Key Aspects in Foramen Magnum Meningiomas: From Old Neuroanatomical Conceptions to Current Far Lateral Neurosurgical Intervention

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Key words
- Far lateral approach
- Foramen magnum meningiomas
- Microsurgical anatomy
- Vertebral artery

Abbreviations and Acronyms
CN: Cranial nerve
FLA: Far lateral approach
FM: Foramen magnum
FMM: Foramen magnum meningiomas
VA: Vertebral artery

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1This publication is dedicated to Dr. Albert Rhoton, Jr. He left us with his amazing spirit of compassion, thoughtfulness, as well as his unique way to see the world and the infinite galaxy that the human train has yet to be discovered.

Citation: World Neurosurg. (2017) 106: 477-483.
http://dx.doi.org/10.1016/j.wneu.2017.07.029

Available online: www.WORLDNEUROSURGERY.org

INTRODUCTION

The foramen magnum (FM) is an anatomical region near the medulla oblongata, at the lower half of the brainstem; importantly, lower cranial nerves (CNs) and vertebral arteries are positioned within this region.1,2 Several techniques describing how to better approach FM lesions have been published. Such techniques include lateral-suboccipital, posterior midline suboccipital, extreme lateral, endoscopic endonasal, and stereotactic radiosurgery, among others.3,4 Foramen magnum meningiomas represent a challenge for neurosurgeons. These tumors require careful surgical manipulation as they are often located in proximity to critical neurovascular structures and the cranial nerves. The far lateral approach is considered the safest neurosurgical approach for excising foramen magnum lesions. It facilitates the access to the anterior foramen magnum and reduces the retraction of vital structures. We describe key historical, epidemiological, genetic, epigenetic, clinical, and neurosurgical aspects of foramen magnum meningiomas. We emphasize the far lateral approach for lesions arising in the foramen magnum, as well as the most appropriate patient positioning for such approach. Caring for these aspects will be rewarded with the best perioperative neurosurgical outcomes.

KEY HISTORICAL ASPECTS

FMMs are histologically considered as common brain tumors within a rare location.5,6 Ancient descriptions on meningiomas were made since Andreas Vesalius’s humani corporis fabrica in 1543.7 The first reported autopsy of a meningioma was made by Salzmann in 1730 on a patient with behavioral changes, headaches, vertigo, and seizures as the main cause of death.8 Illustrations followed in 1849 by Cruveilhier, who published his Traité d’Anatomie Pathologique Générale and his initial description of a fungus duræ mætris.9 In 1851, Lebert named these tumors tumœres fibro-plastiques.10 Further surgical reviews made by Sir James Paget in 1854 classified these tumors as “less malignant than cancer.”11,12 Bennett, in 1858, described the tumor as an epithelial cancer.13 FMMs were first reported as an incidental postmortem finding by Hallopeau from an autopsy made in 1872.14-16 Other key historical descriptions are detailed in Table 1.14,17-20

Harvey Cushing made important advances in the field and in 1902 published an outstanding monography entitled Meningiomas.21 By that time, the removal of a tumor located in the cervicomedullary junction with extension into the posterior fossa was also reported by Frazier and Spiller.22 In 1929, Elsberg and Strauss reported the first surgical resection of a cervicomedullary tumor through a suboccipital craniectomy using a Cr-C3 laminectomy.23 In 1938, Cushing later reported the tumor as a “dural endothelioma,” a tumor arising mainly from the brain and spinal meningeal sheets.24 Cushing also did a comprehensive neurosurgical documentation on the topic based on 313 patients with meningioma, known by that time as a neurosurgical masterpiece.25-37 In 1968, Heros38 introduced the FLA for the management of vertebral artery (VA) aneurysms at the vertebrobasilar junction, proximal basilar trunk, and arteriovenous malformations of the inferolateral cerebellum. In 1988, George et al.39 introduced the FLA for

Table 1:...
Meningiomas are the most common benign and primary brain tumors in adults. The incidence of these tumors is up to 3.2% of all intracranial meningiomas.33 Among them, FMMs represent about 3.2% of all central nervous system malignancies.34

Based on the anatomical compartment, involved FMMs are classified as intradural, extradural, or intradural and extradural.35 Intradural FMMs are then subclassified as anterior, posterior, and lateral.36 Common anatomical locations of FMMs include anterior, followed by anterolateral and posteralateral.37 FMMs are also classified with regard to VA as above, below, or bilateral. In this classification, the most common location of FMM is below the VA.38 Meningiomas represent about 77% of all benign intramedullary, extramedullary, and intradural extramedullary tumors, with most of these tumors located intradural and extramedullary.39 FMMs have also been found incidentally at the craniofacial junction.40-42 The mean age of presentation of FMM in adults is 55 ± 4.2 years old.43-46 Females are usually more affected than males.47-50 Tumor presentation could be classified as aggressive and nonaggressive.44 Usually, benign presentations of these tumors have nonaggressive clinical presentations, whereas local invasion leads to a more aggressive presentation.49

In children, meningiomas in children have an incidence of 0.4%-4.6% of all central nervous system malignancies.40 Almost 4% of these tumors are found in males.45-46 These tumors tend to be located highly at the cervical spine, over the craniofacial junction; when the tumors become symptomatic, symptoms resemble those from adults.57 FMMs are considered a rare type of primary brain tumors in children.40

KEY EPIDEMIOLOGICAL ASPECTS

Meningiomas are the most common benign and primary brain tumors in adults. The incidence of these tumors is up to 3.2% of all intracranial meningiomas.33 Among them, FMMs represent about 3.2% of meningiomas that are located in the FM.34 On the basis of the anatomical compartment, involved FMMs are classified as intradural, extradural, or intradural and extradural.35 Intradural FMMs are then subclassified as anterior, posterior, and lateral.36 Common anatomical locations of FMMs include anterior, followed by anterolateral and posteralateral.37 FMMs are also classified with regard to VA as above, below, or bilateral. In this classification, the most common location of FMM is below the VA.38 Meningiomas represent about 77% of all benign intramedullary, extramedullary, and intradural extramedullary tumors, with most of these tumors located intradural and extramedullary.39 FMMs have also been found incidentally at the craniofacial junction.40-42 The mean age of presentation of FMM in adults is 55 ± 4.2 years old.43-46 Females are usually more affected than men.34-40 Tumor presentation could be classified as aggressive and nonaggressive.44 Usually, benign presentations of these tumors have nonaggressive clinical presentations, whereas local invasion leads to a more aggressive presentation.49

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KEY MOLECULAR ASPECTS

Familial genetic syndromes such as LiFraumeni, Turcot, Gardner, Gorlin, and MEN1 predispose for the presentation of meningiomas.48-50 An association between type 2 neurofibromatosis and mutations in chromosome 22q has been found. In these cases, gene-tumor presentation is related to aggressive tumoral behavior and clinical manifestations rely on histopathological findings and higher malignancy rates.49-53 Epigenetics is another key aspect in FMM. Epigenetics relates to changes in gene expression patterns that do not depend on changes in DNA sequence.53 Epigenetic marks are established during early development and modified during carcinogenesis.54 These cancer-related epigenetic changes are frequently observed in meningiomas that include aberrant promotor methylation, disturbed chromatin organization, and abnormal microRNA expression.55 Some of these aberrant epigenetic changes have been associated with tumor aggressiveness and the potential of invasiveness; such changes have been also associated with clinical stages of the disease.49-55-59 Chromosomal instability, potentially linked to epigenetic aberrations in heterochromatin regions such as centromeric and pericentromeric regions where mutations and aneuploidy correlate, is seen in most solid cancer tumors and should also be considered in meningiomas.57 Cases such as chromosome 1p deletion have been associated with recurrence and progression in meningiomas.50 Additionally, signaling pathway aberrations, such as TFGiROs1 fusion—related tumor growth in the c-ros oncogene 1 (ROs-1) signaling pathway, have often been found in the pediatric population with simultaneous presentation of meningiomas and type 2 neurofibromatosis.39 Gene inactivation of BAM22, BCR, TIM-1 on chromosomal 22q region and MEG3, NDRG2 on chromosome 14q region have been also associated with the presentation of high-grade meningiomas.49-50

KEY CLINICAL ASPECTS

Clinical presentation of meningiomas is location dependent. When the primary lesion is supratentorial, patients tend to develop orthostatic headache, secondary to irritation of the leptomeningeal sheets.60 Should papilledema be found, the course of visual impairment is due to the mass effect these tumors produce over the optic chiasm.61 When a diagnosis of primary extradural meningioma is made, compressive signs and symptoms are present in more advanced stages of the disease depending on location and extent of the tumor.62 Another location-related symptoms include facial nerve palsy, diziness, diplopia, and hearing disturbances.64 In cases where recurrence is found, the Simpson grading system used for meningioma recurrence must be applied.50-57

Symptoms of FMM include gait ataxia, suboccipital headache, and cervical pain. These symptoms are exacerbated by Val-salva maneuvers and cough.65-67 Sometimes FMM may mimic nervous system disorders such as amyotrophic lateral sclerosis, multiple sclerosis, and even compressive-like presentations such as normal pressure hydrocephalus, cervical spondylolysis, carpal tunnel syndrome, and intramedullary tumors.68

FM syndrome due to transoperative changes on lumbar cerebrospinal fluid drainage in Chiari malformation type I has induced quadriaparesis and late-onset cardiopulmonary arrest.16-25 Muscle atrophy due to spinal cranial nerve (CN XI) deficit following FMM compressive effects has been found simultaneously with injury over
lower cranial nerves. Sternocleidomastoid muscle and trapezius muscle atrophy with hypoglossal cranial nerve (CN XII) palsy, hoarseness, dysphagia, and tongue deviation could also appear in cases involving lower cranial nerve compression. Dysarthria, gait ataxia, and hemihyperesthesia have also been described in FMM. In rare occasions, lower limb anomalies along with sudden falls without loss of conscience have also been reported. FMM may also debut as a subarachnoid hemorrhage.

KEY NEUROSURGICAL ASPECTS

Several approaches have been used to remove FMM; however, limitations apply. For instance, the midline approach in suboccipital craniotomy limits the lateral dissection for visualization of the extracranial VA. The suboccipital approach allows visualizing the medulla when a C1 laminectomy needs to be done, as well as when making a posterolateral exposure; however, this approach is limited when an aggressive and extensive exposure is made. The lateral extension of the midline approach is limited due to a major anatomical exposure of key neurovascular structures.

In neurosurgery, patient positioning plays an important role in outcomes. Germane to this work, the head of the patient should be carefully placed in a neutral position. In light of this, prior neck flexion positioning of the individual must be avoided because it worsens the neuroaxis compression. Using the mentioned positioning, the FLA has shown fewer complications compared with approaches such as midline, suboccipital, and posterolateral, which may increase perioperative morbidity due to its aggressive and extensive exposure.

Cerebrospinal fluid fistulae and pseudomeningocele could appear when a major surgical exposure of the hypoglossal canal is made. Intraoperative monitoring of motor and somatosensory evoked potentials is recommended when manipulation of CN VII and CN XI is considered. Skin incision should be carefully addressed considering the approach to the C2 spinous process and throughout the cranio cervical junction.

FLA SURGICAL TECHNIQUE

After adequate patient positioning and trichotomy, antiseptic measures should be carried out with povidone, benzoin, and the like to guarantee safeness when making the approach. A hockey stick–shaped incision is made in the skin with prior zone demarcation; use of bipolar coagulation is recommended to avoid bleeding of scalp vessels. One limb of the incision was made in the midline, beginning below the spinous process of C3 with extension above the inion (Figure 1A). The horizontal incision extends from the inion toward the mastoid process, and then the approach extends below the mastoid process and upward the horizontal incision with ipsilateral scalene exposure (Figure 1B). The transverse sinus and sigmoid sinus is paralleled, providing the possibility of laterally folding a mucocutaneous flap. This approach allows exposing the occiput and arch of C1 out to the transverse process angulation (Figure 1C and D).

After this step is completed, soft tissue is located and periosteal dissectors, as well
as monopolar cautery, are used to expose the anatomical landmarks. Simultaneously, ipsilateral occiput is cleared from the soft tissue to the FM. Next, FM is exposed until the mastoid process curves medially and anteriorly.\textsuperscript{86,87} Ct lamina is exposed laterally until palpation of Ct process below the oblique muscles within the suboccipital triangle.\textsuperscript{88,89} The suboccipital triangle is composed by the rectus capitis posterior major, superior oblique, and inferior oblique muscles; its floor is formed by the posterior atlanto-occipital membrane, where the terminal extradural VA and first and second cervical neural roots are located. The course of the VA is then identified, outlined, and safely mobilized. The intravascular and perivascular plexus should be controlled with cautery. The posterior portion of the transverse foramen is then removed by a drill. It allows releasing of the VA posteriorly and mobilization from the occipital condyle, and protection of the VA with a vessel loop should be carefully addressed.\textsuperscript{8,88-92}

Unilateral suboccipital craniotomy and Ct hemilaminectomy are done by a J-shaped plate of the hemioccipital bone for formation of the bone flap (Figure 1E).\textsuperscript{93} The medial vertical limb of the bone flap then extends upward from the FM to the transverse sinus (Figure 1F). The lateral vertical limb extends from the asterion and then further extends as an inferomedial curved line, reaching the FM as laterally as possible. The horizontal limb connects the upper portions of the vertical limbs and parallels the transverse sinus. A Ct hemilaminectomy lengthens the dural incision, which is made using a side-cutting bur with a footplate (Figure 2A).\textsuperscript{85} Once Ct hemilaminectomy is done, retrosigmoid mastoidectomy is achieved by exposure of the transverse and sigmoid sinuses from the torcular Herophilii to the beginning of the jugular bulb for the superolateral extent of the dural incision. For tumor extraction in the FM, a wider exposure of the dura should be obtained. In this stage, dural dissection is made away from the bone, drill bone is thinned, and removal of the Kerrison punch is completed (Figure 2B).\textsuperscript{85}

In a timely fashion, occipital condyle drilling allows the anterior visualization that the FLA provides. A minimal amount of condyle can be removed before occipito-cervical fusion is considered. Retraction and mobilization of the dural sheet surrounding the VA guarantees preservation of adjacent structures (Figure 2C). Protection of the VA should be carefully done before drilling.\textsuperscript{42} The far lateral exposure also provides access for the following neurosurgical approaches: First, the transcondylar approach directed through the occipital condyle allows access to the lower clivus and premedullary area; second, the supracondylar approach is made through the area above the occipital condyle to the hypoglossal canal and jugular tubercle; and, third, the transcondylar exposure through the lateral area to the occipital condyle is done. It includes drilling of the jugular process in the area lateral to the occipital condyle to access the posterior portion of the jugular foramen and to the mastoid on the lateral side of the jugular foramen (Figure 2D).\textsuperscript{93} A dural incision is made as a J-shaped incision, using a scalpel and continued with Metzenbaum scissors, surrounding the dura from the transverse-sigmoid junction curving medially and inferiorly for posterior crossing of the FM to the intradural entry point of the VA.\textsuperscript{93,94} Cervical dura should be opened linearly and paralinearly downwards to at least the C2 lamina, and the dura should be reflected anteriorly as deeply as possible.

Lastly, microsurgical removal is achieved by using Caviton ultrasonic surgical aspiration, microbipolar coagulation, and microsurgical instruments.\textsuperscript{96} After the tumor is removed and hemostasis is accomplished, closure of the dura is done. When it is not possible, dural substitutes can be used for hemostasis. The occipital bone fragment is replaced and then fixed. The muscles are reattached, and the aponeurosis, subcutaneous tissue, and skin closure are attained.

CONCLUSIONS

FMMs have been a challenge to neurosurgeons for centuries. A perfect knowledge of the surgical anatomy is the most important requisite to safely perform the exposure. Since Andreas Vesalius’s human anatomical dissections to current times,
several techniques have been used for treating FM lesions; however, the FLA has become the safest intervention to solve it. This approach shows less mortality and morbidity rate than other neurosurgical approaches. This microsurgical approach protects vital neurovascular structures, allowing a careful approach to the VA and lower cranial nerves. Having said this, it becomes mandatory in neurosurgical education to distribute tasks in the surgical scenario for the improvement of patient safety, applying state-of-the-art materials and techniques. Future generations of neurosurgeons will benefit by having a more comprehensive view of not only the anatomy of FM based on historical roots of human brain studies but also the advances made to apply safer neuroanatomical approaches such as the far lateral one.

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Conflict of interest statement: The authors of this manuscript report no conflicts of interests concerning this manuscript and have nothing to disclose.

Received 20 May 2017; accepted 6 July 2017


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