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Case Report

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Accessory Nerve Meningioma of the Foramen Magnum: A Rare Neurosurgical Entity

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ABSTRACT

Meningiomas, which are the most common primary intracranial tumors, frequently arise from the dura of the convexities, parasagittal regions, and skull base. The accessory nerve (cranial nerve XI) at the level of the foramen magnum is an exceptionally uncommon site of origin. To date, only a few cases have been clearly documented in the literature. We report the case of a 66-year-old woman who presented with dizziness, headaches, and left-sided shoulder weakness. The lesion was initially presumed to be a regular foramen magnum meningioma. Intraoperatively, the spinal accessory nerve was centrally engulfed by the tumor, with gross invasion of the nerve sheath and no identifiable dissection plane. The final histopathology confirmed the diagnosis of a World Health Organization Grade I meningioma. Accessory nerve meningiomas are exceedingly rare and can closely mimic the more common dural-based foramen magnum tumors.

KEYWORDS: Accessory nerve, Meningioma, Foramen magnum, Cranial nerve, Microsurgical resection

ABBREVIATIONS: MRI: Magnetic resonance imaging, **SSTR2A:** Strong Somatostatin Receptor 2A, **CN:** Cranial nerve

INTRODUCTION

Meningiomas are the most common primary intracranial tumors, accounting for approximately half of all benign central nervous system neoplasms. They arise from the arachnoid cap cells of the meninges and can theoretically occur at any location within the central nervous system (11). While most meningiomas involve the cerebral convexities, parasagittal region, or skull base, some may originate in association with the cranial nerves (CNs) (4). Although optic nerve sheath meningiomas are well-documented and represent approximately 1% of all intracranial meningiomas (2), meningiomas may also originate from other CNs. Reported cases include those involving the trigeminal nerve (CN V) (3) and the oculomotor nerve (CN III) (1), among others, each presenting with distinct clinical features. In contrast, meningiomas arising from or intimately associated with the lower CNs—particularly the accessory nerve (CN XI)—are exceedingly rare.

Extra-axial solid tumors at the level of the foramen magnum are typically diagnosed as meningiomas. Schwannomas are less common but may be considered when there is a single CN deficit and no clear dural tail on imaging. However,

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accessory nerve meningiomas are extremely rare and can easily be overlooked by both radiologists and neurosurgeons. Surgical resection remains the mainstay of treatment for large lesions, but its complex relationship with the accessory nerve poses considerable challenges.

The present report aimed to describe a rare case of a foramen magnum meningioma originating from the accessory nerve, highlighting the diagnostic challenges associated with non-dural-based foramen magnum CNs meningiomas, discussing the surgical approach and nerve preservation strategy, and sharing the histopathological properties and radiological findings of this rare clinical entity.

CASE REPORT

A 66-year-old woman without any relevant medical history was referred to our neurosurgery outpatient clinic due to a 6-month history of dizziness, occipital headaches, and left-sided shoulder pain. Neurological examination revealed mild weakness of the left shoulder upon elevation (Medical Research Council Grade 3/5) and a positive Romberg sign. No other CN deficits or other myelopathic signs were noted.

Brain magnetic resonance imaging (MRI) demonstrated a homogeneously enhancing lesion at the foramen magnum (Figure 1). The lesion was radiologically consistent with a foramen magnum meningioma, although no clear dural tail was noted on imaging. Computed tomography scans showed no hyperostosis or bony involvement.

The patient was offered surgical resection and provided informed consent. She was placed in a lateral position and underwent a left-sided far-lateral craniotomy. Intraoperatively, the tumor was found to encase the accessory nerve (Figure 2). A specimen was sent for frozen section analysis, which revealed that the tumor was most consistent with a meningioma. Notably, no dural attachment of the lesion was observed. Upon careful microsurgical dissection, the tumor was found to invade the nerve sheath of the accessory nerve, and a clear surgical plane could not be identified. Dissection was performed by identifying the intact nerve's proximal and distal ends and carefully navigating through the lesion with the goal of preserving the nerve's anatomical and functional integrity (Figure 2).

Histopathological examination confirmed the diagnosis of a meningioma, with immunohistochemical negativity for S100 and SOX10, thus ruling out the diagnosis of a schwannoma, and strong somatostatin receptor 2A positivity confirming the diagnosis of a meningioma arising from the accessory nerve (Figure 3).

The patient tolerated the procedure well and exhibited no new neurological deficits postoperatively. Follow-up MRI showed complete resection of the lesion, with the 6-month follow-up MRI scan demonstrating no residual or recurrent tumor (Figure 4).

DISCUSSION

Although the foramen magnum is a recognized site for meningioma development, most of these tumors originate from the dura in the lower posterior fossa and foramen magnum. Yasargil et al. have reported that foramen magnum meningiomas originate from the dural entry point of the vertebral artery on the intradural side (12). However, although rare, foramen magnum meningiomas may arise directly from the accessory nerve. The preoperative diagnosis of such lesions can be challenging; yet clinicians should maintain a high index of suspicion for accessory nerve meningiomas when encountering homogeneously enhancing masses that lack a dural tail and lie in close anatomical proximity to the course of the accessory nerve.

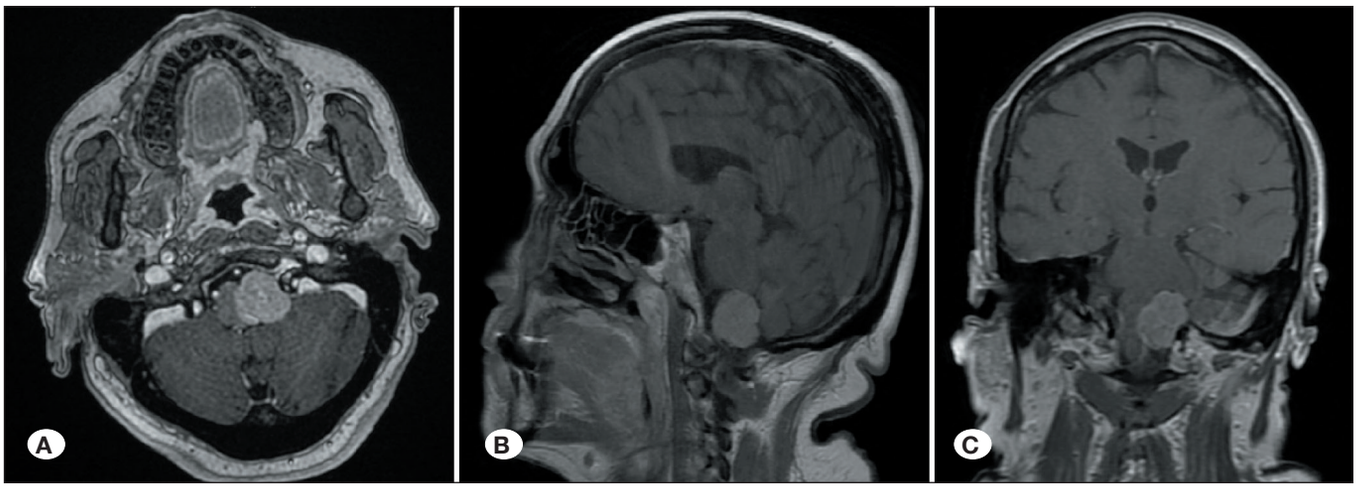

Figure 1: Preoperative contrast enhanced T1W magnetic resonance imaging scans revealed a well-circumscribed, diffusely enhancing left-sided lesion at the level of the foramen magnum (**A:** axial, **B:** sagittal, **C:** coronal sections). The imaging characteristics were suggestive of a meningioma; however, the absence of a clear dural attachment raised the differential diagnosis of other pathologies, including a schwannoma or, less commonly, a meningioma not originating from the dura.

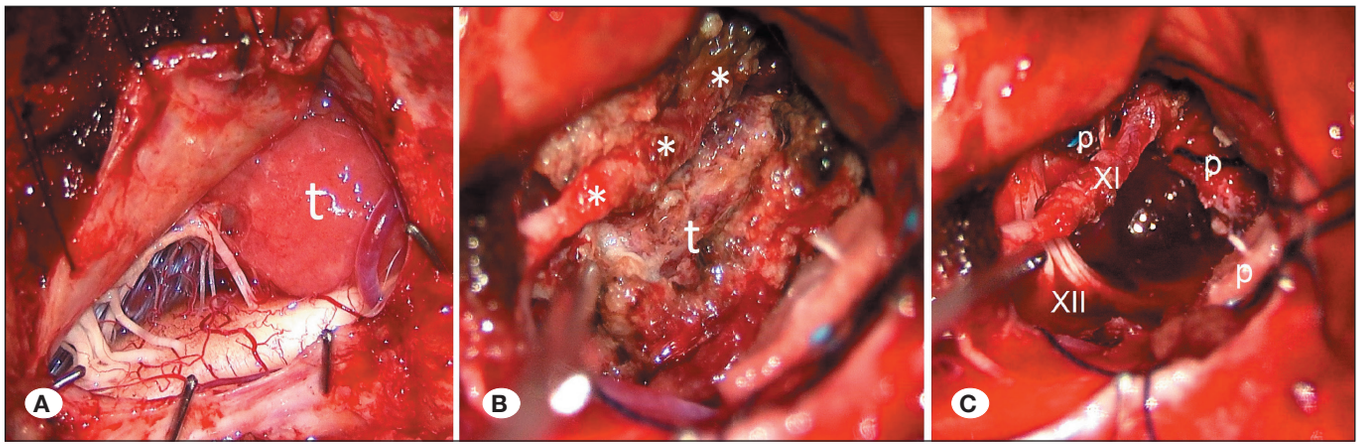

Figure 2: Intraoperative views of accessory nerve meningioma resection via a far-lateral approach. **A)** Intraoperative photograph following surgical exposure via a left-sided far-lateral approach. A well-circumscribed tumor (t) is visualized at the level of the foramen magnum. **B)** Intraoperative view demonstrating the tumor dissection process. The accessory nerve (*) is seen after being dissected from the tumor (t), revealing gross invasion of the nerve sheath and absence of a clear dissection plane. Microsurgical dissection was performed between the proximal and distal nerve segments under high magnification to preserve the nerve's integrity. **C)** Intraoperative view showing the accessory nerve (XI) after complete circumferential dissection and separation of the tumor. The label (p) marks the cottonoid patties within the operative field.

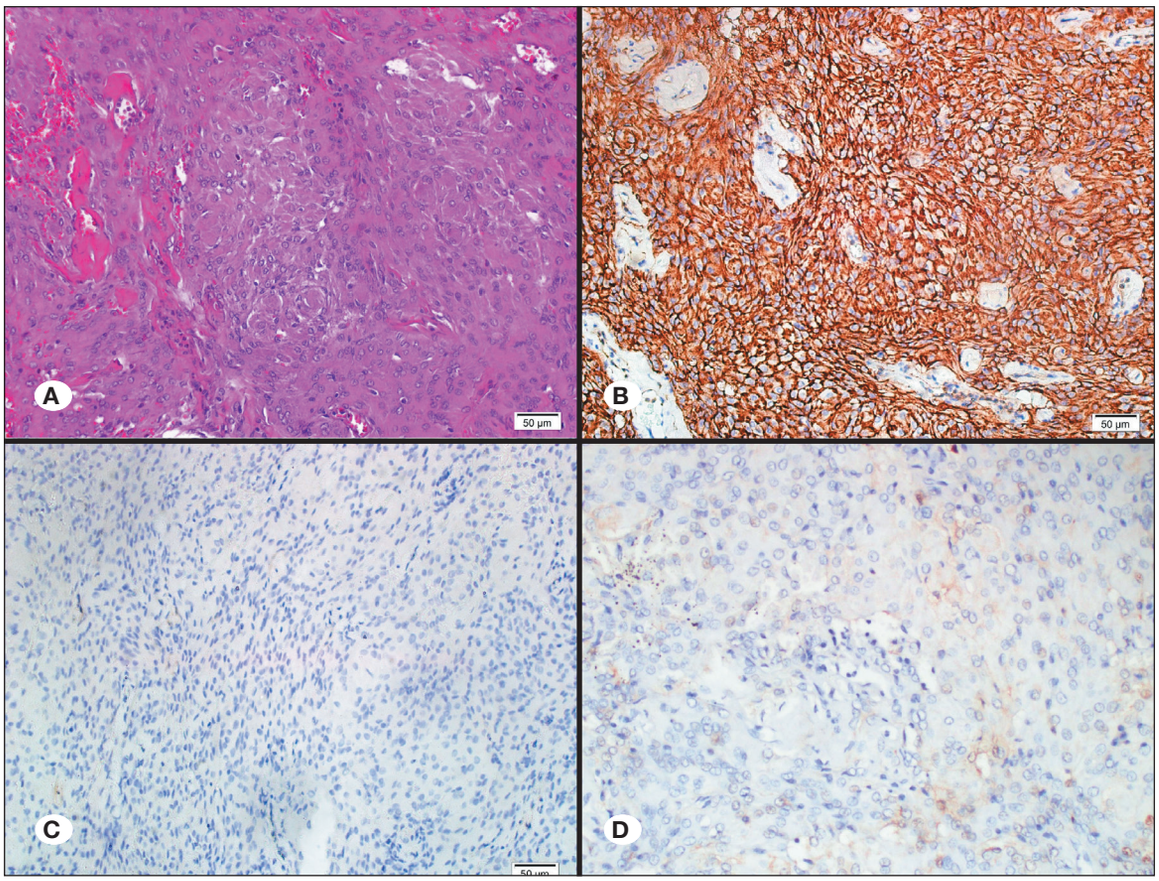

Figure 3: Histopathological and immunohistochemical features of the tumor confirming the diagnosis of meningioma. **A)** Histopathological appearance of the tumor. The lesion comprised cells with abundant eosinophilic cytoplasm, oval nuclei, and a syncytial growth pattern (hematoxylin & eosin staining; magnification ×400). **B)** Tumor cells demonstrate diffuse immunoreactivity for somatostatin receptor 2A (SSTR2A), consistent with a diagnosis of a meningioma (×200). **C)** SOX10 expression was not observed in the tumor cells, supporting the exclusion of a schwannoma or other neural crest-derived neoplasms (×200). **D)** Diffuse strong S100 staining was not observed in the tumor cells (×200).

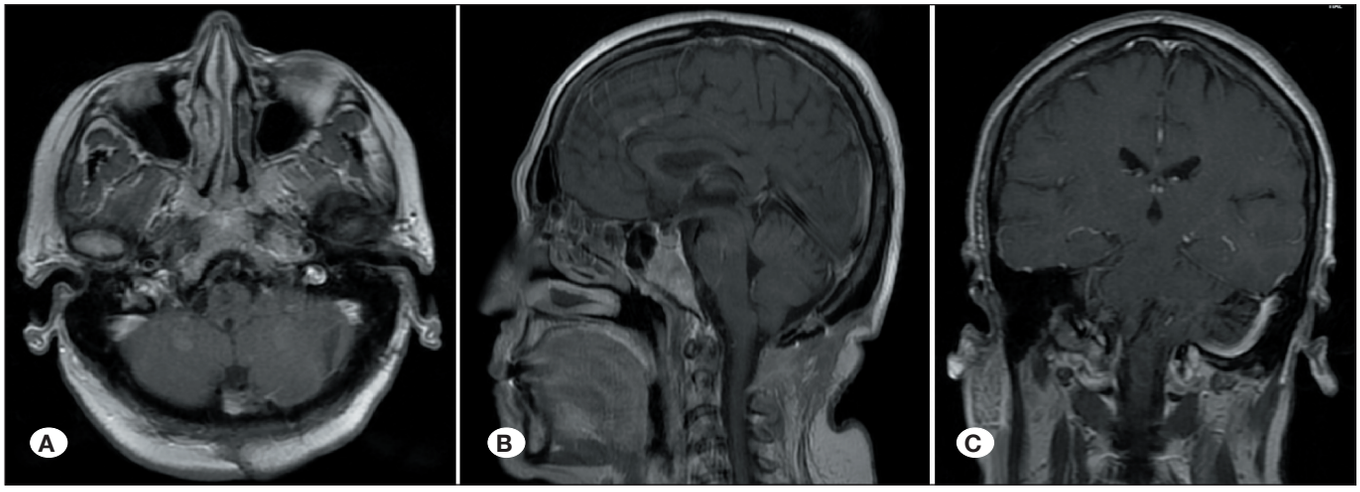

Figure 4: A six-month postoperative magnetic resonance imaging scans demonstrating gross total resection of the lesion, with no evidence of residual or recurrent tumor (**A:** axial, **B:** sagittal, **C:** coronal sections).

Apart from optic nerve sheath meningiomas—which are uniquely encased by true meningeal layers—accessory nerve meningiomas are exceedingly rare and, by definition, lack a dural attachment. The origin of these tumors may involve ectopic arachnoid cap cells along the perineural arachnoid extensions or displaced meningeal elements in the CN sheath.

Unlike most foramen magnum meningiomas, which displace or encase nearby nerves, direct infiltration of the nerve sheath considerably complicates surgical resection. In such cases, the absence of a dissection plane between the tumor and nerve makes microsurgical dissection a difficult and tedious process. A practical approach, as in our case, involves identifying the nerve's proximal and distal ends, followed by intratumoral dissection to preserve the nerve's continuity. In our experience, intraoperative neurophysiological monitoring was valuable in guiding the resection process and avoiding irreversible damage. Ultimately, in these rare cases, a meticulous microsurgical technique remains critical for accessory nerve preservation.

The present case report emphasizes the importance of understanding the normal anatomy, particularly in certain locations such as the foramen magnum region where critical neurovascular structures are densely clustered and lie in close proximity to each other. The accessory nerve is purely a motor nerve that innervates the sternocleidomastoid and trapezius muscles. In foramen magnum lesions, the absence of a dural tail, in addition to their homogeneous enhancement, anatomical proximity to the course of the spinal accessory nerve, and the presence of accessory nerve paresis can help raise suspicion for an accessory nerve meningioma. However, it is still difficult to include accessory nerve meningioma in the differential diagnosis because only a few cases have been reported in the literature. When surgically treating these lesions, comprehensive knowledge of the anatomy and microsurgical principles as well as careful neuromonitoring are of utmost importance to preserve the nerve. When the neurosurgeon is familiar with the anatomical course of the CNs, particularly the accessory nerve, as in the present case, it is possible to identify that the nerve ends within the tumor and originates from the site of engulfment. Neuromonitoring, including nerve stimulation, can easily confirm the nerve and facilitate the surgical course. Another unusual aspect for the neurosurgeon is that this meningioma is not attached to the dura at any point. In the present case, the lesion seemed to be floating, freely movable, and to be easily pulled out; however, this situation can be very deceiving, as forcing the lesion out may lead to the avulsion or transection of the accessory nerve.

To the best of our knowledge, only six cases of accessory nerve meningiomas have been reported in the literature (Table I). Among these, only three cases were foramen magnum meningiomas (5,7,9), whereas the remaining cases were upper cervical lesions. Kobyakov et al. have described a cystic tumor with a solid component arising from the nerve sheath, whereas Mohri et al. reported a solid lesion eccentrically adherent to the accessory nerve with a favorable dissection plane (5,7). Contrarily, our case involved the spinal accessory nerve being centrally engulfed by the tumor, with clear evidence of gross nerve sheath invasion and no identifiable dissection plane (Figure 2). This reinforces the uniqueness of our case, both anatomically and surgically, contributing to the limited literature on this rare entity.

Accessory nerve meningiomas are exceedingly rare and can radiologically mimic common dural-based foramen magnum meningiomas. The present case highlights the importance of including the accessory nerve meningiomas in the differential diagnosis of foramen magnum lesions, particularly when a dural tail cannot be identified on the MRI scan. Intraoperatively, the absence of a favorable dissection plane and gross nerve sheath invasion—as seen in our case—can pose considerable surgical challenges. Nevertheless, functional preservation may still be achievable through meticulous microsurgical dissection performed under high magnification and illumination, guided by anatomical knowledge and intraoperative neurophysiological monitoring. The present case contributes to the limited

Table I: Reported Cases of Foramen Magnum Meningiomas in the Literature with Key Clinical and Surgical Characteristics

Case	Author(s)	Year	Age/ Sex	Presentation	Preoperative Examination	Location and side	Histology	Surgical Approach	Outcome
1	Thomé et al. (9)	2003	61/F	Ataxia, left hemiparesis, myelopathy	Minimal left-sided motor impairment (grade 4–5/5), positive Babinski's reflex, positive Romberg test, slightly decreased proprioception, incoordination and dysmetria on the left side	Bilateral, foramen magnum	S-100 positive, EMA positive, vimentin positive	Midline suboccipital + C1 laminectomy	No recurrence at the 3-year follow-up (presentation was likely secondary to neurodegenerative disease)
2	Tatagiba et al. (8)	2005	35/M	Right shoulder pain and accessory nerve paralysis	Complete accessory nerve palsy on the right side	Right jugular foramen	Meningothelial, invasion of the epineural space,	cervical-transmastoid paracondylar approach to the jugular foramen.	Postoperative hoarseness and dysphagia later improved.
3	Liechty et al. (6)	2007	9/M	Closely followed up for multiple meningiomas/ schwannomas	Right eye blindness, nystagmus on the left side	Right spinal accessory nerve at the C1 level	Psammomatous, neurofibromatosis II	C1 laminectomy	No recurrence at 3 years
4	Mohri et al (7)	2019	69/F	Dizziness and bilateral shoulder pain	Spinal accessory nerve palsy (difficult in raising the shoulder, deficit of 3/5) on the left side	Left spinal accessory nerve at the foramen magnum	Meningothelial meningioma, positive for EMA	Midline suboccipital with C1 laminectomy	Improvement in shoulder strength
5	Ueno et al. (10)	2022	57/F	Headache	No deficit	Right spinal accessory nerve at C1–C2	Atypical meningioma, positive staining for epithelial membrane antigen and progesterone receptors	C1 laminectomy with restricted posterior foramen magnum osteotomy	Neurologically intact postoperatively
6	Kobyakov et al. (5)	2024	53/M	Occipital headache	No focal deficit	Left spinal accessory nerve at the foramen magnum	Angiomatous meningioma, progesterone receptor (PR)-positive, positive epithelial membrane antigen EMA	midline suboccipital approach	Uneventful recovery
7	**Present Case**	2025	66/F	Dizziness, headache	Mild weakness of left shoulder elevation (3/5), positive Romberg sign	Left spinal accessory nerve at the foramen magnum	Meningothelial meningioma, SOX-negative, S-100 negative, gross invasion of the accessory nerve sheath	Left-sided far-lateral approach	Left trapezius/SCM weakness 3/5, later improved

literature on accessory nerve meningiomas and underscores the need for a high preoperative index of suspicion and surgical adaptability in managing atypical skull base tumors.

CONCLUSION

Accessory nerve meningiomas are exceedingly rare and can radiologically mimic regular foramen magnum meningiomas. Although there is no clear cleavage plan between the nerve and tumor, functional preservation may still be achievable through meticulous microsurgical dissection, guided by anatomical knowledge and neurophysiological monitoring.

Declarations

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Availability of data and materials: The datasets generated and/or analyzed during the current study are available from the corresponding author by reasonable request.

Disclosure: The authors declare no competing interests.

Informed consent: The patient's written informed consent was acquired.

AUTHORSHIP CONTRIBUTION

Study conception and design: MSG, MA, MAK

Data collection: MAK, SS, Gİ

Analysis and interpretation of results: MSG, MA, TSA

Draft manuscript preparation: MA, MAK

Critical revision of the article: MSG

Other (study supervision, fundings, materials, etc...): BB, MSG

All authors (MA, MAK, SS, GI, BB, TSA, MSG) reviewed the results and approved the final version of the manuscript.

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