Foramen magnum meningioma (FMM)

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Foramen magnum meningiomas (FMMs) are slow growing, most often intradural and extramedullary tumors.

Epidemiology

They represent 2% of all meningiomas [1)]

Classification

Petroclival and foramen magnum meningiomas are classified according to their Tumor Equivalent Diameter.

The compartment of development was most of the time intradural (101/107, 94.4%) and less frequently extradural (3/107, 2.8%) or both intra-extradural. (3/107, 2.8%). When developed inside the intradural compartment, FMMs were subdivided into posterior (6/104, 5.8%), lateral (57/104, 54.8%), and anterior (41/104, 39.4%), if their insertion was respectively posterior to the denticulate ligament, anterior to the dentate ligament without or with extension over the midline. Anterior and lateral intradural lesions grew below (77/98, 78.6%), above (16/98, 16.3%), or on both sides (5/98, 5.1%) of the VA. Only three cases of extraduralFMMs (3/107, 2.8%) were resected by an antero-lateral approach while all the other ones (104/107, 97.2%) were removed successfully by a postero-lateral approach. Lower cranial nerves were displaced superiorly in FMM growing below the VA but their position cannot be anticipated in other situations.

This classification system helps for defining the best surgical approach but also for anticipating the position of the lower cranial nerves and therefore for reducing the surgical morbidity [2)].

Pathology

They have traditionally been said to involve the lower third of the clivus and the C1 C2 area. However, the last categorizations are arbitrary.

There are some tumors that involve the entire clivus, and others that involve the mid and lower third of the clivus. (The upper clivus is the area above the trigeminal root, the mid-clivus extends to the level of the glossopharyngeal nerve, and the lower clivus is the region below the glossopharyngeal nerve).

Clinical Features

The indolent clinical course of FMMs and their insidious onset of symptoms are important factors that contribute to delayed diagnosis and relative large size at the time of presentation. Symptoms are often produced by compression of surrounding structures (such as the medulla oblongata, upper cervical spinal cord, lower cranial nerves, and vertebral artery) within a critically confined space

Diagnosis

When diagnosed, these tumors are often large. However, due to their slow-growing rate and indolent course, partly attributable to the wide subarachnoid space at this level, clinical diagnosis is difficult, leading to a long interval since the onset of the first symptom [3)].

Magnetic resonance imaging (MRI) with angio-MRI to study the relationship between tumour and vertebral artery (VA) is recommended.

Before the MRI Metrizamide computerized tomography scanning and Pantopaque myelography have been the radiographic tests most commonly used to evaluate the foramen magnum [4)].

Treatment

Since the initial pathological description of a FMM in 1872, various surgical approaches have been described with the aim of achieving radical tumor resection [5)].

Foramen magnum (FM) meningiomas are challenging lesions because of the vicinity of the medulla oblongata, the lower cranial nerves, and the vertebral artery.

The surgical treatment of FMMs has evolved considerably due to the progress in microsurgical techniques and development of a multitude of skull base approaches.

Posterior and posterolateral FMMs can be safely resected via a standard midline suboccipital approach. However, controversy still exits regarding the optimal management of anterior or anterolateral lesions.

Based on anatomical and surgical constations it appears that a complete resection of the occipital condyle (resulting in occipito-cervical instability) should be reserved for those very extensive lesions. Yet a partial drilling of the condyle provides a better angle of approach, minimises the hazards of retraction of nervous structures and enables the surgeon to take the best advantage of the dissection and control of the vertebral artery [6)].

The main advantage of the dorsolateral, suboccipital, transcondylar route is the direct view it offers to the anterior rim of the foramen magnum without requiring brain stem retraction [7)].

Posterior FMMs

Posterior and posterolateral FMMs can be safely resected via a standard midline suboccipital approach.

Independently of technical variations and the degree of bone removal, all modern surgical approaches to the lower clivus and anterior foramen magnum derive from the posterolateral (or far-lateral) craniotomy originally described by Roberto Heros and Bernard George [8].

Ventral and ventrolateral FMMs

Ventrally located tumors have been considered to require other complex approaches to ensure surgical corridor such as the postero-lateral approach, called the far-lateral approach or the antero-lateral approach, also named the extreme-lateral approach [9],

Transoral approach has also been used for ventral FMMs. Conventional posterior suboccipital approach has also been used for anterior FMMs.

Far lateral approach, also called posterolateral approach or lateral suboccipital approach, is commonly used to resect ventral FMMs.

Condyle drilling varies from one third to one half of the condyle, in far lateral and extreme lateral approaches.

Transcondylar approaches provide a significant greater area of exposure than the retrosigmoid approach.

Extent of removal of occipital condyle can be individualized according to the case. In some of the FMMs, removal of condyle might not be necessary.

Large ventral FMMs without spinal extension can be easily approached by conventional retrosigmoid suboccipital craniectomy without requiring partial condylectomy as the size of the tumor provides great surgical corridor for tumor removal.

Since Goel et al. [10]. introduced the posterior approach for FM meningiomas, there have been only a few reports of the posterior approach in ventral FM meningiomas [11] [12] [13].

The far lateral and extreme lateral approaches are difficult and unfamiliar to many surgeons. Lateral bony exposure also may pose risk to the vertebral artery and low cranial nerve injury. The occipital condyle drilling may sometimes be necessary in the far lateral approach. Therefore, conventional posterior approach without lateral bony extension may be good to address these problems.

Ventral foramen magnum (FM) meningiomas can be removed gross totally using a posterior approach without far lateral approach. The arachnoid membrane can then be exploited as an anatomical barrier. However, this approach should be taken with a thorough understanding of its anatomical limitation [14].

Innovation

The primary innovation consists in the use of the lateral approach, with the addition of partial resection of the lateral mass of C1. Baranowski etal., present the case of a patient with a meningioma located at the level of the dens of the epistropheus, on the anterior surface of the spinal cord. The lateral approach used in this case allowed for complete resection of the tumour with minimum operative risk, and the patient's recovery has been excellent [15].

Case series

2015

During two years, Mostofi operated 5 patients.

All the patients had magnetic resonance imaging (MRI) with angio-MRI to study the relationship between tumour and vertebral artery (VA). In all the cases, operated in prone position.

In one case, considering the tumour localization (posterior and pure intradural) the tumour was removed via a midline suboccipital approach with craniotomy and C1-C2 laminectomy. In all other cases, meningiomas were posterolateral (classification of George) with extradural extension in one case. In all cases, VA was surrounded by tumor and opted for a modified postero-lateral approach with inverted L incision, craniotomy and C1-C2 laminectomy without resecting the occipital condyle. Epidural part of VA was identified and mobilized laterally. Once VA was identified he opened dura mater and began to remove the tumour [16].

2014

Thirteen patients (11 Feminine / 2 Masculine with FM meningiomas operated on through lateral suboccipital approach were studied. Clinical outcome were analyzed using survival (SC) and recurrence-free survival curves (RFSC).

All tumors were World Health Organization grade I. Total, subtotal and partial resections were acchieved in 69.2%, 23.1% and 7.7%, respectively, and SC was better for males and RFSC for females. Tumor location, extent of resection and involvement of vertebral artery/lower cranial nerves did not influence SC and RFSC. Recurrence rate was 7.7%. Operative mortality was 0. Main complications were transient (38.5%) and permanent (7.7%) lower cranial nerve deficits, cerebrospinal fluid fistula (30.8%), and transient and permanent respiratory difficulties in 7.7% each.

FM meningiomas can be adequately treated in public hospitals in developing countries if a multidisciplinary team is available for managing postoperative lower cranial nerve deficits [17].

2009

16 patients with foramen magnum meningiomas were operated in all cases by a posterior suboccipital approach with lateral extension of the bone opening according to the position of the tumour. In 14 patients, intraoperative monitoring of the lower cranial nerves was performed. Localisation of the tumours was ventral (3), ventrolateral (10), dorsal (1) and dorsolateral (2). Mean age of the patients was 61 years (ranging from 40 to 85 years). Preoperative and postoperative function was classified according to the McCormick scale.

Eight patients experience a postoperative upgrading of at least one grade, in five patients an unchanged status and a deterioration in only two patients. Complete removal of the tumour was possible in 14 cases (Simpson 1-2). The follow-up period varied from 24 to 119 months (mean 43.5 months), during this time there were no recurrences. In there experience, the posterior suboccipital approach is suitable for the majority of these tumours [18].

2006

Clinical data in a consecutive series of 25 patients experiencing a meningioma attached to dura of the anterior or anterolateral FM rim were retrospectively reviewed.

The most common symptoms of the 19 women and six men (mean age, 59.2 yr) was cervico-occipital pain (72%) and gait disturbance (32%). Clinical examination revealed gait ataxia in 48% of the patients. As depicted from preoperative magnetic resonance imaging (MRI), dural attachment of the meningioma at the FM rim was anterior in 36% and anterolateral in 64% of cases. Tumor removal was accomplished via a posterolateral suboccipital retrocondylar approach in all patients. A Simpson Grade 2 resection was achieved in 96% of the patients. Permanent surgical morbidity and mortality rates were 8 and 4%, respectively. No tumor recurrence was observed after a mean follow-up period of 6.1 years (range, 1-14 yr) with clinical and MRI examination, and 80% of the patients have regained full daily activity.

Anterior and anterolateral FM meningiomas that displace the medulla/spinal cord can be safely and completely resected via a posterolateral suboccipital retrocondylar approach. A tumor remnant should be left on critical neurovascular structures in cases with poor arachnoid dissection planes [19)].

2004

Eleven cases with the foramen magnum meningioma were operated by using posterior approach with lateral extension.

Complete removal of the tumor was performed in 7 patients (7/11, 64%), subtotal resection in 2 cases (2/11, 18%) and partial resection in 2 cases (2/11, 18%). There were no operative death and significant complication.

The posterior approach with lateral enlargement is sufficient to expose and remove foramen magnum tumors without expensive bone resection [20)].

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22 patients underwent 23 surgical procedures with a diagnosis of foramen magnum meningioma at Marmara University, Department of Neurosurgery. The suboccipital approach was used for 2 posteriorly located tumors with radiological total removal. The paramedian suboccipital approach was replaced by the far-lateral modification in the treatment of ventral meningiomas. 1 of the 20 ventral tumors was operated twice. The classical suboccipital approach was followed by the far-lateral modification. A gross-total removal was achieved in 21 patients. The overall morbidity was 32%. No specific and clinically significant complications attributable to the far-lateral modification were observed. The far-lateral approach has improved the success of surgery in ventrally located lesions. The posterior suboccipital approach is still indicated in the removal of lesions placed posterior to the dentate ligament [21)].

2002

Seven cases of foramen magnum meningioma (1.4%). All patients showed various neurological symptoms corresponding with foramen magnum syndrome. The tumor locations were anterior in five

cases and posterior in two. Surgical removal was performed through a transoral approach in one patient, the suboccipital approach in three, and the transcondylar approach in two. Total removal was achieved in all patients, except for one who refused any surgical treatment. The major complications were tetraparesis and lower cranial nerve paresis for tumors in anterior locations, and minor complications for posterior locations. One patient died of atelectasis and pneumonia after a long hospitalization. The transcondylar approach is recommended for anterior locations, and the standard suboccipital approach for posterior locations [22)].

2001

17 patients with foramen magnum meningiomas arising from the anterior or anterolateral rim of the foramen magnum underwent operations in the Department of Neurosurgery at King Edward Memorial Hospital and Seth G.S. Medical College. All patients were operated on in a semi-sitting position by use of a conventional suboccipital approach with a midline incision and extension of the craniectomy laterally toward the side of the tumor up to the occipital condyle.

The patients ranged in age from 17 to 72 years, and the tumors ranged in size from 2.1 to 3.8 cm. The intradural vertebral artery was at least partially encased on one side in eight patients and on both sides in two patients. The brainstem was displaced predominantly posteriorly in each patient. A partial condylar resection was performed in two cases to enhance the exposure. Total tumor resection was achieved in 14 patients, and a subtotal resection of the tumor was performed in the other 3 patients. In one patient, a small part of the tumor was missed inadvertently, and in the other two patients, part of the tumor in relation to the vertebral artery and posterior inferior cerebellar artery was deliberately left behind. After surgery, one patient developed exaggerated lower cranial nerve weakness. There was no significant postoperative complication in the remainder of the patients, and their conditions improved after surgery. The average length of follow-up is 43 months, and there has been no recurrence of the tumor or growth of the residual tumor.

From there experience, they conclude that a large majority of anterior foramen magnum meningiomas can be excised with a lateral suboccipital approach and meticulous microsurgical techniques [23)].

1996

38 patients who were operated on for 40 meningiomas of the craniocervical junction between September 1977 and August 1995

Radiological examinations, clinical data, and operation notes were evaluated, and additional follow-up information was obtained from outpatient examinations, telephone calls, and questionnaires.

Four groups could be distinguished according to dural attachment as follows: 1) 15 spinocranial meningiomas originated from the spinal canal and extended intracranially; 25 craniocervical meningiomas of intracranial origin were divided into 2) meningiomas of the lower clivus (10 patients with 11 tumors), 3) lateral meningiomas (11 patients with 12 tumors), and 4) posterior meningiomas (2 patients). Standard midline or lateral suboccipital approaches with opening of the foramen magnum and laminectomy of the involved cervical segments were sufficient for the great majority of tumors. In seven instances only, drilling the posterior third of an occipital condyle was needed. Twelve of 15 spinocranial meningiomas and 13 of 25 craniocervical meningiomas could be removed totally.

One patient underwent ventriculoperitoneal shunting only. With a rate of 63% of totally removed and 30% of subtotally removed meningiomas in this region, we observed clinical recurrences for two patients only. Complications were encountered in 30% of patients, predominantly with recurrent and/or infiltrative or en plaque meningiomas. Whereas motor weakness and gait ataxia tended to improve postoperatively, cranial nerve deficits usually remained unaltered.

The relationship of the tumor to neighboring structures, i.e., the vertebral artery in particular, determines its resectability [24].

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19 patients with ventral or ventrolateral foramen magnum meningiomas operated on via the dorsolateral, suboccipital transcondylar access route. It is emphasized that the microsurgical management of these lesions includes two important aspects which increase the safety of the procedure: a meticulous preoperative planning based on the microanatomical details of each patient, as well as an individualized tailoring of the surgical approach. There were no deaths, and, in the past 5 years, no neurological complications in this series. Gross total removal of the tumour was achieved in each case. It is concluded that microsurgical removal of ventral or ventrolateral foramen magnum meningiomas with this technique constitutes a safe and recommendable procedure [25].

1990

Three cases of foramen magnum meningioma. The first involved a ventral type tumor extending to the second cervical body. Following bilateral mandibulotomy, surgery was performed via the anterior transoral approach and the tumor was totally removed. Nine days postoperatively, she developed meningitis, which was successfully treated with antibiotics. The second patient's tumor was dorsal type and was deeply embedded in the lateral part of the vermis. The tumor was totally removed via the midline suboccipital approach and she recovered uneventfully, with only slight upper-extremity paresthesia. In the third case, the tumor was ventral type and situated mainly in the clivus. Craniotomy was performed by the bilateral suboccipital approach and extended nearly to the jugular tubercle. The tumor, which severely displaced the lower cranial and upper cervical nerves, was totally removed. The postoperative course was lengthy and complicated. Artificial ventilation was required for 2 months, and difficulty in swallowing persisted during long-term follow-up. As illustrated by the second case, dorsal and lateral type foramen magnum meningiomas can usually be removed via the lateral suboccipital approach. In the case of ventral type tumors, the anterior transoral approach entails the risk of infection, as occurred in the first case. The authors conclude that the lateral suboccipital approach is preferable; craniotomy extending to the jugular tubercle lowers the risk of brainstem damage [26].

1986

Seven cases of the foramen magnum tumors were presented with the clinical manifestations and surgical consideration. Early clinical symptoms of the cases with extramedullary lesions were suboccipital neck pain followed by dysesthesia, clumsiness of hand and weakness. On the other hand, early symptoms of the cases with intramedullary lesions were dysesthesia, often followed by swallowing difficulty or hoarsness, which may have some difference from the clinical course of the extramedullary tumor cases. CT scan was remarkably useful in the diagnosis of the foramen magnum tumor. Surgical treatment was done to 6 cases: Five of these cases were operated by suboccipital

craniectomy, and one case with an anteriorly located meningioma in the foramen magnum region was operated by transoral approach. Total removal could be performed in the case without damage to the medulla or spinal cord [27)].

Case reports

2015

Athanasiou et al. report a rare case of anterolateral meningioma of the foramen magnum (FMM) and high cervical spine presenting both intradural and extradural growth in a 7.5-year-old boy.

The patient presented with progressive tetraparesis and gait instability. Neuroimaging revealed an anterolateral tumor of the foramen magnum, C1 and C2 cervical spine level. The patient was treated in two stages: During the first operation, the extradural part was resected while the intradural part was removed in a second operation. Following the second operation, the patient showed almost complete neurological recovery as a result of cervical spinal cord and brainstem decompression but was complicated with cerebrospinal fluid leakage and infection by Acinetobacter. He sustained two further operations for dural sealing and external ventricular drainage and was treated with intraventricular administration of antibiotics.

Histopathology of the tumor confirmed a meningotheliomatous meningioma. At the 6-month post-op follow-up examination, the patient exhibited complete neurological recovery and no radiological tumor recurrence. To the authors' best knowledge, we report the third case of sporadic pediatric meningioma of the foramen magnum and high cervical compartments with an extradural growth.

Accurate pre-operative estimation of possible extradural growth is crucial towards surgical planning and sufficient treatment. Treatment of choice is total resection in a single operating session to avoid re-operations and increased risk of complications. If not possible, a re-operation should always attempt to secure the desired result [28)].

A 59-year-old woman was admitted with a sudden severe headache that had lasted for 5 days. Neck stiffness was present, but no other neurological deficits were present. Subarachnoid hemorrhage and intra-tumor hemorrhage were not noted on a head computed tomography(HCT). The patient's cerebrospinal fluid was xanthochromic. Magnetic resonance imaging(MRI)demonstrated a gadolinium-enhanced tumor with hemorrhagic changes around the foramen magnum. After conservative therapy, MRI showed a decrease in tumor size and a dural tail sign. This tumor was diagnosed as a hemorrhagic meningioma, and was resected with a posterior suboccipital approach. Histology confirmed that this tumor was a benign transitional meningioma with hemorrhagic change. This is a rare case involving benign meningioma onset by hemorrhagic change. Postoperative tumor recurrence was not present. [29)].

2011

Ghanta and Mohammad report the case of a seventy-year-old female who presented with history of neck pain and weakness in all four limbs for three months. Neurological examintaion showed

quadriparesis with grade 4/5 power, hypertonia and hyperreflexia. Two days before surgery, she devoloped urinary retention and was catheterized. Magnetic resonance imaging (MRI) revealed a large ventral foramen magnum meningioma of size 3.5 × 2.8 × 3.2 cm severely compressing the brainstem.

Pre operative MRI showing large ventral foramen magnum meningioma severely compressing the brainstem

Surgery was done in supine position with head turned to the left and fixed in sugita head frame. The tumor was approached by a linear incision of 9 cm, behind the mastoid process. Right retromastoid suboccipital craniectomy was done. Bone removal was done laterally till the sigmoid sinus was exposed and inferiorly including the foramen magnum. No resection of the occipital condyle was done. Dura was opened in cruiciate manner. Intraoperatively brainstem was severely stretched posteriorly by the tumor. Vertebral artery was seen adjacent to the base of the tumor. Tumor was soft to firm in consistency and intial debulking of the tumor was done. Total excision of the tumor was done(Simpson grade-2) with minimal handling of the brainstem as the tumor size provided great space for microsurgical removal. Post operatively, patient had transient lower cranial nerve palsy and mild left hemiparesis which recovered completely. Post operative MRI revealed complete excision of FMM.

Neurological examination at one year follow-up revealed power of 4+/5 on the left side and no cranial nerve deficits [30].

2009

A 72 year-old woman developed posterior neck pain and a tingling sensation in the left arm. Magnetic resonance imaging showed a well defined and homogenously enhancing mass at the foramen magnum with no dural attachment. Angiography did not demonstrate a blood supply to the tumour via the posterior meningeal branches of the vertebral artery. The mass was totally removed via a midline suboccipital approach. Intraoperatively, the mass was found to adhere to the dentate ligament without a dural attachment. Histopathology findings were consistent with the diagnosis of meningioma.

This case is the first report of a meningioma originating from the dentate ligament and the tumour may have originated from the pia-arachnoidal extension of the spinal cord to the dura [31].

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