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A Typical Intracranial Epidermoid Cyst: A Diagnostic Journey Through MRI

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Abstract: Intracranial epidermoid cysts are rare, benign tumors constituting about 1% of all intracranial tumors, typically presenting in adults aged 20 - 40. Atypical variants are even rarer. This report details two cases of atypical intracranial epidermoid cysts. The first case involves a 43 - year - old female with headaches and ataxia. MRI revealed a T2 hyperintense cystic lesion in the left cerebello medullary cistern, suggesting an atypical white epidermoid cyst. The second case concerns a 69 - year - old female with persistent headaches, neurological deficits, and a seizure. MRI showed a large, irregular extra - axial lesion in the superior cerebellar cistern extending to the right occipito - temporal region, indicative of an atypical epidermoid cyst. These cysts cause symptoms due to mass effect, with headaches being the most common. Diagnosis relies on MRI, showing characteristic T2 hyperintensity and lack of restricted diffusion on DWI. Despite their benign nature, epidermoid cysts can cause significant neurological issues. This article revolves around the findings of both cases with their imaging appearance on various MRI sequences.

Keywords: Atypical Intracranial Epidermoid Cyst, MRI, Benign slow growth, Mass effect, Headache, White epidermoid cyst, Choline Peak on MR Spectroscopy

1. Introduction

Epidermoid cysts are rare, benign growths that occur within the skull, but outside the brain parenchyma, which constitute approximately 1% of all intracranial tumors. [1, 2, 3] Embryologically, they originate from ectodermal inclusions that occur during neural tube closure between the third and fifth weeks of embryogenesis. [4] Although primarily congenital, epidermoid cysts grow very slowly, often taking many years to become noticeable. Typically, patients are between 20 and 40 years old when they present. [5] Males may have a higher prevalence. [6, 7] Symptoms are due to mass effect on adjacent structures. Atypical epidermoid cysts are even rarer, with intra - axial epidermoid cysts making up less than 1.5% of all intracranial epidermoid cysts. [8]

Here we report 2 cases of atypical intracranial epidermoid cysts.

2. Case Reports

Case 1:

History - A 43 - year - old female presented with on and off headache for the past six months and one month history of ataxia.

Report - MRI study of brain showed a well - defined extra - axial T2 hyperintense cystic lesion measuring $\sim 15.0 \times 10.0 \times 15.0 \text{ mm}$ in the left cerebello - medullary cistern extending till the level of foramen magnum. The lesion showed complete suppression on T2 FLAIR images. The lesion appeared mildly heterogenous with signal nearly isointense to brainstem on T1 weighted images. No definite restricted diffusion was noted on DWI. The lesion was seen exerting significant mass effect on medulla pushing it towards the right side. It was also seen pushing the left vertebral artery posteriorly and medially. Epidermoid cyst with atypical features (possible white epidermoid) is the likely diagnosis.

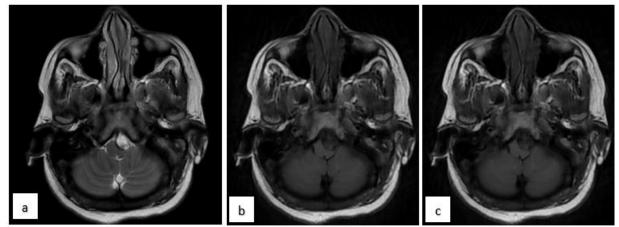


Figure 1: MRI axial sections of brain showing (a) well - defined T2 hyperintense cystic lesion in left cerebello - medullary cistern; (b) lesion appears mildly heterogenous with signal nearly isointense to brainstem on T1 weighted images; & (c) lesion showed complete suppression on T2 FLAIR images.

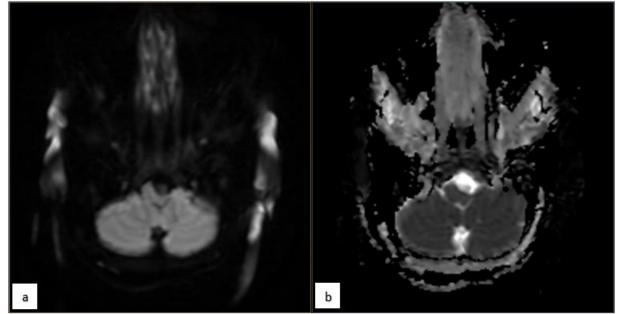


Figure 2: MRI axial sections of brain - (a) diffusion - weighted imaging (DWI) & (b) apparent diffusion coefficient (ADC) images. No definite restricted diffusion was noted on DWI.

Case 2:

History - A 69 - year - old female presented with persistent, gradually worsening headache for two months, focal neurological deficits and one episode of seizure, after which she presented to the OPD.

Report - MRI study of brain showed a well - defined irregular shaped extra axial lesion with smooth lobulated margins showing predominant T1 hypointense & T2/FLAIR hyperintense signal measuring ~ 7.6 x 5.8 x 5.0 cm (CC x AP x ML) in the region of superior cerebellar cistern, right ambient cistern & extending upto parasagittal aspect of right occipito - temporal region. Multiple tiny cystic areas were noted within the lesion. No diffusion restriction / GRE hypointense blooming was noted within the lesion. Post contrast images showed mild patchy areas of enhancement. On MR spectroscopy, the lesion showed choline peak with absent / severely depressed NAA peak. The lesion caused mass effect over midbrain, cerebral peduncle, right occipito temporal lobes, 3rd ventricle, temporal & occipital horns and posterior body of right lateral ventricle which in turn was causing cerebellar tonsillar herniation due to mass effect.

Possibilities include - Atypical epidermoid – More likely.

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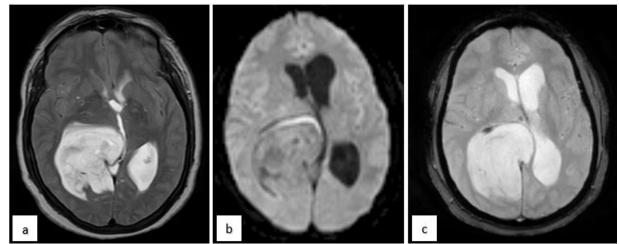


Figure 3: MRI axial sections of brain showing (a) well - defined irregular shaped lesion with smooth lobulated margins showing predominant T2 hyperintense signal; (b) DWI shows no diffusion restriction; & (c) gradient echo (GRE) - no hypointense blooming noted within the lesion.

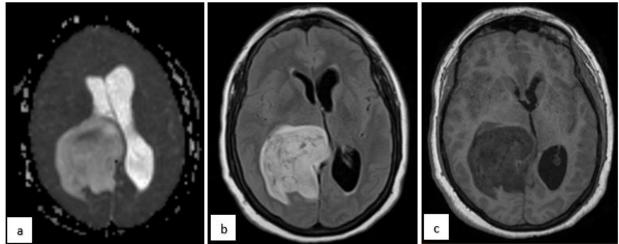


Figure 4: MRI axial sections of brain showing (a) ADC; (b) lesion shows T2/FLAIR hyperintensities; & (c) predominantly hypointense on T1 weighted images

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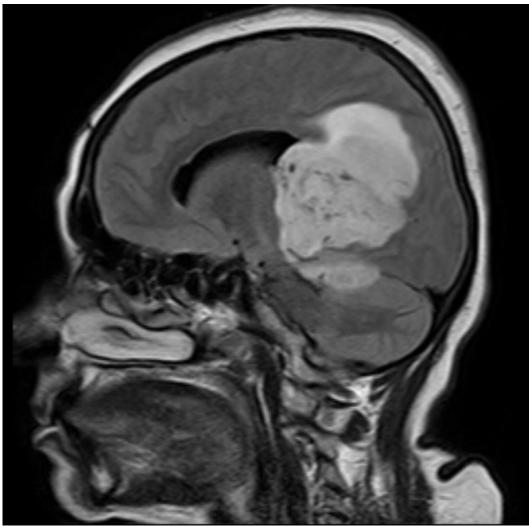


Figure 5: MRI sagittal section of brain T2/FLAIR showing the extent of lesion from the superior cerebellar cistern, right ambient cistern; extending upto parasagittal aspect of right occipito - temporal region.

3. Discussion

Patients present later in life because of the slow growing mass. The signs and symptoms of epidermoid cysts are due to a gradual mass effect and typically include headaches, which are the most common symptom, as well as cranial nerve deficits, cerebellar symptoms, seizures, and increased intracranial pressure. Although recurrent aseptic meningitis is uncommon, it is a recognized complication, similar to the less common dermoid cyst. [9]

Diagnosis of intracranial epidermoid cysts is primarily reliant on imaging modalities, with MRI being the most definitive. The characteristic imaging features on MRI include hyperintensity on T2 - weighted images and isointensity or slight hyperintensity on T1 - weighted images. Epidermoid cysts typically do not show enhancement post - contrast administration, which helps differentiate them from other cystic lesions such as arachnoid cysts and schwannomas. Additionally, the lack of restricted diffusion on diffusion weighted imaging (DWI) is a distinguishing feature. The heterogeneity observed in the presented cases is indicative of atypical variants, which further complicates the diagnostic process. [10] The mass effect observed in both cases underscores the clinical significance of these cysts despite their benign histological nature. In the first case, the mass effect on the medulla and the displacement of the left vertebral artery exemplifies the potential for significant neurological compromise. Similarly, in the second case, the extensive involvement of the superior cerebellar cistern and the resultant cerebellar tonsillar herniation highlight the potential for severe clinical outcomes.

Management of intracranial epidermoid cysts typically involves surgical resection, especially when symptomatic. Complete excision is ideal, however, the tendency of these cysts to adhere to surrounding neurovascular structures often limits the extent of resection, thereby increasing the risk of recurrence and postoperative complications.

Atypical epidermoid cysts, as illustrated in the presented cases, require a nuanced approach to diagnosis because of their rarity and atypical presentation. The differential diagnosis must consider other cystic lesions, including schwannomas and arachnoid cysts, particularly when imaging features are not classic. In such scenarios, MR spectroscopy can provide additional diagnostic clues, as seen in the second case where a choline peak was noted.

4. Conclusion

While intracranial epidermoid cysts are rare and typically benign, they pose significant challenges. Early recognition through detailed imaging studies is crucial for appropriate management and to mitigate potential neurological deficits. Future studies should focus on refining diagnostic criteria and exploring less invasive treatment modalities to improve patient outcomes.

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Authors' Contribution

Dr. Shalmol Thomas: Writing, Investigation, Analysis, Review and Editing.

Dr. Piyoosh Priyadarshee: Writing, Investigation, Analysis, Review and Editing

Dr. Baskar A.: Conceptualization, Supervision, Methodology, Resources, Data Collection, Writing and Formal analysis.

All authors have read and agreed to submit the manuscript. Informed consent, proper written consent obtained from the patient.

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Conflict of Interest

The authors declare that there is no conflict of interests.

Data snd Materials Availability

All data sets collected during this study are available upon reasonable request from the corresponding author.

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