement of the VA).
2. In case of large FMMs and presence of additional CSM: Placement of monitoring tools and devices PRIOR to definitive patient positioning. This includes standard MEP and SEP monitoring to/from arms and legs (median nerve and tibial nerves, and corresponding specific muscles, respectively).
3. Prone position, the head in flexion (Concorde).
4. Linear incision, normally from inion to C3 (depending on extent of caudal tumour growth). Palpating the inion and the spinous processes of C2 and lower mxfhelps to draw an appropriate line on the skin.
5. Placement of one curved self-retaining retractors with the handle pointing cranially at the level of the posterior fossa, which follows the anatomical shape of the suboccipital region as to avoid interference with the surgeon's hands and hindering convenient access to the target region. A second self-retaining retractor may be used in reverse sense, its handle pointing caudally, depending on the necessity and extent of cervical exposition and possible need for resection of posterior arches C1 and C2. Resection of extradural tumours such as chordomas may end at this stage—with resection and drilling of affected bony structures around the FM.

6. Haemostasis, placement of cotton patties, and rinsing of the surgical field.
7. In a midline approach, and depending on the cranial extension of the tumour, it is advised to place two burr holes at the same horizontal level, on either side of the midline, and as far lateral as possible. Posterior fossa exposition is notoriously limited and easily misjudged in the beginning, and each millimetre of exposition counts. These twin burr holes are connected by a blunt dissector. Care must be taken not to run into a possible large venous lacuna or a persisting occipital venous sinus. In that case, drilling of the bone to complete the craniotomy may be safer than the blunt use of a craniotome for the connection of these two burr holes. Adequate haemostatic material and dural titanium clips should be ready to be used if deemed necessary.
8. The posterior atlanto-occipital membrane can be thickened and extremely cumbersome to be separated from the occipital bone.
9. Dural incision is performed in a way which leaves a 2–3 mm rim around the entrance zone of the vertebral artery, so as to have enough room for watertight dural closure at the end of the procedure and to minimize the risk for occurrence of a CSF fistula.
10. Upon dural opening, and in case of direct visibility of an aspect of the tumour it may be advisable to perform direct electrical stimulation of the exposed capsule in order to avoid to miss and to cut through a thinned and stretched CN.
11. If possible, the capsule of the tumour is followed toward its site of insertion. If being confronted with whitish fibres, which cannot be clearly attributed to CN or to denticulate ligament, then direct electrical stimulation is performed again. At the anterolateral insertion site, bipolar coagulation is performed for consequent devascularization of the tumour.
12. In most intradural tumours with no history of previous surgical or radiosurgical treatment, it is possible to find and to define an arachnoidal plane between their capsule and critical neurovascular structures such as CN or the brainstem. It is of utmost importance to guard this plane as it will help during tumour dissection from its surrounding structures. Large tumours may be debulked from internally first, thus facilitating manipulation of residual tumour and its capsule. The situation can be different and far more challenging in recurrent tumours where it might be advisable to leave a thin layer of tumour on CN or the brainstem rather than trying to peel it completely off and thereby causing structural damage and consecutive neurological worsening.
13. In most instances, the zone of dural attachment is resected in this late stage of the surgical procedure. If this concerns dura overlying the clivus, this does not require subsequent duroplasty. If the insertion site was more lateral or posterior, then bovine pericard can be used for duroplasty. It is advised to cover the dura with fibrinoid material at the end of the procedure (i.e. with Tachosil™).

27.6 Technique of Dorsal Paramedian Approach

Alternatively, and for a strictly lateralized approach, the skin incision should be placed on a line halfway between the posterior midline and the virtual projection of the lateral border of the patient's head ("mid of the mid"). Subcutaneous and transmuscular dissection will equally lead to the posterior fossa along its flattening aspect toward the foramen magnum, and laterally to the C0-C1 articulation and the basion.

27.7 Outcome

The posterior midline approach has been used for the treatment of FMMs since decades. This includes meningiomas of all sizes, calcified and non-calcified tumours, and antero-laterally and anteriorly localized FMMs. The rationale behind the use of (far) lateral approaches is the avoidance of traction-related neurological worsening, i.e. by placing retractors on the brainstem with direct impact on long white matter tracts and cranial nerve nuclei, or by transmitted traction on the cranial nerves themselves. Proponents of lateral approaches argue that there is better direct tumour exposition and local control of neurovascular structures around it by additional bone and C0/C1-joint drilling. This should then result in better clinical outcomes in large series of FMMs reported in the literature. In fact, this is not the case when different series from different neurosurgical centres are compared (with all the reservations with regard to such comparisons). The results which were obtained with the use of posterior approaches in respective clinical series of FMMs are as follows: permanent morbidities in the range of 5–10%, and mortality <5% [4, 9–11, 13, 15, 20–22]. This is not different from corresponding numbers for (far) lateral approaches with/without additional osteosynthesis of the cranio-cervical junction. In principle, these numbers show that surgery of tumours at the foramen magnum by experienced teams is entirely feasible. There seems to be an intrinsic rate of morbidity and mortality for the resection of these lesions, which is the same for both principally different types of approaches—dorsal vs. lateral. This is an argument in favour of the posterior approach, because it takes lesser time and resources—presumed that the results on long term are the same. Reported recurrence rates for either approach were similarly low, however [15, 20, 21, 23]. In addition, the application of simple posterior approaches without drilling of the C0/C1 joints does not require additional cranio-cervical osteosynthesis with all its inherent risks and complications (Figs. 27.1, 27.2, 27.3, 27.4, 27.5, 27.6, 27.7, and 27.8).

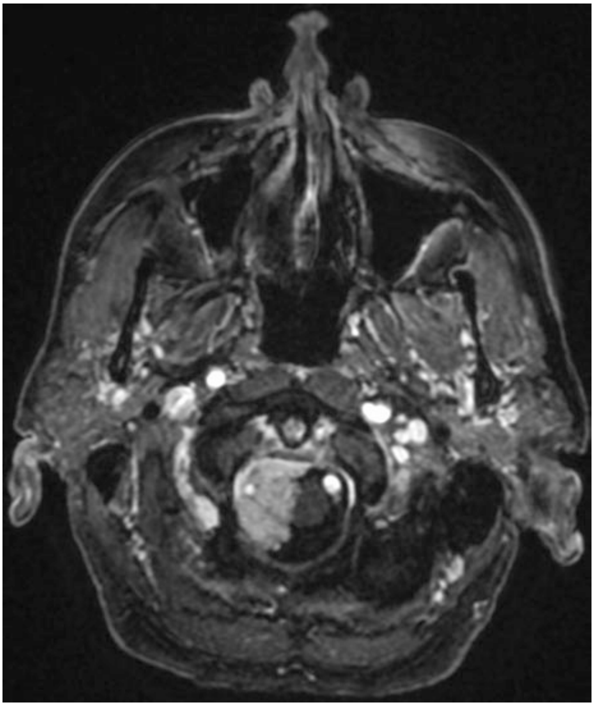

Fig. 27.1 Right-sided laterally localized FMM, with complete encasement of the (non-dominant) right vertebral artery in a 77-year-old female patient. This Gd-enhanced axial cut of a T1-weighted sequence allows to distinguish between the various involved neurovascular structures, the enhanced tumour, which takes approximatively up to 50% percent of the diameter at the level of the cranio-cervical junction. This tumour was approached via a standard midline incision, followed by a suboccipital craniotomy, resection of the posterior arch of C1, partial resection of the arch of C2—all more to the right side than to the left

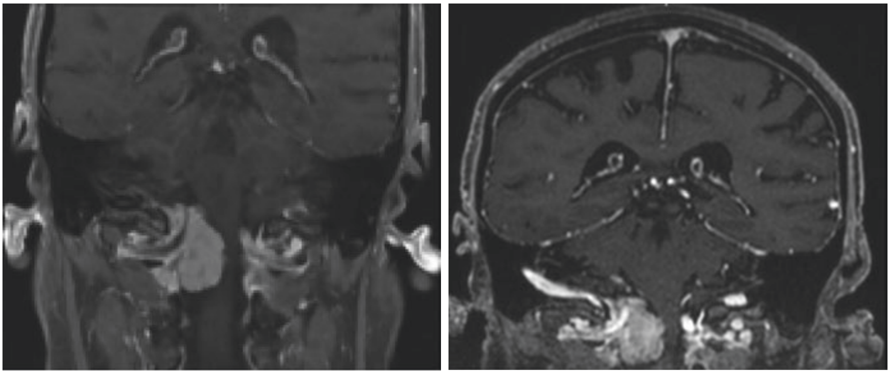

Fig. 27.2 Coronal view in same patient as in Fig. 27.1 of right-sided, laterally localized FMM, with complete encasement of the (non-dominant) right vertebral artery. T1-weighted Gd-enhanced imaging clearly shows the dural attachment and the venous sinuses. All this is relevant for planning of the surgical approach, and for preparation of material for vascular repair

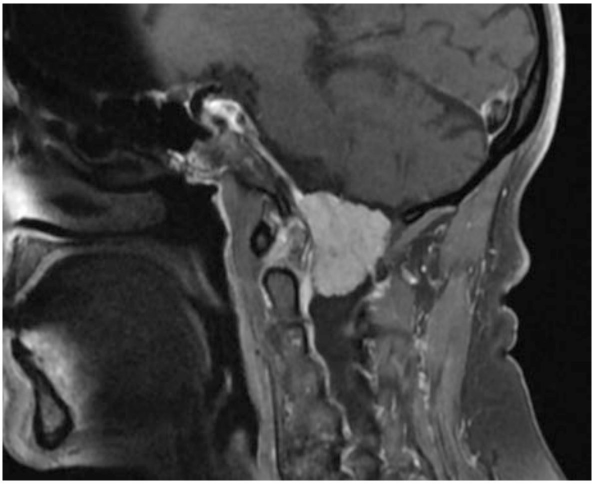

Fig. 27.3 Sagittal view in same patient as in Figs. 27.1 and 27.2 of right-sided, laterally localized FMM. This T1-weighted Gd-enhanced image is of particular value for assessment of the tumour size in the cranio-caudal direction and for presurgical planning, i.e. skin incision, resection of laminae C1 and C2, etc.

Fig. 27.4 Right-sided, anteriolaterally localized highly calcified meningioma in 67-year-old woman with known psychiatric disease and increasing walking difficulties. On this T1-weighted Gd-enhanced axial MR image, the tumours are mainly black. There is severe compression of the brainstem and partial encasement of the vertebral artery. The tumour was approached via a midline skin incision from the inion down to the spinous process of C2. Resection of the posterior arch of C1 and posterior enlargement of the foramen magnum by drilling the infero-posterior part of the occipital bones. This was followed by Y-shaped dural opening. Only subtotal removal was possible, because the tumour had to be drilled out partially, and a margin of security was left around the vertebral artery

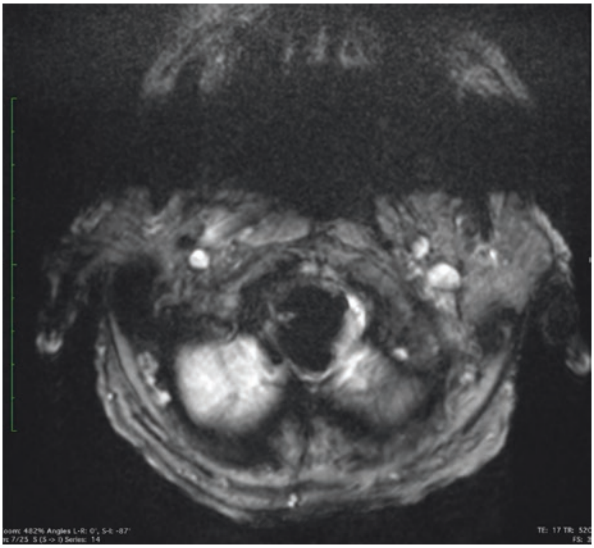

Fig. 27.5 Non-injected T2-weighted axial MR image of the same patient as in Fig. 27.4. The extent of the calcified process is better demarcated

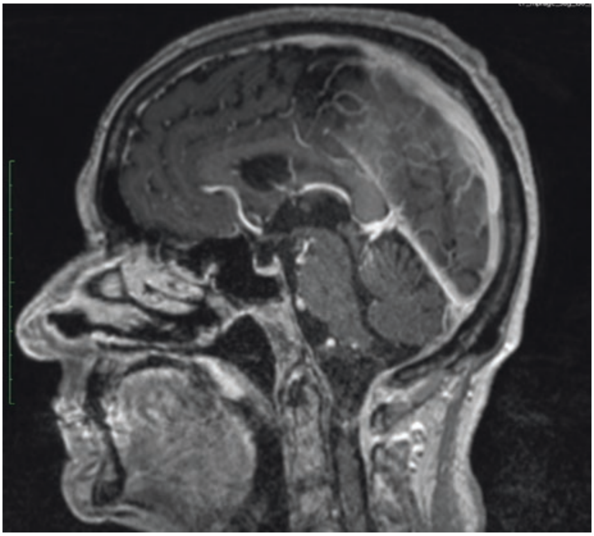

Fig. 27.6 Sagittal Gd-enhanced T1-weighted MR image of the same patient as in Fig. 27.4. The mass at the cranio-cervical junction can be seen as a mere “negative” defect of the medulla oblongata

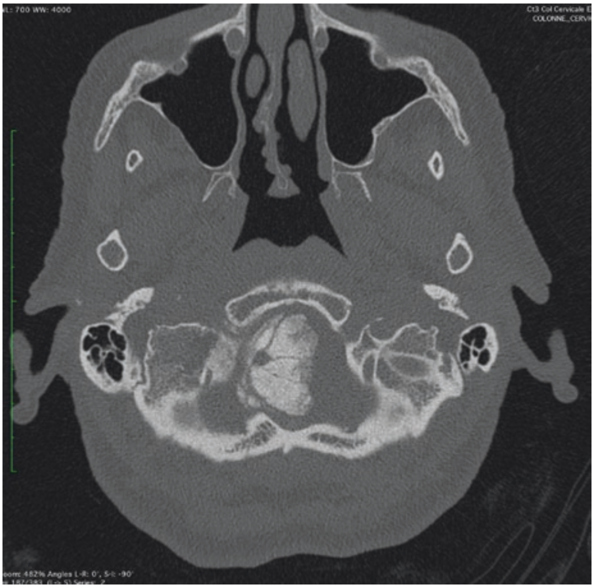

Fig. 27.7 Axial CT bone scan of the same patient as in Fig. 27.4. Extensive calcification can be seen obstructing the foramen magnum in a right-to-left direction

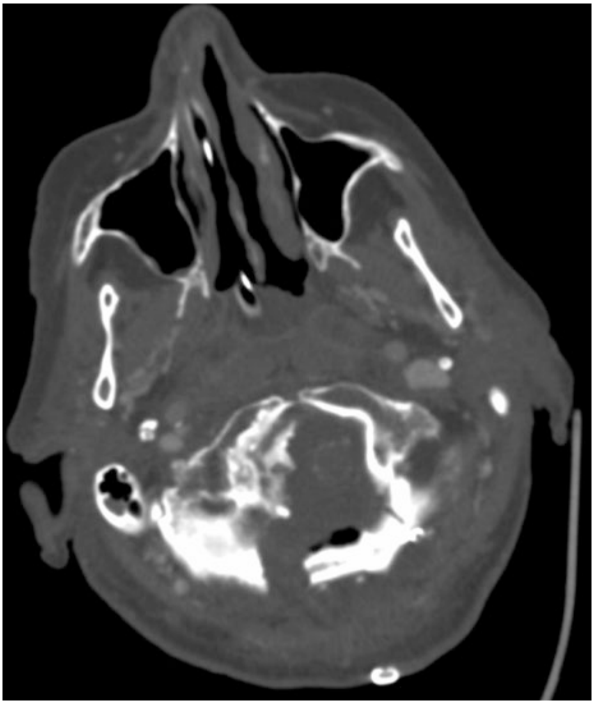

Fig. 27.8 Post-operative axial CT bone scan of the same patient as in Fig. 27.4. A hyperostotic shell is remaining along the dura and around the vertebral artery following subtotal removal of the calcified meningioma, which could be removed with extensive drilling only. Decompression of the brainstem has been achieved

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