

Skull base chordoid meningioma: a case with confounding imaging features

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Introduction

The skull base, which supports the brain, contains vessels and nerves that enter or exit the skull. Skull base lesions can originate from the skull base itself from the adjacent intracranial or extracranial structures. They constitute a wide variety of conditions such as chordomas, cranial nerve sheath tumours and meningiomas, which can complicate radiological diagnosis. Imaging plays a major role in detection, localization and characterization of these lesions and is vital in surgical planning and follow up. Out of the many lesions involving the skull base, meningiomas usually have typical imaging features. However, we report a case with confounding imaging features, which proved to be a chordoid meningioma, which is a rare variant.

Case History

A 25-year-old male presented with progressive headache and vomiting for 3 days duration. He had a gradual onset hoarseness of voice, difficulty in articulating words, difficulty in swallowing, double vision and left sided facial pain over past one month. On examination, he had multiple left side cranial nerve (CN) palsies including CN-V to CN XII. He had no history of fever, and septic screening was negative.

He underwent a Contrast Enhanced Computed Tomography (CECT) of the brain and a subsequent Magnetic Resonance Imaging (MRI) of the brain. CECT showed a heterogeneously enhancing, diffuse extra axial mass in the base of the skull involving the middle and posterior cranial fossae, with its epicenter to the left of the midline. There were no internal calcifications (Figure 1).



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Figure 1: Axial CECT brain showing the heterogeneously enhancing extra axial mass lesion denoted by asterisks.

The medial aspect of the apex of the left petrous temporal bone was eroded and the left jugular foramen was widened (Figure 2).

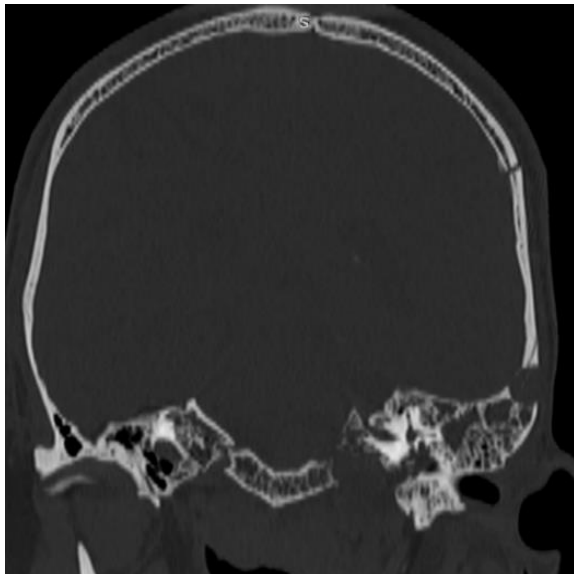


Figure 2: Coronal CECT bone window showing the widened left side jugular foramen with temporal bone erosions.

On MRI, the lesion was well defined and lobulated and showed low signal intensity on T1 weighted imaging, high signal on T2 weighted imaging and FLAIR sequences, and heterogenous contrast enhancement on post contrast imaging (Figure 1-6).

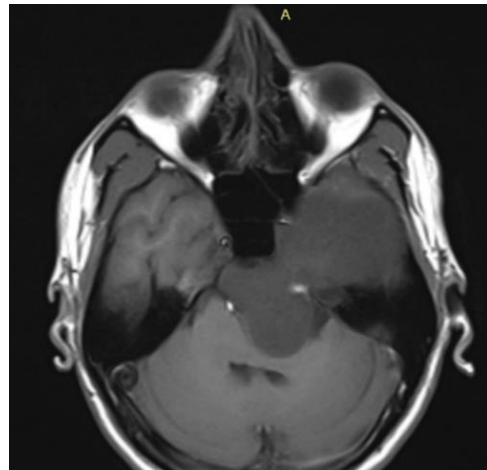


Figure 3: Axial T1W MRI brain showing T1 low signal intensity lesion.

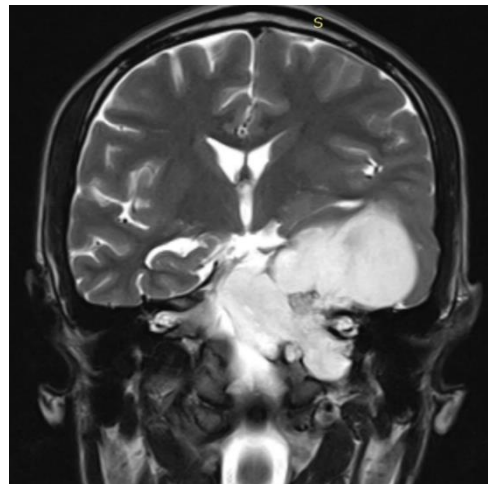


Figure 4: Coronal T2W MRI brain showing the extra axial lesion with high signal intensity, extending out of the left jugular foramen (*).

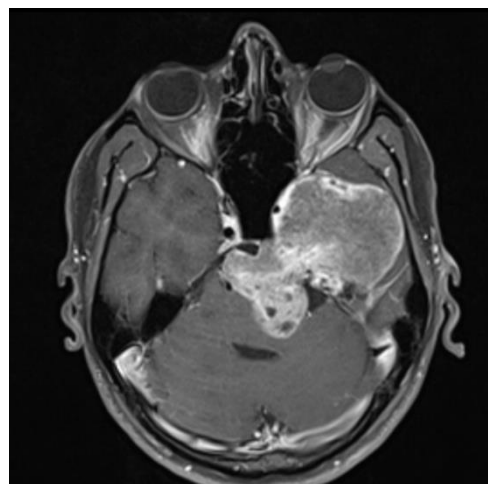


Figure 5: Axial T1W post contrast MRI brain demonstrating heterogeneous contrast enhancement in the lesion and extension into the posterior cranial fossa with mass effect on the brain stem. The continuity of the lesion is also evident.

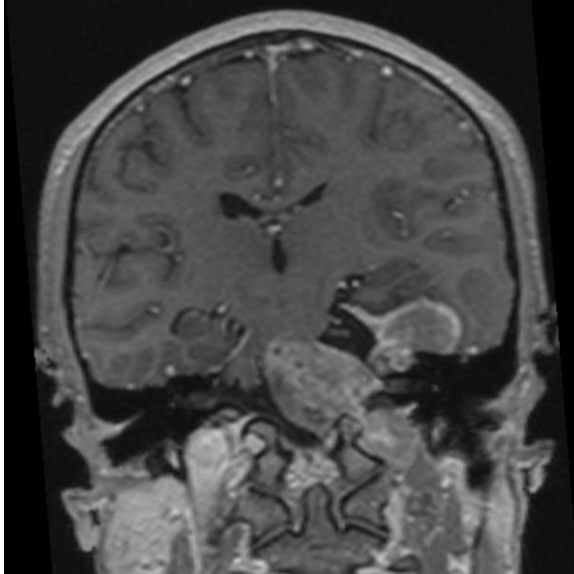


Figure 6: Coronal section of T1W post contrast MRI brain demonstrating the lesion extending out of the left jugular foramen

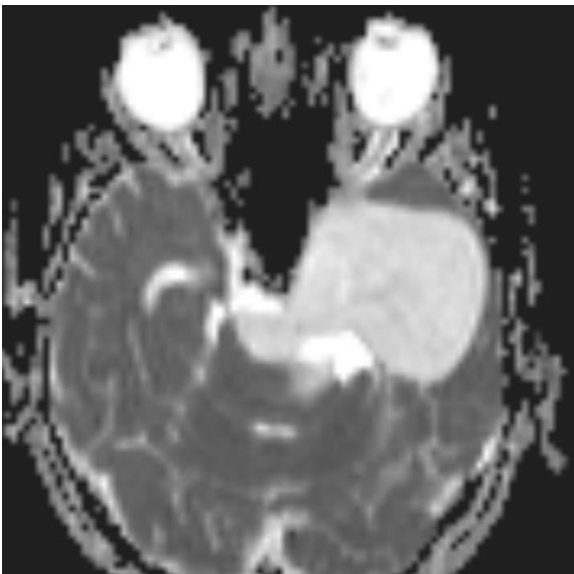


Figure 7: Axial MRI ADC map shows high signal intensity of the lesion compared to adjacent white matter, in keeping with facilitated diffusion

The epicenter of the lesion was in the left petrous temporal region, however, the lesion extended posteriorly to the pre pontine cistern exerting mass effect on the brain stem. It further extended inferiorly through the left jugular foramen (Figure 4, Figure 6) Antero superiorly the lesion extended into the left Meckel's cave compressing the temporal lobe. The

cavernous sinus was intact. There was no hydrocephalus. Diffusion tensor imaging showed that the adjacent projection fibers and association fibers were displaced but not disrupted (Figure 8).

Based on the growth pattern of the lesion, a differential diagnosis of a nerve sheath tumor or an atypical meningioma was made. The patient underwent debulking surgery (Figure 9) and the histological diagnosis was of a chordoid meningioma. He was referred for further oncological management with a view to commencing radiotherapy.

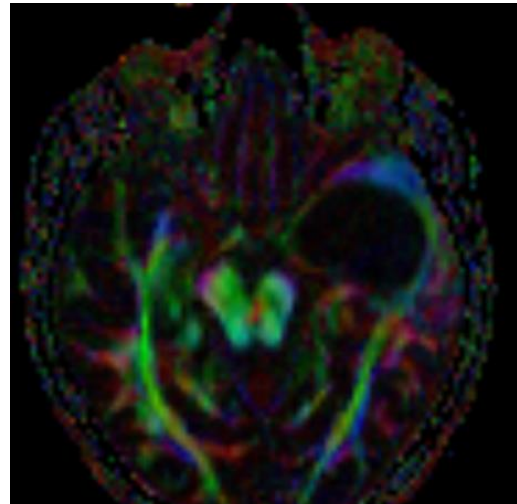


Figure 8: Diffusion Tensor Imaging showing the displaced association and projection fibers.



Figure 9: Post operative CECT showing residual lesion in the posterior fossa.

Discussion

As skull base lesions are complex, it is a diagnostic challenge to both the clinician and the radiologist. Their intricate clinical presentations and varying etiologies highlight the significance of precise diagnostic imaging. Both Computed tomography (CT) and Magnetic Resonance Imaging (MRI) are used in the imaging of the skull base. The combination of these two modalities helps in the lesion characterization and evaluation of the bony involvement, extracranial soft tissue involvement and vascular involvement¹ similar to our case.

Skull base meningiomas account for up to 50% of all meningiomas.^{1,2,3} Similar to meningiomas elsewhere, they are more common in middle aged females and small lesions will often be asymptomatic.¹ A larger lesion on the other hand can cause both nonspecific symptoms as well as site specific symptoms such as anosmia, proptosis and ophthalmoplegia or lower cranial nerve palsies¹ similar to our patient. Typically, meningiomas are well defined, extra-axial lesions which are hyperdense in non-contrast CT, and show iso intensity to grey matter in T1 and T2 weighted sequences and avid homogenous contrast enhancement in both post contrast CT and MRI.¹ En plaque meningiomas will have a more infiltrative growth pattern over the dura.⁴ Apart from the location and the enhancement pattern, the presence of a dural tail, calcification and underlying bony hyperostosis will aid in the imaging diagnosis.

However, some meningiomas can demonstrate atypical imaging features which would be confounding, as in this case. Some rare angiomatous or chordoid sub types will show high signal intensity in

T2 weighted imaging while microcystic sub types will be hypointense T1 W with high signal in T2 weighted sequences.^{4,5} Rare cystic meningiomas can have intratumoral or extra tumoral cystic components.⁴ The enhancement pattern can also be variable. Literature states that there can be heterogenous enhancement or peripheral ring enhancement due to presence of central necrosis and areas of calcifications.^{4,6,7} Meningiomas commonly cause underlying bony hyperostosis, but it is said that sometimes bone osteolysis can also occur.^{1,4} Expansion of foramina are also unusual but reported in literature.⁴ When these atypical imaging findings are encountered, specially in skull base lesions, the imaging diagnosis can be challenging. In our case, the presence of T2 high signal intensity, heterogenous contrast enhancement in the absence of areas of calcification and underlying bony erosion and lysis with expansion of skull base foramina were the key confounding imaging features. These imaging features along with patient demographics were against a diagnosis of a typical skull base meningioma. The lesion had a more lobulated appearance with extension through widened skull base foramina as well, which were suspicious for a nerve sheath tumor. Patient's clinical presentation and patient demographics were also favoring this.

The definitive diagnosis of a chordoid meningioma can only be made histologically. Chordoid meningiomas are a very rare sub type which are WHO grade II lesions.⁵ According to literature, their incidence is 0.5% of all meningiomas.^{8,9} Initially these chordoid meningiomas were described as affecting primarily young patients, in association with Castleman syndrome.^{8,10} However, a case series

published by Couce et al, found that in the adult population, chordoid meningiomas were not associated with Castleman syndrome.⁹ It is also of note that chordoid meningiomas are extremely rare in the posterior fossa. Pond et al, in a case series analysis, found that almost all chordoid meningiomas show hyperintensity in T2 images with the majority showing avid homogenous contrast enhancement.¹¹ Marked peritumoral oedema was also noted in some cases.¹¹ There are studies that have reported the presence of facilitated diffusion (which is seen as high ADC values), as highly characteristic of chordoid meningiomas.^{11,5} Pond et al, also concluded that there is a statistically significant increase in ADC values in chordoid meningiomas when compared to other subtypes, thus aiding in the differentiation from other subtypes.¹¹ The clinical implication of being able to predict this subtype preoperatively is that when it comes to atypical meningiomas, the extent of the resection will largely predict the recurrence rates.¹¹ Studies have shown that chordoid meningiomas which were totally resected did not show evidence of recurrence, whereas those which underwent subtotal resection almost always recurred.^{11,9} Therefore, if we can identify atypical meningiomas and predict the histological sub type through imaging, it will help in the surgical planning to achieve a total resection. Even in the cases of difficult total resections, the preoperative prediction will help in initiating more rigorous post operative treatment and follow up.¹¹ In conclusion, skull base lesions can have a wide range of differential diagnosis and imaging plays a key role in identification and characterization of lesions. Meningiomas are a common entity in the skull base, but atypical imaging findings

can pose a diagnostic challenge. The awareness of atypical imaging features can not only aid in accurate preoperative diagnosis, but also help in predicting rare histological variants such as chordoid meningiomas, which would in turn play a major role in surgical planning and follow-up.

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Conflicts of interest

The authors have no conflicts of interest to declare.

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