

Pontomedullary white epidermoid: a rare cause of tinnitus

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ABSTRACT Intracranial epidermoid cysts are relatively rare congenital inclusion cysts. They are the third most common cerebellopontine angle cistern mass after vestibular schwannoma and meningioma. White epidermoid is a rare variant. We present the first case, to our knowledge, of a white epidermoid cyst anterior to the pontomedullary junction, emphasising its imaging appearance, location and the importance of pre-operative diagnosis, which may reduce operative complications.

Keywords: pontomedullary, T1 hyperintense, white epidermoid
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INTRODUCTION

Intracranial epidermoid cysts account for 0.2%–1.0% of all primary intracranial tumours.⁽¹⁾ Typically, an intracranial epidermoid cyst has signal intensities that are similar to those of cerebrospinal fluid (CSF), i.e. hypointense on T1-weighted magnetic resonance (MR) images and hyperintense on T2-weighted MR images. A rare variant that shows reverse signal characteristics is known as ‘white epidermoid’.⁽²⁾ Here, we report the first case of white epidermoid anterior to the pontomedullary junction, which was hyperdense on computed tomography (CT), hyperintense on T1-weighted and hypointense on T2-weighted MR images. So far, to our knowledge, this is also the first case with a pre-operative diagnosis.

CASE REPORT

A 17-year-old boy presented with complaints of a gradual loss of hearing and tinnitus on the right side over the last five years. He had unsteadiness of gait and a tendency to fall on the right side for the past one year. Neurological examination revealed sensorineural hearing loss.

CT imaging showed a well-defined, lobulated, non-enhancing hyperdense lesion in the right prepontomedullary region (Fig. 1a). MR imaging revealed a 2.0 cm × 1.1 cm × 1.7 cm lesion that was hyperintense on T1-weighted image (Fig. 1b) and hypointense on T2-weighted image, anterior to the right pontomedullary junction (Fig. 1c). The lesion was isointense to minimally hyperintense relative to CSF on diffusion-weighted images (DWI) (Fig. 1d). However, the mean apparent diffusion coefficient (ADC) value within the lesion was significantly low ($0.16 \times 10^{-3} \text{ mm}^2/\text{s}$), suggestive of restriction (Fig. 2a). Gadolinium-enhanced T1-weighted images revealed no enhancement. The lesion was seen in close proximity to the origin of the right VIII cranial nerve, encasing the bilateral vertebral arteries, and causing mild mass effect on the medulla and pons anteriorly. These features suggested a differential of early sub-acute haemorrhage. 3-D time

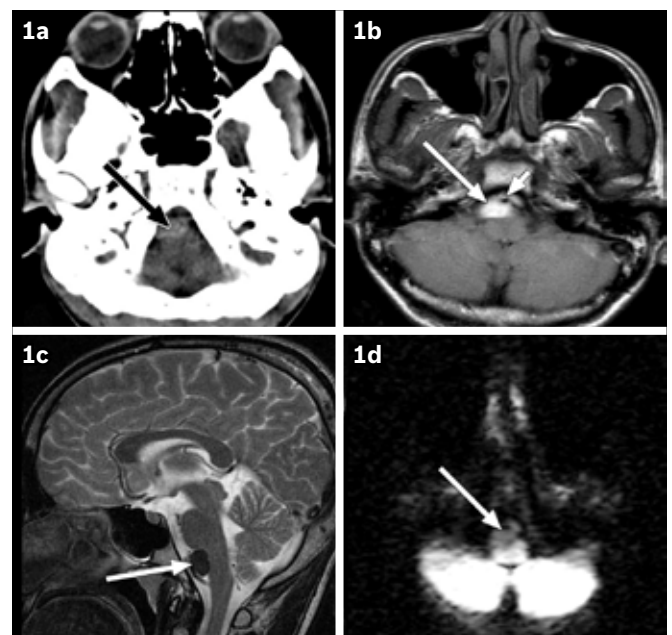


Fig. 1 (a) Non-contrast axial CT image near the pontomedullary junction shows a hyperdense mass (arrow) with average HU 70; (b) Axial T1-W MR image shows a well-defined hyperintense mass (arrow) anterior to the medulla with a vertebral artery (arrowhead) traversing anterior to the mass; (c) Sagittal T2-W MR image shows a hypointense intradural extra-axial mass anterior to the pontomedullary junction (arrow); (d) Axial diffusion-weighted MR image shows the lesion to be isointense to minimally hyperintense relative to cerebrospinal fluid (arrow).

of flight MR angiography disclosed no vascular abnormality. In comparison to the previous MR imaging performed six months back, the lesion showed no temporal change in size and signal characteristics, ruling out haemorrhage. Hence, a white epidermoid cyst was considered as the working diagnosis.

On surgery, the lesion was lobulated, with a pearly white appearance (Fig. 2b). The patient underwent a right retromastoid suboccipital craniectomy and total excision of the lesion. The

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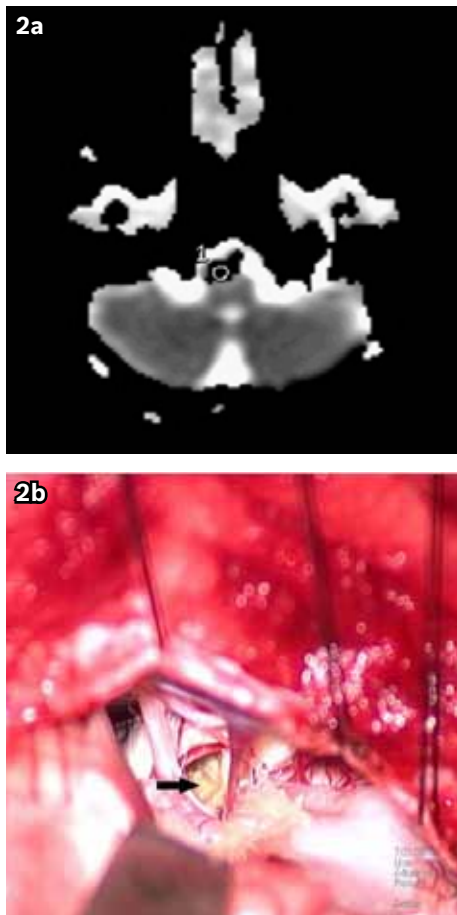


Fig. 2 (a) ADC map shows a mean value of $0.16 \times 10^{-3} \text{ mm}^2/\text{s}$ within the lesion; (b) Intra-operative photograph shows a pearly white lesion with lobulated margins (arrow).

lesion was in contact with the VII-VIII nerve complex superiorly, and inflammation of the cranial nerve (CN) VIII had likely caused the tinnitus. The lower CNs were ventral to the lesion, with the posterior inferior cerebellar artery draped dorsal to it. The patient had an uneventful postoperative period and was relieved of tinnitus. Histopathological examination revealed a cystic tumour with concentric lamellae of anucleated keratin (Fig. 3a) lined by stratified squamous epithelium (Fig. 3b), thus confirming our diagnosis.

DISCUSSION

Intracranial epidermoid tumours are rare congenital inclusion cysts that develop during the third to fifth week of embryogenesis.⁽³⁾ They occur off midline and are commonly located in the cerebellopontine angle (CPA) cistern (40%–50%), fourth ventricle (17%) and the sellar-parasellar region (10%–15%). They are also known as ‘mother-of-pearl tumours’ due to their well-defined, lobulated and glistening surface.⁽⁴⁾ The cysts contain a waxy keratohyalin material, crystalline cholesterol, lined by stratified squamous epithelium.

Typically, epidermoids are hypodense on plain CT image. On MR imaging, they are hypointense on T1-weighted and hyperintense on T2-weighted images and DWI. The mean ADC values of epidermoid tumours are lower than those of CSF.^(3,5,6) White epidermoids, which are the rarer variety, are hyperdense

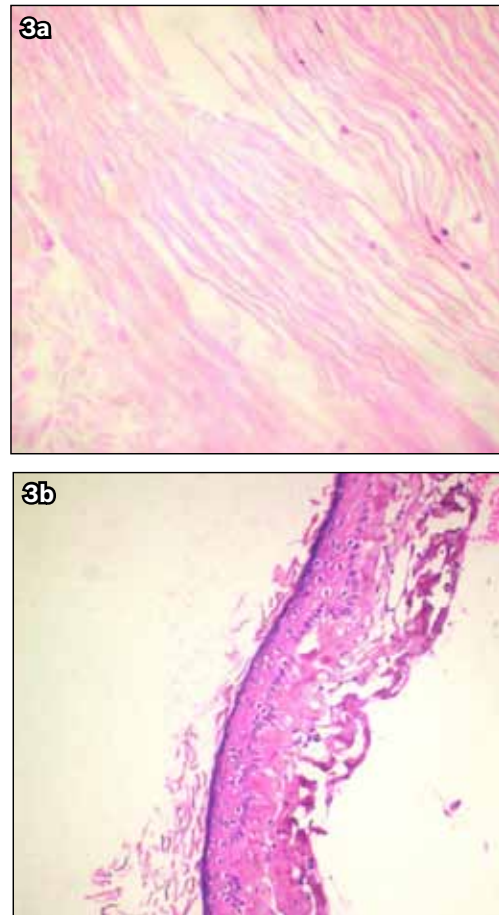


Fig. 3 Photomicrographs of epidermoid cyst show (a) concentric lamellae of anucleated keratin; and (b) occasional focus showing stratified squamous epithelium (Haematoxylin & eosin, $\times 400$).

on CT, hyperintense on T1-weighted images and show variable signals on T2-weighted images.⁽²⁾

White epidermoids have been described in the CPA, posterior fossa and frontotemporal region.⁽¹⁻⁴⁾ Chen et al⁽⁴⁾ described intracystic haemorrhage with breakdown products of haemoglobin causing paramagnetic effect as the cause of hyperintensity in both T1- and T2-weighted images. Epidermoid cysts are relatively avascular tumours; however, partial leakage of the cystic materials and subsequent surrounding granulation reaction may lead to neovascularisation, causing haemorrhage. This corroborates with another study,⁽²⁾ which found a high lipid content, methaemoglobin and haemosiderin as the causes of unusual signal in white epidermoid.

On the basis of chemical analysis of cystic fluid in a white epidermoid, Timmer et al⁽¹⁾ described a high total protein concentration and a large fraction (67%) of albumin as the causes of hyperdensity on CT and hyperintensity on T1-weighted images. They also emphasised that the amount of cholesterol and triglycerides was too low to cause a significant T1 shortening. The low signal intensity on T2-weighted images may be explained by the high viscosity of the fluid, similar to that described in chronically inspissated sinonasal secretions. These characteristics are similar to those in our case and were thought to be the cause of the unusual appearance. The differentials in our case were early subacute stage of haematoma, neurenteric cysts and

craniopharyngioma due to similar imaging characteristics.⁽⁷⁾ However, the absence of temporal resolution of the lesion over a period of six months ruled out haematoma. Intracranial neurenteric cysts are isodense to hypodense on non-contrast CT. They may be hyperintense on T1-weighted images but are usually hyperintense on T2-weighted images and FLAIR sequences,⁽⁸⁾ unlike our case. Craniopharyngioma is rare in this location and shows peripheral/nodular enhancement on contrast administration. Knowledge of the unusual imaging features of epidermoids suggested this entity as the likely pre-operative diagnosis. It is important to recognise epidermoids presurgically, as intra-operative rupture of the cyst may cause aseptic Mollaret's meningitis⁽⁹⁾ due to chemical reaction. These lesions may be misdiagnosed as bone tumours.^(2,4)

The uniqueness of this case is the location of the epidermoid anterior to the pontomedullary junction and near the midline. The presenting symptom of tinnitus had led to early imaging, which enabled us to obtain a pre-operative diagnosis and favourable outcome with appropriate presurgical planning for epidermoid resection. In conclusion, white epidermoids should be considered in the differential diagnosis of extra-axial T1-weighted hyperintense lesions. Knowledge of the imaging features of white epidermoids aids in

pre-operative diagnosis, resulting in improved surgical planning and patient outcome.

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