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Case Report

A rare case of jugular foramen meningioma

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Introduction

Meningiomas are the commonest nonglial neoplastic lesions of the brain and constitute about 20% of all neoplastic intracranial lesions.¹ Posterior fossa meningiomas constitute 10% of all intracranial meningiomas and their locations include the cerebellar convexity, lateral tentorium, cerebello-pontine (CP) angle, jugular foramen, petroclival surface, and foramen magnum.² Jugular foramen meningiomas (JFM) are exceptionally rare with only a few cases reported in the literature.³ We report one such rarity here.

Case report

A 38-year-old male patient presented to our hospital with chief complaints of headache and vomiting of 3 months duration.

These symptoms were gradually progressive and were associated with one-month history of gait disturbance, hearing loss, and tinnitus. On examination, he had positive cerebellar signs on the right side. However, there was no neurological deficit.

The patient was referred for noncontrast computed tomography (NCCT) of head, which revealed a lobulated, fairly well-defined heterogeneously hypodense extra-axial mass lesion with epicenter in the right jugular foramen. The lesion was seen to extend superomedially to the right CP angle. The right jugular foramen was widened with associated destruction of its walls (Fig. 1). Few areas of necrosis were seen in the lesion. However, no calcifications were seen. The imaging characteristics were suggestive of posterior fossa meningioma, likely jugular foramen meningioma, and the patient was referred for contrast-enhanced magnetic resonance imaging (CEMRI) for confirmation.

CEMRI of brain revealed a lobulated, mixed solid and cystic extra-axial mass lesion in the right jugular foramen with destruction of its walls. Superomedial extension of the lesion was seen into the right CP angle. The lesion was heterogeneously hypointense on T1WI, heterogeneously hyperintense on T2WI, and showed moderate enhancement after administration of intravenous gadolinium contrast. Central non-enhancing areas were seen in the lesion. Mass effect was seen because of the lesion with compression of pons, right middle cerebellar peduncle, and right cerebellar hemisphere. Partial obliteration of the right CP angle cistern was also seen (Figs. 2 and 3).

The patient was taken up for surgery and the tumor was extirpated via right retromastoid suboccipital craniectomy. Complete macroscopic removal of the intradural tumor, without resection of its dural attachment and of its extradural extension, was done – consistent with Simpson's

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Fig. 1 – (A) Noncontrast computed tomography (NCCT) of head at the level of jugular foramen in cerebrum window showing a heterogeneously hypodense mass lesion in the right jugular foramen. (B) Noncontrast computed tomography (NCCT) of head at the level of jugular foramen in bone window showing widened and destructed walls of right jugular foramen.

Fig. 2 – (A) T1W axial image showing heterogeneously hypointense mass lesion in the right jugular foramen with superomedial extension into the right cerebellopontine (CP) angle. (B) T1W postcontrast axial image showing moderate enhancement of the lesion with central nonenhancing areas.

grade III excision. All the cranial nerves were anatomically preserved. The tumor in the jugular foramen was not removed, as it would have resulted in further morbidity. However, postop, the patient developed palsy of right IX, X, and XI nerves.

Histopathological examination of the tumor showed well-demarcated lobules of meningotheial cells with many whorls. In addition, a few psammoma bodies were seen interspersed within (Fig. 4). The findings were highly suggestive of a meningotheial meningioma. Based on the imaging and histopathological findings, the patient was diagnosed to be a case of jugular foramen meningioma.

The patient underwent Gamma Knife Stereotactic Radiosurgery (GKRS) for the residual tumor in the jugular foramen.

Follow-up imaging 6 months after GKRS showed mild decrease in the size of the tumor in jugular foramen. Presently the patient is stable and his right-sided IX, X, and XI cranial nerve palsy is gradually improving. The patient is planned for regular clinical and imaging follow-up.

Discussion

Meningiomas are benign tumors, which are derived from the arachnoid cells.⁴ JFM form a small group of intracranial meningiomas, with only a few cases reported in the literature.⁵ JFM most likely arise from arachnoid villi associated with the jugular bulb.⁶

Fig. 3 – (A) T1W postcontrast sagittal image showing enhancing mass lesion in the right jugular foramen with superomedial extension into the right cerebellopontine (CP) angle. (B) T2W coronal image showing heterogeneously hyperintense lesion in the right cerebellopontine (CP) angle with compression of pons and right cerebellar hemisphere.

The jugular foramen is a canal in the petrous temporal bone and consists of pars nervosa (nervous part) and pars vascularis (vascular part). The main contents of the canal are IX, X, XI cranial nerves, internal jugular vein, and meningeal branches of ascending pharyngeal artery.³

JFM may present with multiple symptoms depending on the size and location, including hearing loss, tinnitus, and lower cranial nerve deficits. Some other presentations can be with headache, visual disturbance, gait abnormality, dysphagia, hoarseness, and dysphonia. Our patient presented with gait disturbance, hearing loss, and tinnitus.

A high degree of suspicion, while taking history and while performing investigations, is required for the diagnosis of JFM. Investigations that can be done for diagnosing JFM include computed tomography (CT), magnetic resonance (MR)

imaging, MR venography, MR angiography, and conventional catheter angiography. JFM are primarily centered on the jugular foramen with centrifugal spread to CP angle, middle ear, and neck. On CT, JFM are iso- to hyperdense and show dense enhancement after administration of i.v. contrast. Hyperostosis may be seen in the surrounding bones, which may show destruction. On T1-weighted MR imaging, they are iso- to hypointense; on T2-weighted MR imaging, they show intermediate to hyperintense signal, and on postcontrast imaging, they show moderate to intense enhancement. MR imaging may also reveal enhancement of adjacent dura, consistent with dural tail sign.⁵ MR angiography and MR venography are extremely useful adjuncts to conventional MR imaging for mapping the cerebral arterial and venous anatomy prior to surgery. Conventional angiography has an important role to play, if one is contemplating presurgical embolization. However, the role of embolization is debatable. In our patient, we had not contemplated preoperative embolization, and hence he was not advised conventional angiography.

The differential diagnosis of JFM includes paragangliomas, schwannomas, epidermoid tumors, chordomas, aneurysms, metastases, and ependymomas, with two main differentials being paragangliomas and schwannomas. Paragangliomas usually erode and destroy the adjacent bone and do not show calcifications. Schwannomas appear iso- to mildly hyperdense on CT and may cause enlargement of jugular foramen without any bone destruction.⁷

Bakar classified JFM into the following types: Type I – primarily within jugular foramen, Type II – primarily extension to the CP angle, Type IIa – extension to the CP angle and middle ear, Type III – primarily extension to neck, Type IIIa – primarily extension to neck and middle ear, Type IV – extension to CP angle and neck, and Type IVa – extension to CP angle, neck, and middle ear.³ JFM in our patient fitted into Type III.

Treatment of JFM includes observation, surgery, radiosurgery, and combination of surgery and radiosurgery. Multiple surgical approaches have been described out of which lateral

Fig. 4 – Photomicrograph of H&E stained section of the tumor showing well-demarcated lobules of meningothelial cells with interspersed psammoma bodies.

retromastoid suboccipital craniectomy is commonly employed.² The same approach was employed in our case with Simpson's grade III excision with residual tumor treated with GKRS. Regular follow-up of these patients is suggested to look for recurrence.

Conclusion

Although jugular foramen meningioma is an extremely rare disorder, it should be kept in mind in the differential diagnosis of posterior fossa mass lesions. The purpose of this paper is to highlight the fact that a high degree of suspicion while performing investigations leads to a correct preoperative diagnosis, better surgical planning, and better patient outcome.

Conflicts of interest

The authors have none to declare.

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