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Insidious Presentation of a Foramen Magnum Meningioma – A Case Report

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Abstract

Foramen magnum meningiomas are rare intracranial tumors that commonly present with lower cranial nerve deficits and motor/sensory disturbances of the extremities. Their signs and symptoms can be attributed to their close relation to the brainstem, particularly at the non-distendable area of the foramen magnum (the foramen at the skull base that connects brain and spinal cord). Diagnosis is usually delayed due to the rarity of the disease and its often not so pronounced symptoms. We present a case of a foramen magnum meningioma with an insidious onset that was treated in our department.

Keywords: Foramen magnum; Meningioma; Surgical treatment

Introduction

Meningiomas are common primary neoplasms arising from meningothelial (arachnoidal) cells. They comprise about 25% of all primary tumors of the central nervous system (CNS) and 15-20% of all intracranial tumors [1,2]. Despite their rather high incidence, only 2.5% (range 1.8-3.2%) arise at the foramen magnum level, but still meningiomas are the most commonly observed tumors of the foramen magnum, accounting for 70% of cases [3-5]. They may develop intradurally (94.4%), extradurally (2.8%) or both intra-extradurally (2.8%)[6].

Clinical presentation usually includes lower cranial nerve palsies and long tract signs and symptoms due to medulla oblongata compression. Due to the rarity of the disease and its often insidious onset, patients are often misdiagnosed or develop severe neurological deficits until proper diagnosis and treatment can be established. We present the case of a 55-year-old female with an anterior foramen magnum meningioma who was treated in our department after a long work-up from other disciplines.

Case Report

A 55-year-old female with a free medical history presented to our department complaining of nearly nine months onset neck pain. She had previously visited an orthopedic surgeon and underwent a cervical spine CT scan which revealed a small C5-C6 disc herniation but her neck pain did not respond to the prescribed analgesics neither to physiotherapy. On admission to our department she had no extremity weakness or sensory disturbances (other than the neck pain, especially during neck movements but without any movement limitation). After a careful and detailed medical history and physical examination she also reported recent (a few weeks') onset of mild swallowing disturbances (dysphagia) and subtle left cheek and tongue numbness. She underwent a cerebral and cervical magnetic resonance imaging (MRI) scan which revealed a mass lesion (isointense on T1, hyperintense on T2 and hypointense on diffusion-weighted sequences, with homogenous contrast enhancement, distinct borders and a characteristic dural tail) at the anterior margin of the foramen magnum which compressed medulla oblongata and displaced it posteriorly (figures 1 and 2). The maximum dimensions of the lesion were 21mm (anteroposterior), 28mm (transverse) and

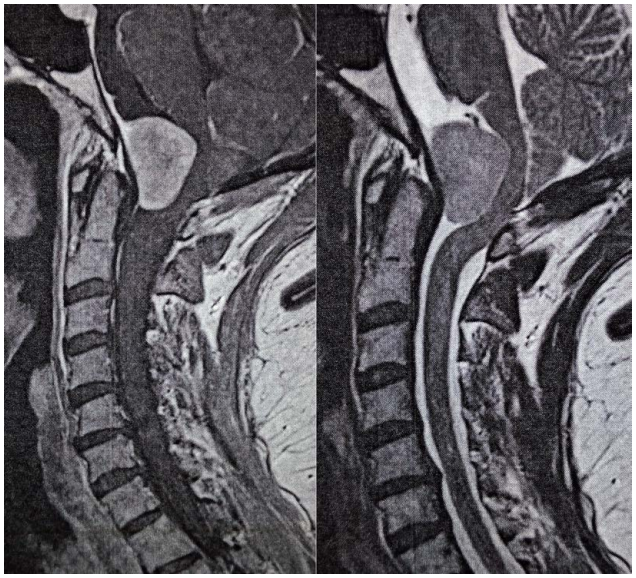

Figure 1: Preoperative sagittal contrast-enhanced T1WI and T2WI MRI scan

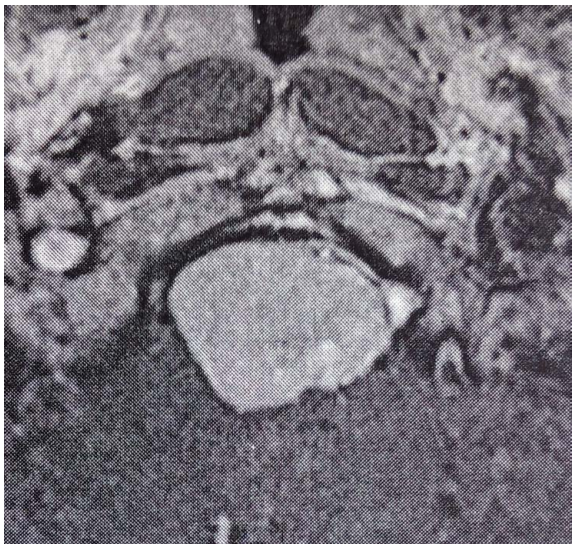

Figure 2: Preoperative axial contrast-enhanced T1WI MRI scan

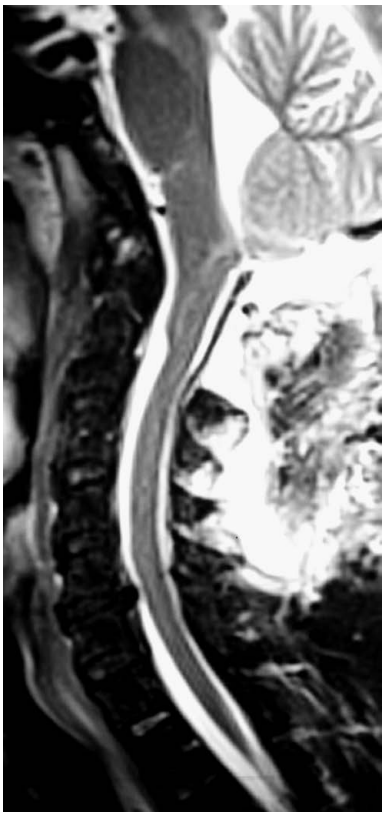

Figure 3: Post-operative sagittal contrast-enhanced T2WI MRI scan

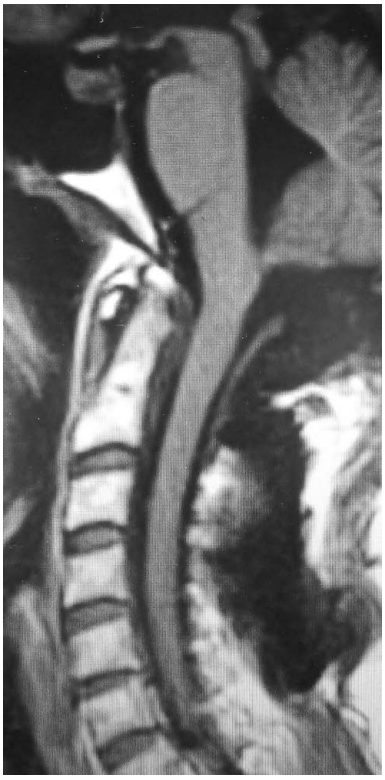

Figure 4: Post-operative sagittal T1WI MRI scan

28mm (vertical). The dimensions of the foramen magnum were 35mm (anteroposterior) by 28mm (transverse). A preoperative digital subtraction angiography showed that the tumor was supplied by the left ascending pharyngeal artery.

The patient was operated on at the prone position and a suboccipital craniectomy and C1 laminectomy were performed. Through a posterolateral approach a complete piecemeal removal of the tumor was achieved without any intraoperative events (figures 3 and 4). Her postoperative recovery was uneventful, with the exception of hiccups which persisted for a few days. Pathology report of the surgical specimen was "Grade I meningioma".

Discussion

Foramen magnum meningiomas are undoubtedly challenging tumors in regard to both their diagnosis and treatment. Due to their close relation to the brainstem (and particularly the medulla oblongata), the lower cranial nerves and the vertebral artery and branches, perfect knowledge of the surgical anatomy is imperative when treating such cases [7]. Preservation and minimal (if possible not at all) retraction of lower cranial nerves (glossopharyngeal, vagus, spinal accessory and hypoglossal) during surgical excision of foramen magnum tumors is crucial for keeping surgical morbidity low. Gamma knife radiosurgery could be used in cases of small meningiomas of the foramen magnum, resulting in tumor control and preservation of neurologic function, but only when there is no mass effect on presentation.

When foramen magnum meningiomas are finally diagnosed they are usually rather large because of their slow-growing rate, the wide subarachnoid space at this level and their insidious clinical presentation. Therefore, there is often a long interval of many months between onset of symptoms and diagnosis [5,8]. Differential diagnosis includes multiple sclerosis, amyotrophic lateral sclerosis, syringomyelia and cervical spondylosis [9,10].

In a not so careful physical examination early signs and symptoms of a foramen magnum meningioma might be missed and the progression of the disease can lead to severe and possibly permanent neurological deficits. Early symptoms include occipital and upper cervical pain, often exacerbated by neck movement, due to compression of the upper cervical nerves that innervate the infratentorial dura mater. Progression of the disease is characterised by development of unilateral arm motor and sensory deficits, which progress to the ipsilateral leg, then the contralateral leg and finally the contralateral arm. Later findings include spastic quadriparesis and lower cranial nerve palsies. Terminal progression includes quadriplegia, inability to maintain airway protection with secondary pneumonitis, and finally respiratory arrest [5].

Conclusion

Foramen magnum meningiomas are rare intracranial tumors characterised by insidious onset of symptoms. Due to their potential progression to severe and irreversible neurological deficits, physicians should be aware of its clinical presentation and suspect it when encountering patients with corresponding symptoms. It cannot be overemphasized that perfect knowledge of the surgical anatomy is a prerequisite for treating foramen magnum meningiomas.

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