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Case Report

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Case Report on White Epidermoid Cyst and Review of its Literature

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Abstract

Background: Epidermoid cysts are rare slow-growing benign tumors showing typical imaging features in CT and MRI. However, few epidermoid cysts were reported to have atypical imaging features. Such epidermoid cysts are called 'white epidermoids'. Hereby we report a case of white epidermoid localized in the right cerebellopontine angle.

Case Description: 26-year-old female without any known co-morbidities presented with occipital headache and vomiting for the previous 1 month. Headache was associated with neck pain on the right side. Her clinical examination shows a positive Romberg test. Further, Magnetic resonance imaging was advised. On MR, a well-defined lobulated T1 hyper-intense and T2 hypo intense extra-axial lesion was noted involving cisterna magna. The lesion was showing a focal area of blooming within, with no area of diffusion restriction. Radiologically it was diagnosed as a white epidermoid cyst. The patient underwent surgery and the excised mass was sent for histopathological examination. Histopathology was consistent with an epidermoid cyst showing a cyst wall lined by keratinizing stratified squamous epithelium with hemorrhage into the cyst cavity. The patient had an uneventful post-operative period.

Conclusion: White epidermoid cysts are extremely rare. Atypical imaging features of epidermoid cysts should always be considered while differentiating an epidermoid cyst from other lesions.

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Introduction

Epidermoid cysts are rare slow-growing benign tumors constituting about 1.8% of all intracranial tumors [1,2]. These are congenital tumors, that arise from tissue sequestrated during the closure of the neural tube in embryogenesis [3]. Typically, an epidermoid cyst shows CSF density on CT and signal intensities broadly similar to CSF on T1 and T2 weighted MR sequences with heterogeneous signal suppression on FLAIR [4,5]. However very few cases have reported atypical imaging features and are called white epidermoid cysts. We report here a case of the epidermoid cyst with atypical imaging features in the cerebellopontine angle, which was radiologically diagnosed and further confirmed with histopathology and thereby briefly review its literature.

Case Presentation

Chief Complaints

36 years old female presented with occipital headache and vomiting for previous 1 month.

History of Presenting Illness

26 years old female presented with occipital headache and vomiting for previous 1 month. Headache was assosciated with neck pain.

History of Past Illness

Patient had no history of diabetes or hypertension.

Clinical Examination

On examination, she was conscious and alert. Her higher mental functions were normal. Bilateral pupils were normal with 3mm diameter and equally reacting to light. Extra ocular movements were full and power was 4/5 in all the limbs. Grade of deep tendon reflexes were normal in all the limbs. Sensory system was normal.

Diagnostic Assessment

Magnetic resonance imaging revealed a well-defined lobulated extra-axial mass measuring 3.1×4.2×5.5 cms in cistern Magna. It showed hyperintensity on the T1 weighted sequence and hypo intensity on T2 weighted sequences. Gradient sequences showed few punctate foci of blooming within. No areas of diffusion restriction or post-contrast enhancement noted within the lesion. The lesion is causing splaying of the paramedian aspect of bilateral cerebellar hemispheres and falx cerebelli. Mass effect is noted in the form of effacement of the median outlet of the 4th ventricle, anterior displacement of the medulla, V4 segments of bilateral vertebral arteries along with effacement of premedullary cistern. Focal edema was noted in the cervical cord, extending inferiorly up to the C3 level. Moderate dilatation of the supratentorial ventricular system with minimal trans ependymal

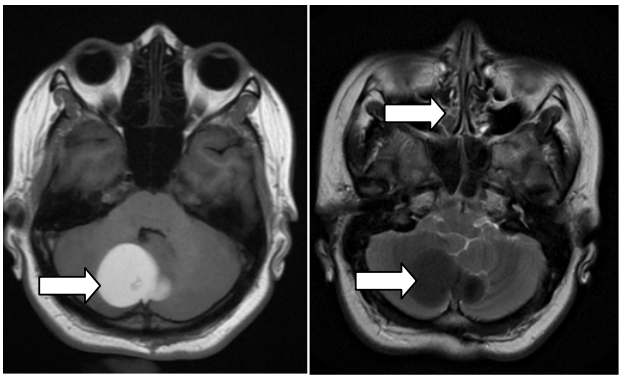

Figure 1: Magnetic resonance imaging - T1 weighted axial image showed hyper intense lesion (White arrow) in cisterna magna and T2 weighted axial image showed hypo intense lesion (White arrow) in the corresponding region.

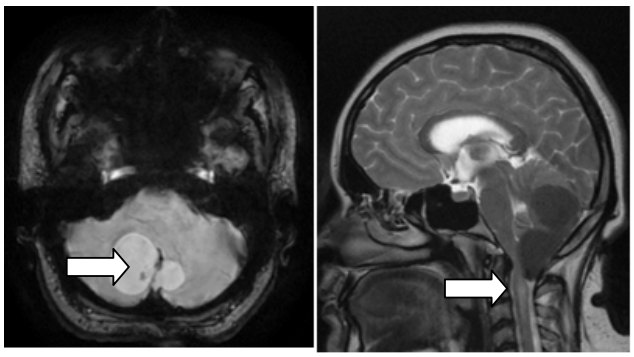

Figure 2: Magnetic resonance imaging - Gradient sequence (GRE e SWAN) showing focal area of blooming (white arrow) in the lesion and Saggital T2 weighted image shows cervical cord edema. E. DWI sequence shows no diffusion restriction.

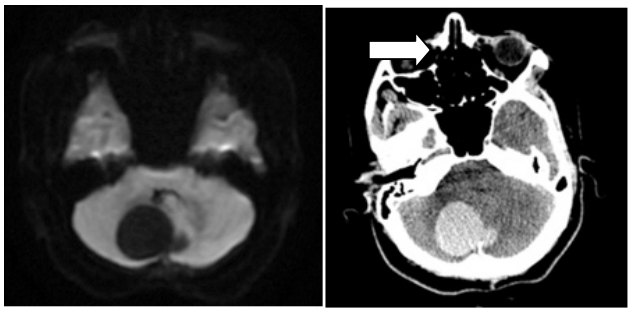

Figure 3: Magnetic resonance imaging - DWI sequence shows no diffusion restriction and Axial non contrast CT image shows hyperdense nature of the lesion.

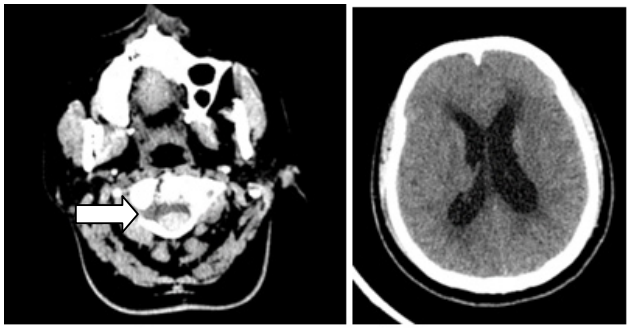

Figure 4: Magnetic resonance imaging - Axial non-contrast CT image at the level of upper cervical spine shows focal edema, with extension of lesion into spinal canal and Axial non-contrast CT image at the level of lateral ventricles shows moderate dilatation of the supratentorial ventricular system with minimal transependymal seepage.

seepage of CSF. The lesion shows partial suppression on the FLAIR sequence.

Follow up and Outcome

Patient underwent gross total excision of the tumor by right retromastoid suboccipital craniotomy and right C1 arch and posterior one third occipital condyle excision. The excised specimen was sent for histopathological examination. On gross appearance, the tumor was encapsulated, soft and suckable, yellowish in color showing mild vascularity. Microscopic examination revealed cyst wall lined by keratinizing stratified squamous epithelium and also bits of organizing hemorrhage with cholesterol granulomas and foamy macrophages. These features were consistent with epidermoid cyst with hemorrhage into cyst cavity.

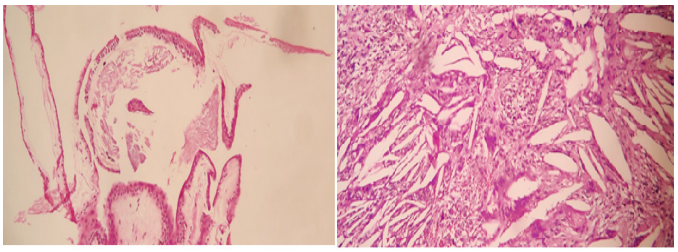

Figure 5: Fragmented strips of cyst wall lined by keratinising stratified squamous epithelium (Black arrow) and Organising hemorrhage showing cholesterol granulomas and foamy macrophages surrounded by lymphocytes.

Discussion

Epidermoid cyst is a rare slow growing extra axial tumour comprising for about 1.8 % of all intracranial tumours [1,2]. They are benign congenital inclusion cyst, believed to arise between third and fifth week of gestation. During the division of neuroectodermal and cutaneous ectoderm in the intrauterine development, ectodermal cell rests get sequestered within the neural groove [3]. These misplaced ectodermal cells can develop into epidermoid cyst, dermoid cyst, or dermal sinus [1]. Dermoid cysts prefer midline location whereas the epidermoid cysts can occur throughout the neuroaxis, most commonly in the cerebellopontine angle and parasellar region. Other rarely reported cases are pineal region epidermoid, intra axial and intra diploic epidermoid cysts [1,4].

Growth of epidermoid occurs at a linear rate as that of normal skin. These tumors tend to expand within the subarachnoid spaces and encase the major arteries and the perforators coursing within the cisterns. Owing to its slow growth rate, these tumors attain relatively large size before the patient becomes symptomatic [5].

Variable imaging appearances of the epidermoid cyst are seen owing to the amount of cholesterol content, protein content and hemorrhage within the cyst. Typically, an epidermoid cyst shows CSF density on CT and signal intensities broadly similar to CSF on T1 and T2 weighted MR sequences with heterogeneous signal suppression on FLAIR. However, few pathologically verified epidermoid cysts showing hyper density on CT, increased signal intensity on T1 and decreased signal intensity on T2-weighted images, as in our case, are reported in the literature [5,6]. These are termed as Atypical or white epidermoid cyst. White epidermoids account for only 3% among the epidermoid cysts [7]. These atypical epidermoid cysts appear hyperdense on CT. Hence, they were termed as dense epidermoid cysts by Braun IF, et al. (1977) [8].

Saponification of cyst debris leading to dystrophic calcification is likely to be the most common process causing hyperdensity in CT according to Braun IF, et al. (1977) [8]. Thickened capsule with dense calcification and increased proteinaceous cystic content due to liquefactive necrosis were also the other theories postulated as the causes for its dense nature. Liquefactive necrosis occurs as the result of immediate inflammatory reactions to the recurrent leakage of cystic contents. These recurrent inflammatory reactions also cause perilesional vascular granulation tissue that compresses the blood vessels around the lesion leading to spontaneous haemorrhage. Aseptic meningitis is another complication of such cysts due to its spontaneous rupture into the CSF fluid [9].

Hyperintensity of atypical epidermoid cyst on T1 weighted sequences is either due to its high protein or lipid content or rarely due to intracystic haemorrhage as [10]. Reduced signal on T2 weighted sequence with 'shading sign' is due to either blood products or proteinaceous debris within the cyst. Shading sign is the T2 shortening of spontaneous hyperintense T1 weighted cysts, originally described in endometriomas, is also a feature of white epidermoid cyst [9]. In our case, intracystic hemorrhage and cholesterol granulomas accounted for T1 hyperintensity and T2 hypo intensity.

Typically, an epidermoid cyst shows true restricted diffusion with no surrounding edema and no internal enhancement on post contrast study. However, peripheral rim enhancement has been reported in few cases. Atypical epidermoid cyst, as in our case do not show restricted diffusion. Liquid nature of the cystic content and xanthochromic fluid within the cyst due to micro bleeding or degrative changes may explain this nature in these epidermoid cysts [9].

The differential diagnosis for white epidermoid cyst in the cerebellopontine angle is cystic schwannoma, cystic meningioma, arachnoid cyst and neurogenetic cyst.

Internal auditory canal extension and strong contrast enhancement of the cyst wall are few differentiating features found in schwannoma. Also, pure cystic nature of vestibular schwannoma is extremely rare. This lesion can be differentiated from cystic meningioma as the latter shows broad dural base attachment, dural tail and strongly enhancing solid component.

Arachnoid cyst has better defined margins and cause bone thinning. Arachnoid cyst shows total fluid suppression in FLAIR sequence and lack of diffusion restriction [10].

The main point of the surgical treatment of the atypical epidermoid cyst, as like classical one, is radical excision with its capsule and microsurgical evacuation of cystic contents. However due to the localization, fragments of the adherent capsule to the deep veins of this region are left in situ to avoid risk. Choice of operative approach depends upon the extent of the cyst [9].

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