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Clinical Article

Management of benign craniovertebral junction tumors

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Summary

Background. We report our surgical experience in the treatment of fifteen consecutive patients with benign craniovertebral junction tumors, observed from 1993 to 2000 at our department.

Method. We treated 7 meningiomas, 3 epidermoids, 3 C1 neurinomas and 2 neurinomas of the lower cranial nerves. Clinical results were evaluated by Karnofsky Performance Scale and all patients underwent preoperative neuroradiological evaluation with CT, MRI and MRA; angiography was not routinely performed and was considered for each individual case.

Findings. 11 partial transcondilar and 4 retrocondilar approaches were performed. Total removal was achieved in 11 cases (73,3%) and subtotal removal in 4 patients (26,7%). None of the patients required occipitocervical fusion. Patients were followed for an average period of 24 $\pm$ 31 months. Clinical and radiological follow-up showed no recurrence in cases with total removal. In all patients a statistically significant postoperative increase of KPS scores was recorded. The treatment of epidermoid tumors presented particular issues: debulking the lesion, we obtained a surgical window, avoiding a large removal of bone. In Nakasu grade 1 or 2 meningiomas, we carried out total removal by piecemeal resection and without complete condylectomy and bone fixation.

Interpretation. The choice of these approaches and the extent of bone resection should be defined according to the tumor's location and size. Moreover we emphasize that preoperative neuroradiological evaluations on presumptive tumor type could be helpful to the surgeon in order to tailor the technique to different lesions, providing the required exposure, without unnecessary surgical steps.

Keywords: Craniovertebral junction tumors; occipitocervical fusion; craniocervical approach.

Introduction

The most frequent neoplastic lesions of the craniovertebral junction are meningiomas, neurinomas, chordomas, paragangliomas, epidermoids, dermoids and chondrosarcomas. Each lesion has a different growth pattern, with different bony destruction and neurovascular structures involvement, requiring a specific surgical approach and technique [16, 17]. Indeed, these lesions often involve and encase very important neural and vascular structures, such as the vertebrobasilar system and lower cranial nerves (IX, X, XI and XII) [4, 7, 11]. Preoperative neuroradiological evaluation with CT, MRI and MRA is necessary in order to tailor the approach to each lesion [5]. Surgical management of craniovertebral junction tumors is challenging for the neurosurgeon. Nevertheless, new posterolateral and anterolateral approaches to craniovertebral junction tumors have been recently developed, with the aim of reducing operative mortality and morbidity [16, 17, 19]. Our aim is to define the surgical approach on the basis of histopathological considerations on neuro-imaging appearance of these lesions and in our experience we emphasize the importance of this aspect in order to choose the appropriate technique for the surgical management. Nowadays, many variants of the lateral approach to craniovertebral junction are available. Nevertheless, retrocondylar and partial transcondylar approaches should be preferred to minimize postoperative neurological deficits and reduce surgical time. We prefer the partial transcondilar approach (PTCA) because of better access and control of the vertebral artery, as well as a shortened operating time.

Methods and patients

We retrospectively analyzed our series of fifteen consecutive patients in whom surgery for benign craniovertebral junction tumors was performed over a 7-year period (January 1993 through February 2000) (Table 1). There were 7 meningiomas, 3 epidermoids, 3 C-1 neurinomas and 2 lower cranial nerves neurinomas. These lesions developed both within the posterior fossa and cervical region intra

Table 1. *Patients*

Age	16–70 years (mean age: 52,26)
Sex	11 women, 4 men
	female/male ratio = 2,5 : 1
Tumors	meningiomas: 7 (46,6%)
	epidermoids: 3 (20%)
	C1 neurinomas: 3 (20%)
	lower cranial nerve neurinomas: 2 (13,3%)
Symptoms and signs on admission	gait disturbance: 7 (46,6%)
	occipital pain/headache: 7 (46,6%)
	diplopia: 2 (13,3%)
	swallowing difficulties: 4 (26,6%)
	XI c.n. palsy: 2 (13,3%)
	tongue weakness or spasms: 2 (13,3%)
Surgical approach	retrocondylar (RCA): 4 (26,6%)
	partial transcondylar (PTCA): 11 (74,4%)

and extradurally. The average age of patients was 52,26 years. The series was comprised of 11 women (73,3%) and 4 men (26,7%); female sex predominated by a factor of 2,5 : 1. The average length of hospitalization was 15 days. The time between the first occurrence of symptoms and the surgery ranged from 1 month to 5 years. On admission the most frequent symptoms and signs were gait disturbances (7/15), occipital pain/headache (7/15), swallowing difficulties (4/15), diplopia (2/15), tongue weakness or spasms (2/15) and XI c.n. palsy (2/15). No patient had undergone previous surgery. Patients with meningiomas presented with gait disturbances in 57,14% (4/7), headache/occipital pain in 28,57% (2/7), swallowing difficulties and diplopia in 14,28% (1/7). Gait disturbances and occipital pain/headache were the most frequent presenting symptoms in patients with epidermoid 66,6% (2/3). Headache/occipital pain was present in 66,6% (2/3) of patients with C1 neurinoma. Eventually, deficit of XI c.n. was present in all the patients with lower cranial nerve neurinoma; these patients presented also with a deficit of XII and IX cranial nerves. All patients preoperatively underwent cerebral CT, MR and MRA. The Karnofsky Performance Scale was used to evaluate the patient's clinical course: the preoperative median value was 50. The differences between the preoperative and a follow-up KPS scores were examined by conducting a Wilcoxon Signed Rank test.

Patients have been operated on through the following approaches: 11 partial transcondilar approach (PTCA) and 4 retrocondilar approach (RCA). All meningiomas, except case No. 4, were approached with PTCA (Figs. 1–3). All epidermoids were resected via the RCA (Figs. 4, 5), whereas all neurinomas were operated on by PTCA (Figs. 6, 7). Total removal was defined as complete resection of the tumor mass. Subtotal removal was defined as resection of the tumor mass with small tumor remnants left behind near important structures, such as the vertebral artery, perforating arteries and cranial nerves. In PTCA procedures the portion of drilled condyle did not reach the 50%, mark to minimize the risk of craniovertebral instability. At surgery, management of the vertebral artery, identification and preservation of XII cranial nerve and condyle resection were three principal steps. The management of the vertebral artery was laborious: in our PTCA cases we drilled the occipital condyle, reflecting medially and inferiorly the extradural segment of VA, away from the C-1 foramen. Depending on the disease, the posterior arch of the atlas as well as the posterior aspect of foramen transversarium and the lateral atlas mass were resected. After completing the bone removal, we opened the dura, leaving a small dural cuff around the VA. The posterior transposition of the intradural segment of the VA and also to mobilize its branches and the XI cranial nerve, creating a large working window to the lower clivus between the XI cranial nerve and C-2 for the removal of the lesion. Postoperatively, all patient underwent clinical examination; enhanced CT and MR were performed before discharge and after 3 and 12 months. Surgical morbidity was defined as a new postoperative neurological deficit or a worsening of pre-existing symptoms and signs without subsequent recovery.

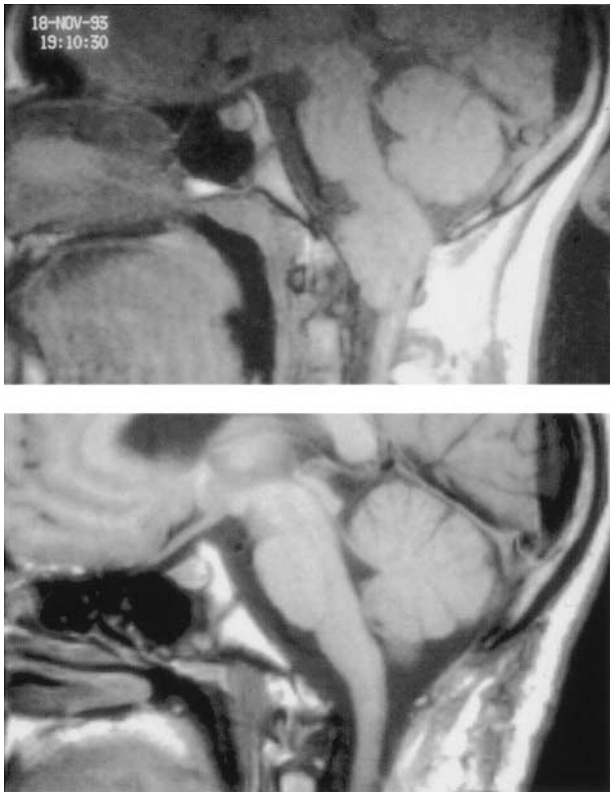

Fig. 1. Case no. 1. Preoperative (*upper*) and postoperative (*lower*) midsagittal MR T1-weighted images of craniocervical meningioma

Results

In our series total removal was achieved in 11 cases (73,3%), subtotal resection in 4 cases (26,7%). Total removal was obtained in 91% of cases undergoing PTCA and in 25% of patients operated via RCA. Total removal was achieved in 6 (85,7%) of the 7 patients with meningioma, in all C1 neurinomas and in two lower cranial nerve neurinoma. Subtotal resection was obtained in one meningioma and in all epidermoids; in these four patients, the tumor capsule was tightly adhered to the brainstem, the vertebral and basilar arteries and their perforating branches. We never needed to perform occipitocervical fusion.

Surgery-related complications occurred in eleven patients. Seven patients (46,6%) presented swallow-

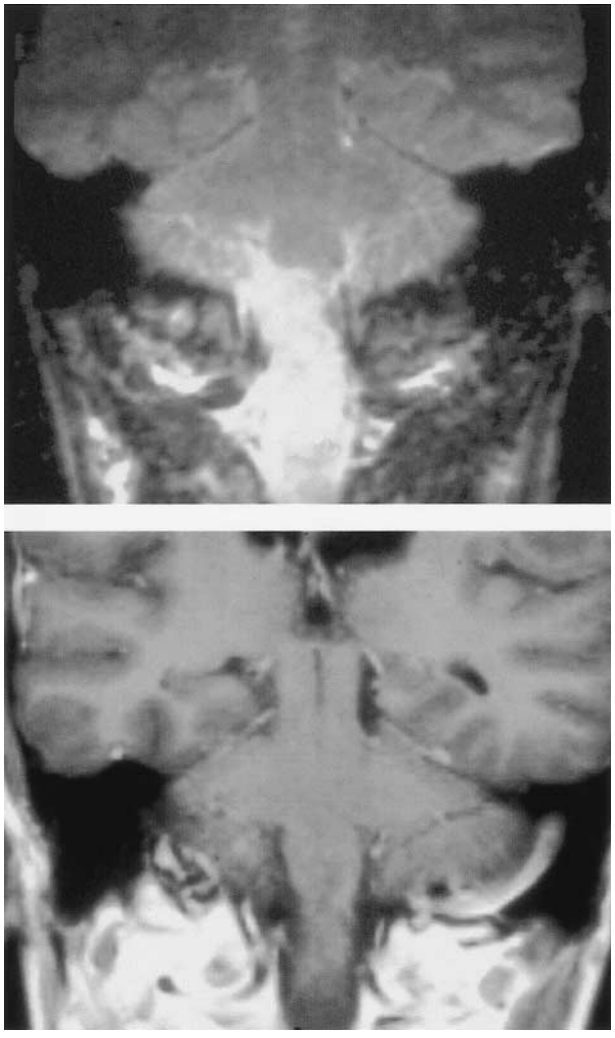

Fig. 2. Case no. 1. Preoperative (*upper*) and postoperative (*lower*) coronal MR images of craniocervical meningioma

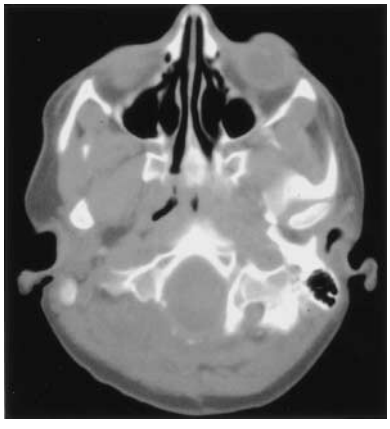

Fig. 3. Case no. 1. Postoperative bone algorithm axial CT scan showing partially drilled right condyle

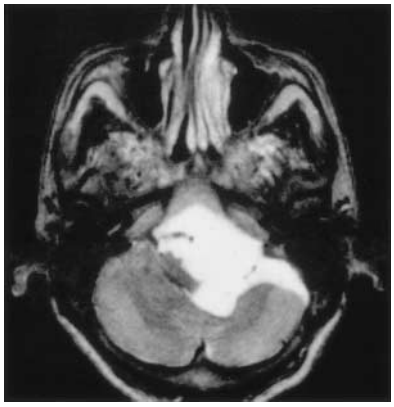

Fig. 4. Case no. 9. Preoperative axial MR T1-weighted image of left epidermoid

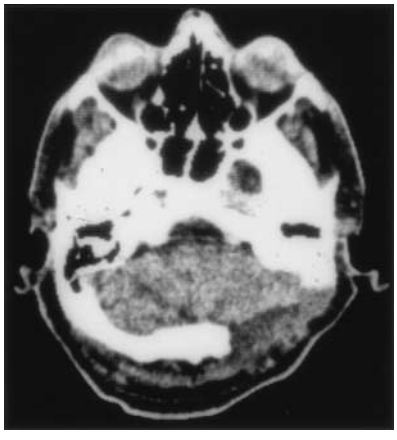

Fig. 5. Case no. 9. Postoperative axial CT scan of left epidermoid

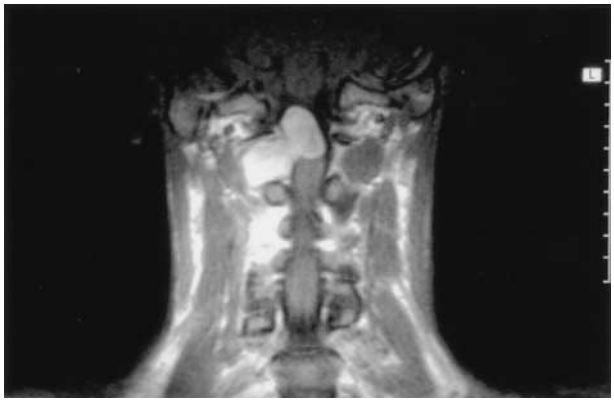

Fig. 6. Case no. 12. Preoperative enhanced coronal MR image of C1 neurinoma

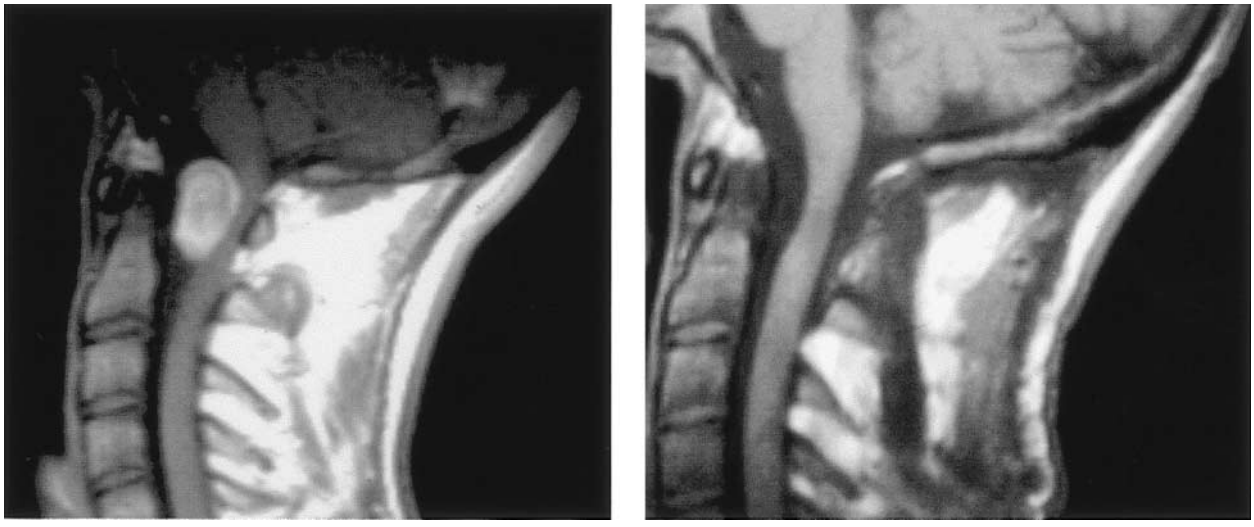

Fig. 7. Case no. 12. Preoperative (*left*) and postoperative (*right*) midsagittal enhanced MR images of C1 neurinoma

ing difficulties. Among these, we observed worsening of preoperative dysphagia in 3 patients (20%), and appearance of new swallowing difficulty in 4 cases (26,6%). According to tumor type, dysphagia occurred in 28,5% of meningiomas, 100% of epidermoids, 33,3% of C1 neurinomas and 50% of lower cranial nerve neurinoma. According to the surgical approach, dysphagia occurred in 36,3% (4/11) of PTCA and 75% of RCA. The postoperative worsening of pre-existing dysphagia was from mild to severe in two cases and from mild to moderate in one case. In all these patients resolution of swallowing difficulties was complete, except in two patients who needed dysphagic diet after 6 months. Two patients developed hydrocephalus which required placement of a ventriculoperitoneal shunt; both patients were affected by epidermoid. CSF leakage was observed in three cases, treated by external lumbar drainage; CSF leak occurred in two patients with meningioma and in one case of C1 neurinoma.

In all but two patients (86,6%) an improved KPS score was obtained during the follow-up period compared with that obtained preoperatively; the median value increased from 50 to 80, indicating that most patients were able to live independent lives postoperatively and benefited significantly from surgery. Overall, however, the improvement varied from scores of 10 to 40. A KPS score improvement of 10 points was observed in 2 cases (13,3%), 20 points in 4 (26,6%), 30 points in 6 (40%), and 40 points in case No. 1. The postoperative improvement of KPS scores was highly statistically significant with a tied Z-value of $-3{,}225$ and a tied P-value < 0.0013. There were no peri-operative deaths. Two patients died in the follow-up period, both because of diseases independent from the craniovertebral tumor. An overview of clinical and surgical results is presented in Table 2.

With regard to histological findings, the most common histolgical type among meningiomas was the meningotheliomatous type (80%). The transitional, psammomatous and angiomatous type were much less common. In all cases predicted tumor type was confirmed by postoperative histological evaluations.

Clinical and radiological follow-up did not evidence of any lesion recurrence in the cases in whom we obtained total removal.

Discussion

The treatment of craniovertebral junction tumors usually presents surgical difficulties related to: 1) access to ventral surface of the lower clivus and craniocervical junction, 2) involvement of the VA at its entry into the dura and involvement of the lower cranial nerves. Hammon in 1972 and Heros in 1986 proposed a lateral suboccipital approach to perform a more lateral suboccipital exposure for vertebrobasilar aneurysms [12, 13]. Since then, the extreme-lateral craniocervical approach has been progressively developed as a more and more lateral approach to provide access to lesions in the midline on the anterior aspect of the foramen magnum. The indications for the posterolateral approach are the presence of intradural neo-

plasms located entirely anterior to the brain stem and cervical spinal cord, and the presence of aneurysms of the vertebral artery and proximal basilar trunk. Extradural lesions of the clivus and upper cervical canal might be approached with this exposure [1, 3, 9, 18, 21–23]. This route, when combined with the retrosigmoid approach, allows control of both intradural and extradural tumors involving the jugular foramen and the upper clival region [18, 21]. Sekhar and colleagues have recently described six variants of the extreme-lateral approach to occipito-cervical junction lesions: retrocondylar, partial transcondylar, complete transcondylar, extreme-lateral transjugular and transtubercular in 69 patients [19]. We used, as Sekhar, the retrocondylar approach (RCA) to reach intradural lesions located anterolateral to the spinomedullary area. The partial transcondylar approach (PTCA) allows one to remove intradural lesions located anterior to spinomedullary area. We never performed, in these cases, the transfacetal approach (TFA), neither the extreme-lateral transjugular approach (ETJA) or the complete transcondylar approach (CTCA). In our PTCA cases we drilled the occipital condyle, reflecting medially and inferiorly the extradural segment of the VA, away from the C-1 foramen. The posterior transposition of the intradural segment of the VA also mobilized its branches and the XI cranial nerve, creating a large working window to the lower clivus between the XI cranial nerve and C-2 for the removal of the lesion [4, 19]. The approach to craniovertebral junction tumors, located anterolaterally to the brainstem, required extensive posterior and lateral bone resection which includes the lateral rim of foramen magnum and the posterior aspect of the occipital condyle. Depending on the disease, the posterior arch of the atlas as well as the posterior aspect of foramen transversarium and the lateral atlas mass can be resected [8, 16, 17]. However, if a significant portion of the lateral mass of C1 and/or a large facetectomy is made at C2, postoperative instability will result; for this reason, a fusion must be created on the opposite side. Fusion can also be performed on the same side, spanning the operative defect and extending one vertebral level below it. Occipitocervical fusion was performed in 21,7% (15/69) of patients treated by Salas, 11 of them after the CTCA, 2 after the TFA and two after the PTCA [19]. Vishteh and colleagues have recently presented a biomechanical study on the stability of the craniovertebral junction after unilateral occipital condyle resection [24]. They found that resection of 50% or more of the occipital condyle produced statistically significant hypermobility at the occipitocervical junction. After a 75% resection, the biomechanics of Oc-C1 and C1–2 motion segments change considerably. Therefore, performing fusion of the craniovertebral junction should be considered if half or more of one occipital condyle is resected. When a dumbbell neurilemmoma is encountered (Figs. 6, 7), the surgeon exposes the neural foramen of the involved level by drilling away the adjacent, superior, and inferior bone. The intradural extended neurilemmoma determined the level at which the posterior facetectomy may be performed [16].

A careful preoperative neuroradiological evaluation of the lesions is essential to identify some histopathological aspects leading to better surgical management of these lesions [5]. The choice of approach and the extent of bone resection should be defined according to the location and size of the tumor. Moreover we emphasize the role of predicting histological tumor type. Indeed, the treatment of epidermoid tumors presents particular aspects (Figs. 4, 5): debulking the lesion, we obtain a surgical window, avoiding large removal of bone. In 1989 Yasargil emphasized that in patients with extensive tumor growth, a single-stage operation was possible because of the manner in which it expands through the subarachnoid spaces [26]. On removing the tumor, a ''surgical channel'' was created, allowing successive access to more extensive areas. Furthermore, in case of large tumors displacing the midbrain controlaterally, a ''natural'' widening of the surgical way is available. In Nakasu grade 1 or 2 meningiomas, we obtained the total removal, after deafferentation of arterial supplies, by piecemeal resection and without complete condylectomy and bone fissation. With regard to foramen magnum meningiomas (Figs. 1–3), it is crucial to evaluate the tumor-brainstem interface and vascular feeders in order to minimize postoperative neurological deficits and reduce surgical time. Meningiomas with a favourable cleavage plane require a more limited approach than en-plaque growing or recurrent meningiomas, in which the finding of arachnoid scarring should require a more extensive approach with eventual stabilization [2, 6, 10, 14, 15, 20, 25].

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Comments

This is a retrospective report on 15 patients with craniocervical tumours operated on during 8 years in one centre by the lateral transcondylar or retrocondylar approach. There were 7 meningiomas, 5 neurinomas and 3 epidermoids; most of the patients were in very bad preoperative condition with mean Karnofsky (KPS) scale 50! All epidermoid tumours were successfully operated on by the retrocondylar route, all other tumours except one meningioma needed at least partial resection of the condyle. The limit of condylotomy was 50% and no instability was later seen. The follow-up was mean 2 years, and no recurrences were seen during that time, and the KPS scores improved in all. The message is, that a satisfactory result may be achieved with these neurologically invasive and technically difficult tumours without perhaps too heroic and destructive lateral bone resection. The message is well argued, however, it could have been emphasised more, that actually all the epidermoid tumours were operated on without condylotomy, and PTCA was used in all meningiomas and neurinomas (except one).

M. Vapalahti

The authors report their surgical experience in 15 patients with benign craniovertebral junction tumors (7 meningiomas, 5 neurinomas and 3 epidermoids). In a lengthy discussion they discuss the merits and demerits of the different approaches they used and which are described in the literature. The clinical information presented in this paper is not without interest, however, because of the very restricted number of patients no firm conclusions can be based on it. The combination of their own clinical experience as presented in this paper and the information from the literature enabled them to suggest different approaches for different types of tumors and for different locations around brainstem-spinal cord transition.

My personal opinion is that most of the benign tumors located at the craniovertebral junction can be removed by way of a simple posterior fossa decompression including the posterior aspect of the foramen magnum and the posterior arch of C1. It is always a surprise how easy a meningioma that is located at the anterior rim of the foramen magnum can be removed by this approach without the necessity to take refuge to the transcondylar or related, very elaborate, approaches. This holds true of course also and in a higher degree for neurinomas and (epi)dermoids.

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