Case report

Primary jugular foramen meningioma with unusual extensive bone destruction: case report and review of literature

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Jugular foramen meningiomas are exceedingly rare tumors of the fossa jugularis. These meningiomas are characterized by an invasive growth pattern with extensive skull base infiltration in all directions and by the mixed permeative-sclerotic appearance of the bone margins of the jugular foramen. We report an unusual case of a primary jugular foramen meningioma in a 30-year-old woman. The unenhanced high-resolution CT of the temporal bones revealed extensive bone destruction around the left jugular foramen as well as bone destruction of the basilar part of the left occipital bone without sclerosis. These findings are unusual for meningiomas and correspond more to glomus jugulare tumors. In the literature, we did not find a case similar to ours. Conclusion. MRI and CT imaging provide accurate distinction between meningioma and glomus tumors or schwannomas in most cases. From high-resolution CT scans, in the case where a permeative-destructive pattern is dominant, and with the absence of hyperostosis and bone thickening around the jugular foramen, the differential diagnosis between jugular foramen meningiomas and other tumors, especially glomus jugulare tumors, is difficult. In that case the correct diagnosis should be based on the MRI findings.

Key words: Jugular foramen, Meningioma, Glomus jugulare tumor, Bone destruction.

Introduction

The most common tumor that develops in the jugular foramen (JF) is a glomus jugulare tumor. Neural sheath tumors follow, such as schwannomas and neurofibromas, which share a common site of development in the JF, while meningiomas are the most rare tumors (1). Jugular foramen meningiomas (JFM) can be classified as primary if the tumor originates from the JF, although these are exceedingly rare, or secondary, when the tumor is centered in the posterior fossa, most commonly in the cerebellopontine angle or petroclival region, with extension into the JF (1, 2). Primary JFM appear

to behave differently from meningiomas that involve JF secondarily (3, 4, 5). These meningiomas are characterized by an invasive growth pattern with extensive skull base infiltration in all directions (3, 6, 7), and by the mixed permeative-sclerotic appearance of the bone margins of the JF (2, 8). In this article, we describe a very unusual case of primary JFM with permeative erosion of JF margins and extensive destruction of affected bones without sclerosis, which is not typical for JFM. We also review the literature on this rare entity.

Case report

Three years ago, a 30-year-old woman with a one year history of progressive left hearing loss, pain in the left half of the face, headache and dizziness, came to the Department of Radiology, and computed tomography (CT) of the temporal bones was performed. CT detected a widened left JF with erosions of its margins without sclerosis and soft tissue mass into the mesotympanum and hypotympanum. The ossicles were intact. The ostheolitic changes were seen on the posterior and inferior wall of the petrous portion of the temporal bone. The radiologist suspected a glomus jugular tumor. Digital subtraction angiography (DSA) revealed the vascular tumor with dual feeding arterial supplies, from the left external carotid artery and the left vertebral artery (posterior inferior cerebellar artery), without intensive tumor blush. Without preoperative magnetic resonance imaging (MRI) the patient underwent surgery at another hospital. Left suboccipital craniectomy via retrosigmoid approach was performed and subtotal tumor removal was achieved. Histopathological examination revealed meningothelial meningioma. In the next few months, she underwent gamma knife radiosurgery twice at another hospital. After surgery, the previous symptoms persisted, with swallowing

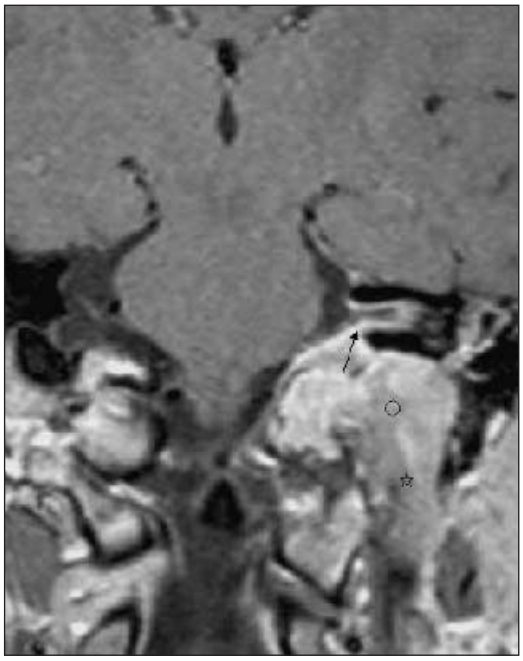

Figure 1 The coronal contrast-enhanced T1-weighted image shows intense enhancement in the mass centered in the jugular foramen (circle) with extensive infiltration of surrounding skull base. The mass spreads into the posterior fossa, the internal auditory canal (arrow) and inferiorly into the nasopharyngeal carotid space (star). High-velocity flow voids in the mass are absence. Also, the MR image shows different signal intensities between the intra- and extracranial components of JFM

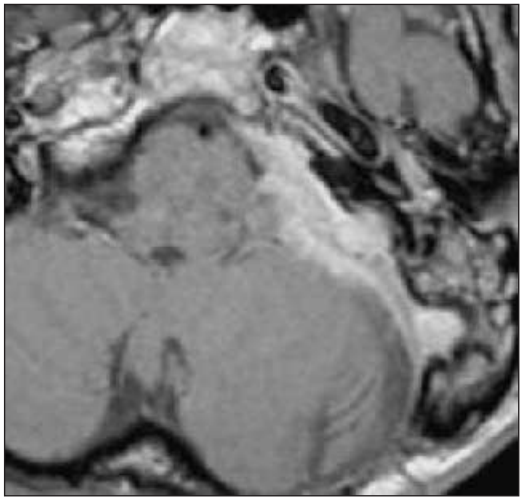

Figure 2 The axial contrast-enhanced T1-weighted image shows en plaque involvement of the posterior fossa with prominent dural tails and medial spread in the skull base to the mid clivus

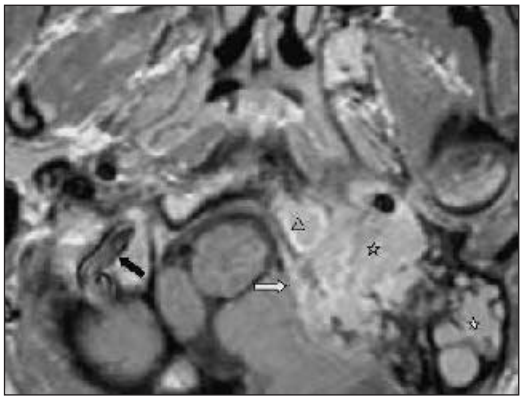

Figure 3 The axial contrast-enhanced T1-weighted image shows the mass in the left fossa jugularis and the carotid space (star) with medial spread into skull base to the mid clivus (triangle), and en plaque involvement of the posterior fossa (white arrow). The left internal jugular vein is incorporated into the mass with the absence of its normal flow void. Note normal flow voids on the right (black arrow). The mastoid air cells are filled with fluid (white star)

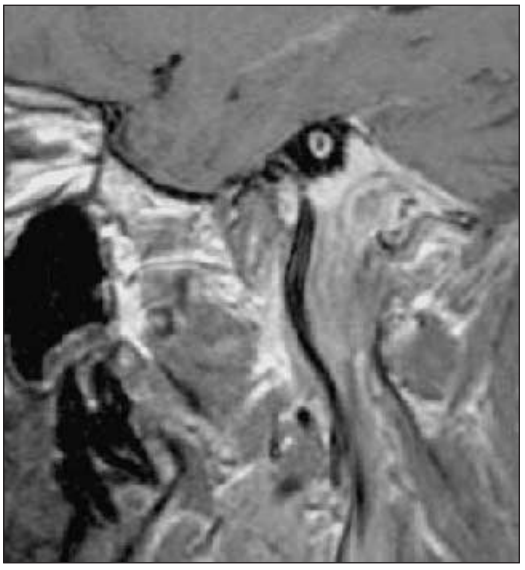

Figure 4 The sagittal contrast-enhanced T1-weighted image shows the primary jugular foramen meningioma with inferior involvement of the nasopharyngeal carotid space and anterior displacement of the carotid artery

dysfunction and dysphonia in addition. Ten months after surgery, postoperative MRI of the brain was performed. The MRI findings showed an extra-axial mass isointense to hypointense on T1-weighted sequences and intermediate on T2-weighted sequences, with strong enhancement after contrast administration. The mass was centered in the left JF with extensive en plaque involvement of the posterior fossa and prominent dural tails, which spread into the internal auditory canal (Figure 1, 2). The mass medially involved the clivus, while inferiorly it involved the carotid space (Figure 2, 3, 4). The MR signal intensity of the intracranial component for JFM was higher than of the extracranial component (Figure 1, 4).

One year after MRI, a control CT showed widening of the left JF with permeative erosion of its margins without sclerosis and extensive bone destruction around the left JF, as well as bone destruction of the basilar part of the left occipital bone, including the left occipital condil, the clivus and the lateral wall of the foramen magnum (Figure 5, 6). The walls of the carotide canal, the vestibular aqueduct, as well as the internal auditory canal were eroded (Figure 7). At this time the tympanic cavity was completely opacified while the ossicles were intact as they were on the first CT scans (Figure 5).

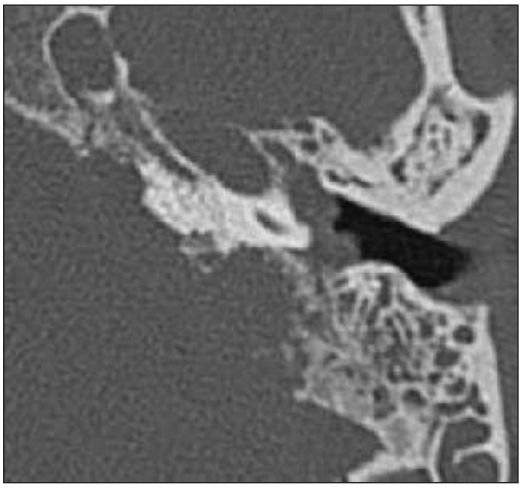

Figure 5 The bone window image from the axial CT scan shows widening of the left jugular foramen with permeative erosion of its margins without sclerosis. The image shows a soft tissue mass in the cavum tympani, as well as fluid in the mastoid air cells on the left

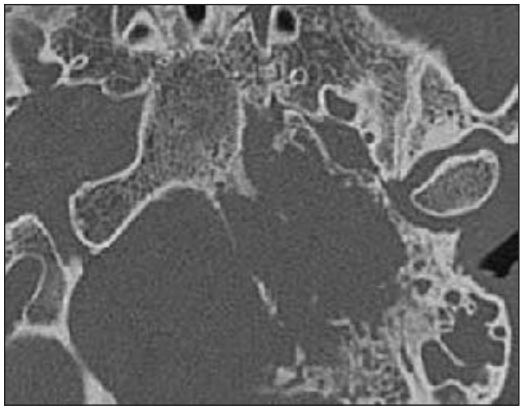

Figure 6 The bone window image from the axial CT scan shows extensive bone destruction of the basilar part of the left occipital bone

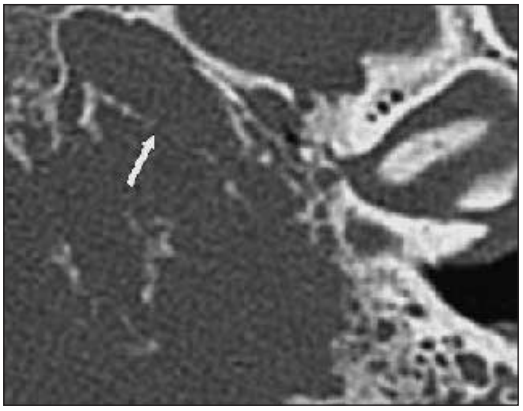

Figure 7 The bone window image from the axial CT scan shows the eroded wall of the left carotid canal (arrow)

Discussion

The jugular fossa is an anatomically complex region which consists of major vessels, the jugular bulb and vein, and multiple cranial nerves, including the cranial nerve IX, X, and XI (1). Due to this fact, patients with jugular fossa tumors (JFT) may have multiple cranial nerve dysfunctions. JFT are rare in clinical practice. JFM are most often considered in the differential diagnosis of glomus jugulare tumors (GJT), which account for 90% of JFT. They are followed by schwannomas of the lower cranial nerves, and then JFM, which account for about 0.7–9.3% of posterior fossa meningiomas (9, 10). Approximately, 112 cases of JFM have been reported in literature (8) and 40 of them are primary JFM (3). Samii and Ammirati in their series of 420 skull base meningiomas found only three primary JFM (0.7%) (8). In many of the reported cases, JFM mimicked GJT and lower cranial nerve neuromas (11). This is very important because different JFT have different surgical risks, and preoperative differential diagnosis is important for surgical planning and evaluation of postoperative morbidity (12). The identity of most lesions can be determined by a combination of spiral CT and MRI (2, 13). CT is useful for analysis of the JF bone margins as well as of adjacent skull base foramina. MRI with gadolinium shows the characteristics of a tumor, its vascularization, extension and its relationship to neighboring structures (1). Primary JFM often present very invasive features, infiltrating the surrounding skull base in all directions (3, 5, 6, 7, 8). This pattern of spread can be referred to as "centrifugal" and usually involves the temporal bone, including the middle ear cavity laterally and invading the skull base, including the jugular tubercle, hypoglossal canal, occipital condyle, and clivus medially. Inferior extracranial spread occurs into the nasopharyngeal carotid space of the deep suprahyoid neck. Further superior intracranial spread is seen along the intracranial dural reflections. This spread along the dura is termed "en plaque" and is characteristic of primary JFM. A globose appearance is seen less commonly in tumors with intracranial extension (2, 8). Theoretically, the pattern of spread allows differentiation between primary meningiomas, and the other fossa jugularis tumors (8). GJT typically involve the hypotympanum superolaterally, with limited involvement of the carotid space inferiorly. Infrequently, they extend medially into the jugular tubercle, hypoglossal canal,

and clivus (2). Unlike paragangliomas, JF schwannomas follow the course of the IX, X, and XI cranial nerves from the brainstem, with variable inferior spread. However, the pattern of spread is not totally reliable and can not form a basis on which to make a distinction between these entities (8). A helpful differentiating feature of JFM is the absence of the flow voids which are characteristic of GJT (2, 9). Macdonald et al. reported five cases of primary JFM. All of them were characterised by centrifugal infiltration of the surrounding skull base. Posterior fossa involvement had an "en plaque" appearance in four cases. All cases showed prominent dural tails. Flow voids were absent in all cases (2). We found all these features in our case.

On DSA, paragangliomas unlike meningiomas have typical angiographic appearance- a hypervascular mass, with enlarged feeding arteries, intense tumor blush, and early draining veins (13). In our case DSA revealed a vascular tumor without intense tumor blush what excluded GJT from the differential diagnosis of JFTs.

A recent study by Shimono et al. demonstrated differences in MR signal intensity and contrast enhancement between the intra- and extracranial components of JF meningiomas. The signal intensities of the intracranial component of JFM were significantly higher than those of the extracranial component on T1-, T2-, and postcontrast T1-weighted images (14). We also noted these different signals that are the best visualised on the postcontrast T1-weighted images (Figure 1). Primary JFM cause irregular enlargement of the JF. On CT scans, the JF margins have a mixed permeative-sclerotic appearance. On the other hand, GJT causes a permeative-destructive pattern, with erosion of the JF margins and infiltrated bone, without preservation of the underlying architecture or bone density. Neuroma gradually enlarges the JF by pressure erosion and gives an expanded and scalloped, but well-defined corticated margin to the JF (2, 9). In our case primary JFM caused permeative erosion of the JF margins and extensive bone destruction of affected bones (the petrous part of the left temporal bone and the basilar part of the left occipital bone) without any sclerosis. These findings are unusual for meningiomas and correspond more to GJT. Chen et al. reported a case of an angiomatous type of JFM with bony destruction around the JF without sclerotic change, which may be related to the histologic type of this tumor (12). In our case the etiology is unknown. It is not related to the histologic type of the tumor because it was a meningothelial meningioma, which is the most common tumor of the fossa jugularis (6).

Conclusion

Correct preoperative differential diagnosis of JF tumors is important for surgical planning and helps to avoid surgical pitfalls. MR and CT imaging provide an accurate distinction between meningioma and glomus tumor or schwannoma in most cases. High-resolution bone window CT is helpful for diagnosis, but in the case of the absence of hyperostosis and bone thickening around the jugular foramen, and when a permeative-destructive pattern is dominant, differential diagnosis between JFM and other tumors, especially GJT, is difficult by high-resolution CT. In that case the correct diagnosis should be based on MRI finding.

Conflict of interest: The authors declare that they have no conflict of interest. This study was not sponsored by any external organisation.

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