

Cerebellopontine angle meningioma extending into internal auditory canal presenting with unilateral facial weakness and sensorineural hearing loss: Case presentation with review of literature

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Abstract

Tumour types other than schwannomas are uncommonly encountered in the cerebellopontine angle, and unusual lesions extending into the internal auditory canal are even rarer. We herein present a case of a 50-year-old woman with a right cerebellopontine angle meningioma (measuring 34mm x 35mm x 36mm), arising from the posterior surface of the petrous bone or the petrotentorial junction, secondarily involving the internal auditory canal. Contrast enhanced MRI showed typical appearance of a meningioma, with calcified flat surface against the petrous bone, a hemispheric or ice-cream-cone shape sign, adjacent hyperostosis, and 'dural tail' extending from the tumour. Initial clinical evaluation revealed continued presence of a stable neurological deficit such as unilateral headache, hearing loss, reduced taste sensation, positive corneal and conjunctival reflex along with right facial weakness (House-Brackmann grade III) signalling towards the involvement of V, VII and VIII cranial nerve. The variation in the anatomy of the faciocochlear nerve bundle in relation to the tumour has to be kept in mind, and preservation of these vital neural structures should be the goal in every case. Tumour resection was performed through a suboccipital-retrosigmoidal approach with a good outcome. Ours is an unusual case report analysing the diagnostic and therapeutic issues, surgical management strategies and treatment outcomes related to this rare disease entity.

Key words: Cerebellopontine angle meningioma, Internal auditory canal, Sensorineural hearing loss, Unilateral facial weakness

INTRODUCTION

Meningiomas are the second most common intracranial neoplasms that account for 15% of brain tumours¹⁻⁷. Among intracranial meningiomas, 5%–10% are located in the anatomic region of cerebellopontine angle (CPA)^{8,9}. Although involvement and enlargement of internal auditory canal (IAC) is a common sign of schwannomas of the CPA, meningioma extension into the IAC can be seen⁸. Extension into the

IAC is rare and should be treated like a separate disease entity because preoperative diagnostic identification permits alterations in surgical strategy planning that allow for the more complete resection of the tumour and better clinical outcome. Due to this exceedingly rare occurrence, only few cases have been reported in world literature²⁻¹². Our patient, harbouring a CPA meningioma extending into IAC, pose a diagnostic and therapeutic challenge on account of anatomic location and site of dural attachment, diverse intimate relationships to important neurovascular structures and a variable constellation of presenting clinical symptoms and signs like unilateral headache, hearing loss and abnormal facial motor function^{9,10}. This case report describes in detail the clinical findings, typical signal characteristics, more specific imaging features such as presence or absence

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of calcification, a hemispheric or ice-cream-cone shape sign, adjacent hyperostosis, a 'dural tail', extension into one or more skull base neural foramina and enlargement of the IAC, which aids in a more specific diagnosis and skilled surgical management¹³⁻¹⁶.

CASE REPORT

A 50 year old woman with no prior medical history presented with two month history of persistent unilateral right sided symptoms of headache, gradual decrease in hearing and progressively developing right facial weakness over a month ago. Neurologic examination showed right facial weakness (House-Brackmann grade III) facial twitching and drooping of liquid diet from right angle of mouth. Patient also complained of decreased taste sensation in anterior two-third of tongue. Audiological examination with pure tone audiography showed that hearing on the right side was decreased to 50 dB, pure tone average (500–3000 Hz), with a word recognition score of 60%. Neuroimaging examination (CT and MR imaging) was performed which revealed a well-defined extra axial abnormal lesion (measuring 34mm x 35mm x 36mm) in right C.P. angle region appearing iso- to hypointense on T1 (Figure 1a) and hypo to hyperintense on T2W sequence (Figure 2a) with areas of blooming (haemorrhage/ calcification) on gradient sequence (Figure 5). There was moderate

heterogeneous enhancement on post contrast study (Figure 1B and figure 3). The lesion extended laterally into ipsilateral internal auditory canal causing widening of the same. The lesion caused widening of C.P. angle cistern and showed significant mass effect on adjacent pons, right middle cerebellar peduncle and compressed origin of right fifth cranial nerve. Thus, diagnosis of meningioma confirmed from 'dural tails' (tumour infiltration along the dura shown by thickened and enhanced dura) and 'the meningeal sign' (broad contact with the meninges and the tumour capsule shown by meningeal contrast enhancement adjacent to the tumour (Figure 1-5). The patient underwent surgery, including involved dura and bone, by means of lateral suboccipital-retrosigmoidal approach in the semi-sitting position with drilling of the posterior wall of the IAC to obtain the best visualization. The tumour showed hypervascularity and the typical appearance of meningioma with numerous small calcifications under the operating microscope. The frozen histologic section also confirmed a meningioma. Immunohistochemistry showed that the tumour cells stained strongly for vimentin and did not stain for S-100 protein, features consistent with meningioma. Hearing was preserved but postoperative facial weakness (House-Brackmann grade I) was encountered temporarily although faciocochlear nerve bundle in relation to the tumour was preserved.

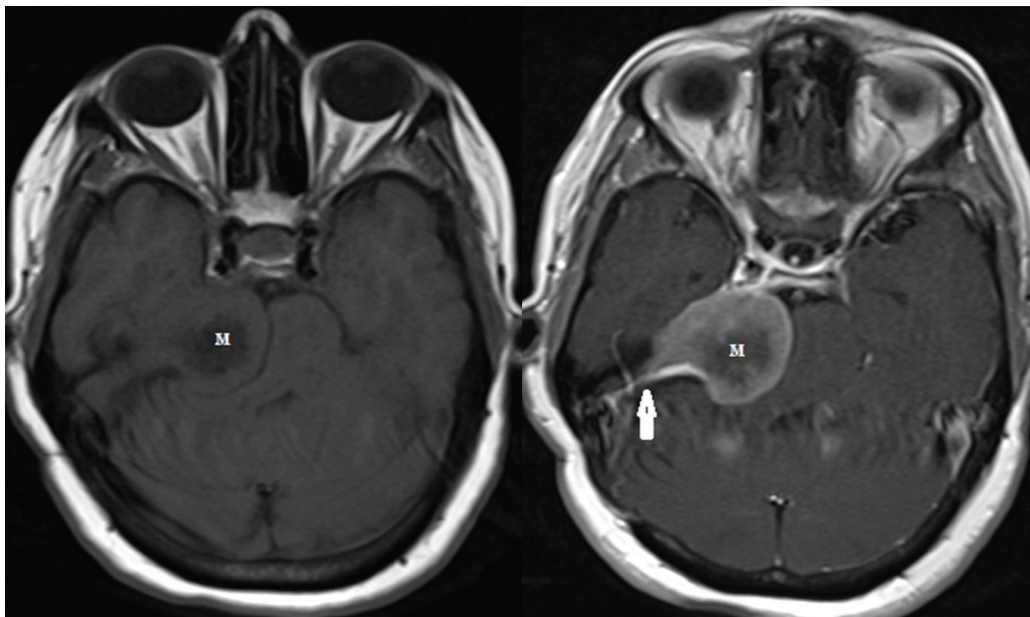


Figure 1(a): Axial T1W image showing extra-axial abnormal lesion (M) in right CPA region appearing isointense at periphery and hypointense in center.

Figure 1(b): Contrast-enhanced axial T1-weighted MR image reveals extra-axial abnormal lesion in right CPA region showing heterogeneous enhancement and dural tail along tentorium.

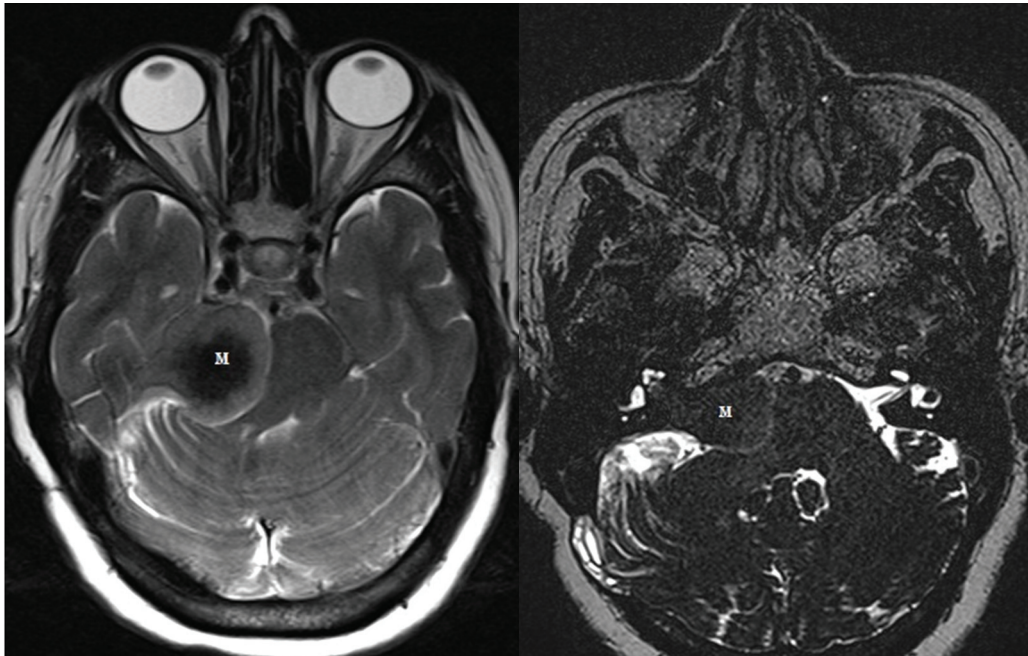


Figure 2(a): Axial T2W image showing extra-axial abnormal lesion (M) in right CP angle region appearing hyperintense at periphery to hypointense in center.

Figure 2(b): Axial T2W SPC image showing extra-axial abnormal lesion (M) in right CP angle region extending in to ipsilateral internal auditory canal.

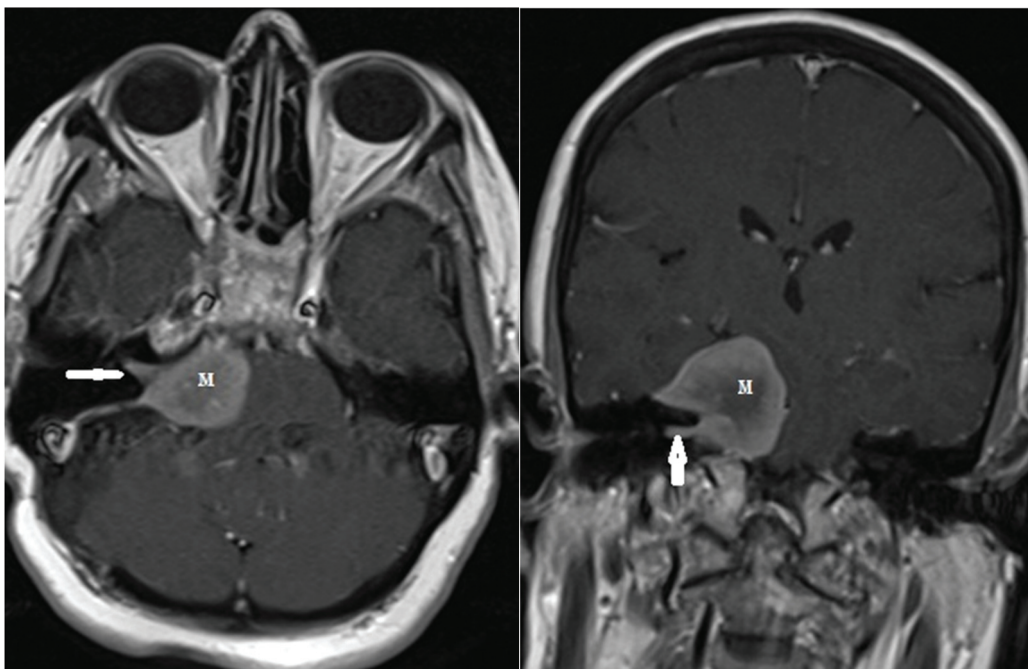


Figure 3(a): Contrast-enhanced axial T1-weighted MR image and **Figure 3(b):** Contrast-enhanced coronal T1-weighted MR image showing extra-axial abnormal heterogeneously enhancing mass (M) in right CP angle region extending in to ipsilateral internal auditory canal (marked by white arrow), increasing its conspicuity.



Figure 4: Axial NCCT image showing extra-axial calcified lesion (M) in right CP angle region, lesion density measures more than 550 H, consistent with calcification within meningioma.

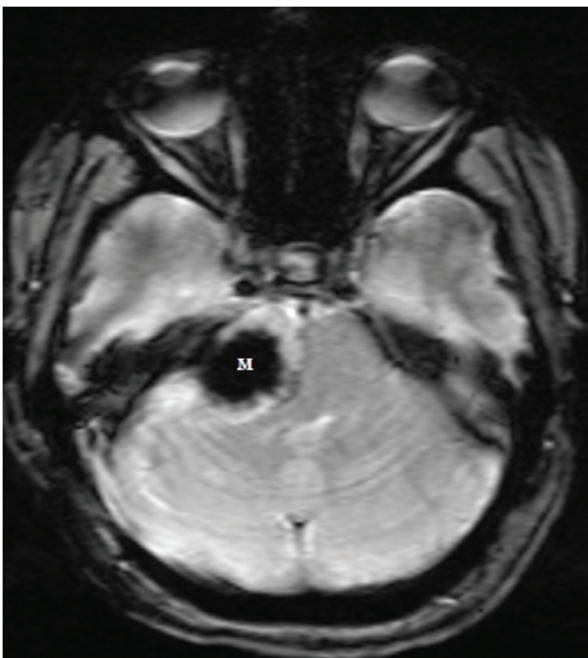


Figure 5: Axial GRE image showing extra-axial abnormal lesion (M) in right CP angle region showing areas of blooming consistent with calcification.

DISCUSSION

Although meningiomas are the second most common tumour of the CPA, CPA meningiomas extension into IAC is exceedingly rare¹⁷⁻²¹. Meningiomas arise from arachnoid villi, which are invaginations of the arachnoid mater along the walls of the dural and venous sinuses especially around the superior sagittal sinus. Previous studies reported that arachnoid villi were distributed not only along the dural sinuses and in the gasserian envelopes but also occasionally exist along the neural structures and their foramina, including the IAC^{22,23}. This can be explained embryonically. The seventh and eighth cranial nerves arise from a common primordium. At 5 weeks of gestation, the fibres of the facial nerve exit the neural tube along with a sheath of arachnoid and dura. CPA meningiomas most likely arise from the arachnoid villi along the porus acusticus (opening between the CPA cistern and IAC) and gasserian envelope.

CPA meningiomas extending into IAC clinically mimic vestibular schwannomas (VS) and other lesions that intrude the IAC. Most of the cases initially present with a hearing problem. The subtle difference is that facial nerve symptoms like facial paresis are more likely to occur with meningiomas than with vestibular schwannomas when the size is small. High-spatial-resolution Magnetic Resonance Imaging (MRI) is the imaging of choice. Bonneville et al.¹³⁻¹⁴ reported categorisation of tumours of the cerebellopontine angle (CPA) on the basis of key neuroimaging features on contrast-enhanced MRI, which aid in more specific diagnosis and limit the differential diagnosis. Enhancing extra-axial lesions, such as in the case presented here, account for 80% to 95% of CPA tumours. The differential diagnosis for enhancing extra-axial lesions includes lesions of the nerves, lesions of the arteries, and lesions of the meninges. Accurate radiographic diagnosis of CPA tumours with intracanalicular extension from VS is often difficult solely on the basis of signal intensity of MR images of these masses. Both lesions show isointense to hypointense on T1-weighted MR images and are of variable signal intensity on T2-weighted MR images. Contrast enhanced MR suggest bright enhancement in both lesions. Needless to say, consistent with these findings, when a patient is found to harbour an enhancing mass in the IAC, it is usually assumed to be a vestibular schwannoma. However, there are few radiodiagnostic findings that should raise the suspicion of intracanalicular meningioma. On the unenhanced MR, meningiomas are often isointense with brain on T1 and T2 weighted images. Extra-axial mass effect suggested by white matter buckling, a

rim of CSF around the mass, a pial vascular rim and a shorter T2 of the mass are described as characteristics of meningioma. Calcification and a 'dural tail' may be nonspecific^{16, 24} but broad-based extension into the petrous bone and a rugged medial tumour surface are significant clues to the diagnosis of meningiomas, whereas vestibular schwannomas usually have a more spherical shape and have a smoother surface. Nager and Masica²⁵ reported that intracanalicular meningiomas can invade the labyrinthine segment, surrounding petrous bone and cochlea by following their individual nerve fibres to their ends. Conversely, extensive bone invasion is unusual in vestibular schwannomas. Thus, the presence of wide bone infiltration around the IAC is suggestive of diagnosis of meningioma. However, VS compresses the facial nerve and displaces it rostrally and medially while intracanalicular meningiomas—the tumour did not just compress the facial nerve but intimately involved it. Contrast enhanced MR suggest that MR is better suited for identifying the extra-axial location of the tumour, the broad contact with the meninges, rugged medial tumour surface, the tumour capsule and meningeal contrast enhancement adjacent to the tumour, i.e., the meningeal sign. CT is, however, superior in demonstrating calcification and atypical tumour density. Both methods provided nearly equal results in demonstrating mass effect, hyperostosis and contrast enhancement. Contrast enhanced MR (CEMRI)

is particularly superior in the diagnosis of meningiomas of the skull base, posterior fossa and high convexity.

Nevertheless, resection with attempts at anatomical continuity is the gold standard. Meticulous skilled surgical sharp dissection is very important to avoid damage to the trigeminal, cochlear and facial nerve. Surgical approaches include the most commonly used translabyrinthine approach, as well as the retrosigmoid approach, transmastoid or middle fossa approach, combined middle fossa–transmastoid approach, and transmastoid–transparotid approach. This is often performed with a cable nerve graft interposition of either the sural or greater auricular nerve.

CONCLUSION

Surgical management aims at radical resection of the tumour mass along with the attached dura and the invaded petrous bone to prevent recurrence, despite maintaining the anatomical continuity and preserving the faciocochlear nerve bundle. Preoperative suspicion of intracanalicular CPA meningioma should be confirmed by intraoperative histologic analysis.

CONFLICT OF INTERESTS

The authors declare that they have no conflict of interests.

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