ANGIOMATOUS TYPE OF JUGULAR FORAMEN MENINGIOMA WITH NECK EXTENSION: DIFFERENTIAL DIAGNOSIS FROM PARAGANGLIOMA AND SCHWANNOMA

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Abstract: Background. Meningiomas involving the jugular foramen and parapharyngeal space are extremely rare. They most commonly occur intracranially and then extend to the extracranial region through the foramen of the skull base, such as jugular foramen. Clinically, these tumors mimic the more common glomus jugulare tumor. Preoperative diagnosis can be correct on the basis of the characteristic imaging findings.

Methods. A 52-year-old woman was seen with a left neck mass and mixed-type hearing loss. She underwent physical examination, MRI, high-resolution CT, and angiography.

Results. Physical examination revealed a retrotympanic, pulsatile red mass in the left ear, and mild bulging of the left oropharyngeal wall. The patient was found to have the spreading, carpet-like, meningioma with extracranial extensions via jugular foramen to parapharyngeal space. Preoperative imaging strategy allowing accurate preoperative diagnosis is discussed.

Conclusions. Accurate distinction between meningioma and glomus tumor or schwannoma is possible in most cases, with attention to fine radiologic detail. ©2007 Wiley Periodicals, Inc. Head Neck 29: 793–798, 2007

Keywords: jugular foramen; neck mass; paraganglioma; meningioma; schwannoma

Meningiomas, which arise from the lining cells of the arachnoid villi, are often located in the dural venous sinuses and their large tributary veins. They are classified into 3 World Health Organization (WHO) grades. Of them, the angiomatous type is rare, accounting for about 2% of all meningiomas. The jugular foramen, which contains the internal jugular vein, can be affected by meningiomas, but is more commonly affected by paragangliomas and schwannomas. Preoperative differential diagnosis cannot be made on the basis of clinical presentation, because these various jugular foramen tumors have similar symptoms and signs. But accurate preoperative diagnosis is still feasible, because these tumors have distinguishable imaging findings. Here, we report a case of angiomatous type of jugular foramen meningioma with extension to the neck and discuss the imaging findings of MRI, CT,
and angiography, which allow accurate preoperative diagnosis.

CASE REPORT

A 52-year-old woman was seen with complaints of progressive left hearing loss for 1 year and a left neck mass for 1 month. Preoperative pure tone audiogram (Figure 1A) revealed mixed-type hearing loss of 88 dB deficiency with air-bone gap of 40 dB. Postoperative audiogram shows left-side severe sensorineural hearing loss of more than 95 dB.

FIGURE 1. (A) Preoperative pure tone audiogram shows left-side mixed-type hearing loss of 88 dB deficiency with air-bone gap of 40 dB. (B) Postoperative audiogram shows left-side severe sensorineural hearing loss of more than 95 dB.

T1-weighted, gadolinium-enhanced MRI demonstrated a uniformly enhanced tumor in the left jugular foramen, extending upward to the cerebellopontine angle and downward to the parapharyngeal space of the neck with encasement of the bifurcation of the common carotid artery (Figures 2A and 2B). High-resolution CT revealed bony destruction around the left jugular foramen with fluid accumulation in left mastoid air cells (Figures 3A and 3B).

FIGURE 2. (A) T1-weighted, gadolinium-enhanced MR image, axial view, shows left cerebellopontine angle tumor with neck extension via jugular foramen. The mastoid air cells were filled with fluid. (B) Axial view of T1WI, gadolinium-enhanced MR image shows parapharyngeal space involvement with obvious salt and pepper sign (arrow). The common carotid artery was also encased. Differentiation between paraganglioma and meningioma is not possible in this image.
Angiography demonstrated a hypervascular tumor in the left jugular fossa with downward extension to the neck. The supplying vessels were mainly from the left ascending pharyngeal artery and partly from left distal vertebral artery. The venous phase of the angiogram demonstrated the left transverse sinus and prolonged tumor stain but did not show the left internal jugular vein and the left sigmoid sinus, which was totally compressed by the tumor (Figure 4). Transartery embolization (TAE) was performed with 150 to 250 μm Contour, which was injected into the left ascending pharyngeal artery. Post-TAE angiogram showed more than 70% decrease of tumor stain (Figures 5A and 5B).

The preoperative diagnosis based on imaging and angiography characteristics revealed meningioma. Far lateral approach of craniotomy extended to the neck was done. A reddish, soft-elastic, and hypervascular tumor arising from the dura at skull base between the left internal acoustic meatus and left jugular foramen was noted with involvement of the lower cranial nerves VII, VIII, IX, X, and XI and compression of the cerebellum and the sigmoid sinus. The tumor was carefully removed away from the lower cranial nerves and sigmoid sinus. The left venous flow from the transverse sinus to the internal jugular vein was well preserved and reestablished.

Histologic diagnosis revealed an angiomatous type of meningioma (Figure 6). After surgery, a nasogastric tube was used to prophylactically prevent aspiration and provide adequate nutrition during the first 2-week operative period. Thereafter, the patient was able to eat per oral route and swallow well without any kind of gastric tube, despite that a left-side vocal paresis was noted postoperatively. Six months later, the patient's hoarseness was much improved, and the laryngoscopy showed that the movement of the left vocal fold had returned to normal. Except for cranial nerve VIII, whose function did not recover (Figure 1B) despite the nerve integrity being maintained during surgery, cranial nerves VII, IX, XI, and XII were all well preserved with good function. To date, the patient is well in the follow-up period of 2 years.

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DISCUSSION

Meningiomas account for 15% to 18% of all primary intracranial tumors, and most of them affect the parasagittal, falx, convexity, and sphenoid wing, the site where cerebrospinal fluid is returned to the venous system. But meningiomas rarely occur in the jugular foramen. Because the jugular foramen consists of greater vessels and multiple cranial nerves, including the cranial nerve IX, X, and XI, patients with jugular foramen meningiomas may be seen with multiple cranial nerve dysfunction such as jugular foramen syndrome (Vernet's syndrome), which is characterized by loss of taste in the posterior third of the tongue (cranial nerve IX), vocal cord paralysis and dysphagia (cranial nerve X), and weakness of the sternocleidomastoid and trapezius muscles (cranial nerve XI). Larger tumors may cause other nonspecific symptoms (eg, headache, increased intracranial pressure) and specific symptoms due to involvement of the auditory pathway and compression of the cerebellum and the brain stem (eg, tinnitus, hearing loss, nystagmus, ataxia, disequilibrium, long tract signs).

Different jugular foramen tumors have different surgical risks, and therefore, preoperative differential diagnosis is important for surgical planning and evaluation of postoperative morbidity. But it is difficult to distinguish between meningiomas and other more common tumors in the jugular foramen, such as paragangliomas and schwannomas, by clinical presentations alone, because they have similar symptoms. Moreover, the pathologic diagnosis of tumors in the jugular foramen cannot be ascertained from preoperative histologic examination because it is difficult and dangerous to obtain tissue biopsy in this region. Fortunately, image studies are useful for determining the most likely diagnosis preoperatively, because these jugular foramen tumors have different tumor vasculature and cause different changes of the adjacent structure that images can demonstrate.

Among the imaging modalities, MRI allows a more comprehensive analysis of the nature and the extent of the tumor, the changes of the adjacent soft tissue, and tumor vasculature using magnetic resonance angiography technique. In our case, MRI revealed that the tumor had intracranial and extracranial parts, which are connected to each other through the jugular foramen. The signal intensity changes of the intracranial part on MRI appeared hypointense on T1-weighted images and homogeneous enhancement with dural tail sign and absence of signal voids on intracranial angiography.

![FIGURE 5. (A) Angiography showed prolonged tumor stain into draining veins in venous phase. (B) Post-transartery embolization angiogram shows more than 70% decrease of tumor stain.](image)

![FIGURE 6. Angiomatous meningioma dominated by excessive vascularization interspersed with small meningothelial tumor cells.](image)
contrast-enhanced scans; these are characteristic findings of the intracranial meningioma. These findings are different from those of paragangliomas, which appear hyperintense on unenhanced T1-weighted images, with flow voids within the tumor manifesting as salt and pepper appearance on contrast-enhanced T1-weighted images.

Schwannomas appear hypointense on T1-weighted images, like meningiomas; but, with contrast enhancement, they frequently display a similar salt and pepper pattern, which may be due to cystic degeneration in the background of the well-enhanced solid component. Besides, schwannomas usually have a characteristic well-circumscribed, elongated configuration following the course of the cranial nerves of origin, which is not characteristic of intracranial meningioma. T2-weighted imaging is less useful because different histologic types of meningiomas show variable signal intensities. For example, the angioplastastic type of meningioma has high signal intensity in T2-weighted sequence, whereas the fibrous type shows intermediate signal intensities.

Because meningiomas grow slowly without obvious symptoms during their development, 20% of intracranial meningiomas have grown outside of the cranial fossa by the time of diagnosis. It is reported that intracranial and extracranial components of meningiomas have different signal intensities on MRI. Unlike the MRI findings of the intracranial component in this case, the cervical component showed a salt and pepper appearance on contrast-enhanced T1-weighted images, with encasement of the carotid bifurcation, just like that seen in a schwannoma, and had a fusiform shape with smooth margins, just like that seen in a schwannoma. The former appearance may be due to rich intratumorous vasculature in the angiomatic type of meningioma, and the latter may be due to the cervical part of this meningioma confined within the tight carotid sheath when it extended downward from the jugular foramen. But because less than 1% of intracranial meningiomas extend to the lower part of the parapharyngeal space, other imaging studies are needed to delineate the nature of this tumor.

In general, unenhanced high-resolution CT of bone window is helpful for surgical planning and for diagnosis, because 3 tumors cause different bony reactions. It is reported that meningiomas usually have irregular margin of the jugular foramen with loss of the normal cortex, which looks mixed permeative-sclerotic pattern. Paragangliomas cause mixed permeative-destructive bony reaction, with erosion of the jugular foramen margin without preservation of the underlying architecture. Schwannomas give an expanded and scalloped, well-defined, corticated margin of the jugular foramen, because the jugular foramen is gradually widened by pressure erosion. But in this case, CT scan demonstrated that the jugular foramen meningioma also widened the jugular foramen with smooth margin, like that seen in a schwannoma, and caused pure permeative bony reaction without sclerotic change. The etiology is unknown and may be related to the histologic type of this tumor.

Angiography is very helpful for differential diagnosis of jugular foramen tumors and essential for management of these lesions. On angiography, paragangliomas appear as rapid intense tumor stain with enlarged feeding arteries and early draining veins. Angiography of schwannomas usually shows the absence of strong tumor stains. Angiography of meningiomas shows rich tumor vasculature with obvious feeding vessels, like that seen in paraganglioma, but it shows prolonged tumor stain well into draining veins in venous phase, as shown in our case. Preoperative embolization of this tumor with rich vasculature helped to reduce bleeding intraoperatively, and the tumor had a gelatin-like appearance and was easily with preservation of the entrapped cranial nerves and carotid artery.

**CONCLUSIONS**

Meningioma of the jugular foramen is a rare tumor that clinically mimics glomus jugulare and schwannoma. Accurate differential diagnosis between meningiomas and paragangliomas or schwannomas is possible in most cases with attention given to radiologic findings, including a permeative-sclerotic bony reaction around the jugular foramen on CT, lack of salt and pepper sign, and the presence of dural tails on MRI in general meningiomas. A large angiomatic type of meningioma as seen in our case may have CT and MRI findings overlapping with those of paragangliomas and schwannomas. In this case, venous drainage phase of tumor stain shown by angiography is another useful imaging finding.

**REFERENCES**