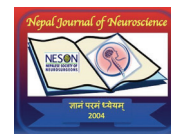


Malignant Epidermoid Cyst: MRI Evaluation and Review of Literature

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Abstract

Epidermoid cysts are commonly encountered intracranial neoplastic masses in clinical Radiology practice. They are mostly benign but may undergo malignant transformation or may present as de novo malignant epidermoid tumor, a rare but ominous phenomenon associated with poor prognosis. Malignant epidermoids may masquerade as glial tumours, ependymal neoplasms, or choroid plexus masses, and are prone to be misdiagnosed. We describe a case of malignant epidermoid cyst or tumor (de novo) in a young adult who presented with the complaints of intermittent headaches which had increased in frequency and severity.

Key words: epidermoid cysts, malignant epidermoid cysts, brain tumours, MRI

Introduction

Epidermoid cysts, derived from embryonal ectodermal elements, are inclusion cysts and comprise 1% of all the primary intracranial neoplasms.¹ They are most commonly located at the cerebellopontine angle cisterns, followed by middle cranial fossa, parasellar region, and the ventricles. They tend to insinuate between the brain structures.² On magnetic resonance imaging (MRI), they appear hyperintense as compared to cerebrospinal fluid (CSF) on both T1 and T2 weighted images and rarely show thin peripheral post-contrast enhancement, while a pronounced heterogeneous patchy enhancement is the characteristic which suggests malignant transformation of the epidermoid cyst and portends a worse prognosis.³

This case demonstrated the characteristic MRI (DWI/ADC and post contrast T1W) imaging findings corresponding to the intracranial malignant transformed

epidermoid cyst infiltrating and encasing the adjacent structures.

Case report

A 27 year old man presented with complaints of intermittent headache with recent increase in frequency and severity. Physical examination and routine blood investigations showed no significant abnormality.

Contrast enhanced MRI of the brain was performed which showed an infiltrative pattern of T1 hypointense (Fig 1) and T2 hyperintense lesion (Fig 3) with partial suppression on FLAIR images (Fig 2) is seen in the prepontine as well as perimesencephalic cisterns (Fig 1). The lesion is extending from floor of lateral ventricle superiorly to foramen magnum inferiorly and upto fourth ventricle posteriorly (Fig 3 and 4). The lesion is seen infiltrating into the right temporal lobe (Fig 3) as well as temporal horn of right lateral ventricle (Fig 2). The lesion is seen closely abutting and displacing midbrain and pons (Fig 1). Contralateral midline shift of 8mm towards left is seen (Fig 2). The lesion is noted to encase basilar artery, bilateral PCA, M1 and M2 segments of right MCA (Fig 4). Encasement of right trigeminal, facial and vestibulocochlear nerves are seen. The lesion shows patchy restriction on DWI (Fig 6) and few areas of nodular and ring enhancement on post contrast images (Fig 5). No obvious meningeal enhancement is seen.

Due to the marked involvement of eloquent and surgically complex structures by the mass, the patient was referred to a higher centre where it was operated. The histopathology report showed multilayered keratin cells with pleomorphic nuclei which was consistent our imaging diagnosis.

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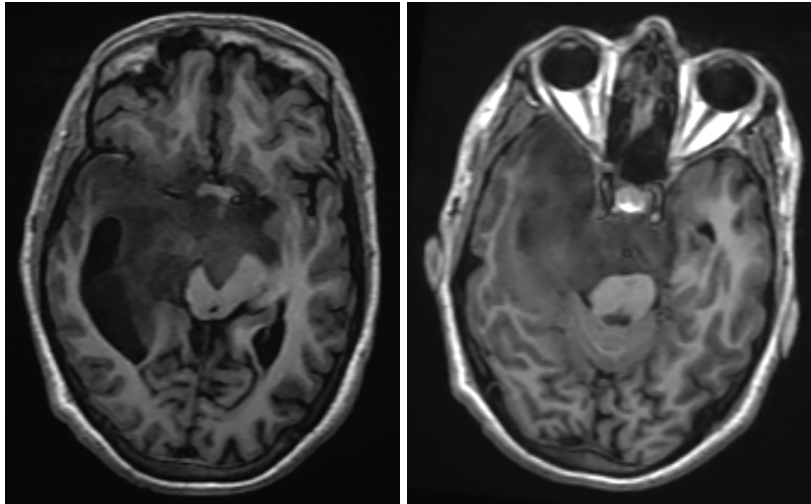


Figure 1: T1 hypointense lesion seen in perimesencephalic cistern. Pons and midbrain are seen closely abutted and displaced by the lesion.

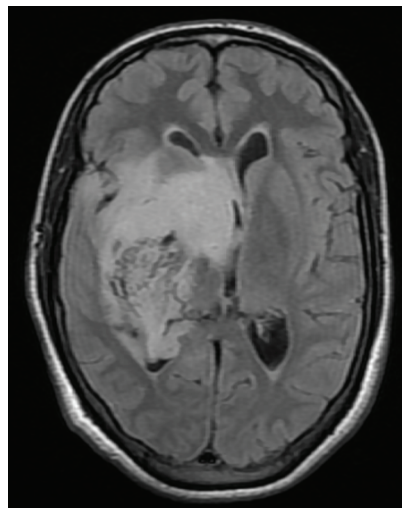


Figure 2: T2 FLAIR image shows partial suppression and infiltration in temporal horn of right ventricle is seen. Significant midline shift towards left is seen.

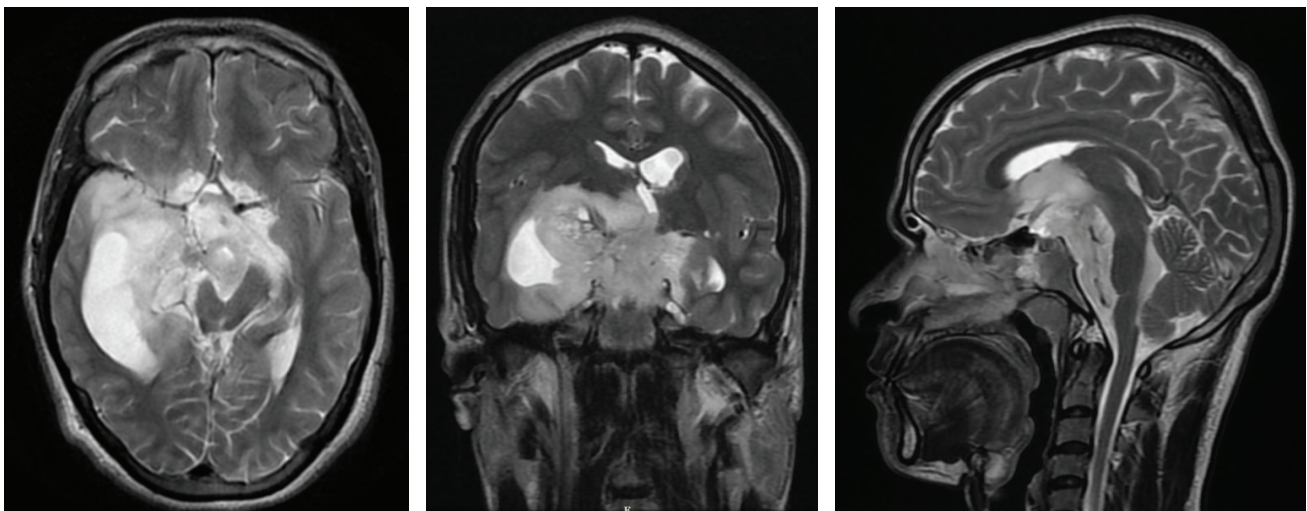


Figure 3: Axial T2 and Coronal T2 weighted images show hyperintense lesion infiltrating into right temporal lobe. Sagittal T2 weighted image showed extension from floor of lateral ventricle superiorly to foramen magnum inferiorly and fourth ventricle posteriorly.

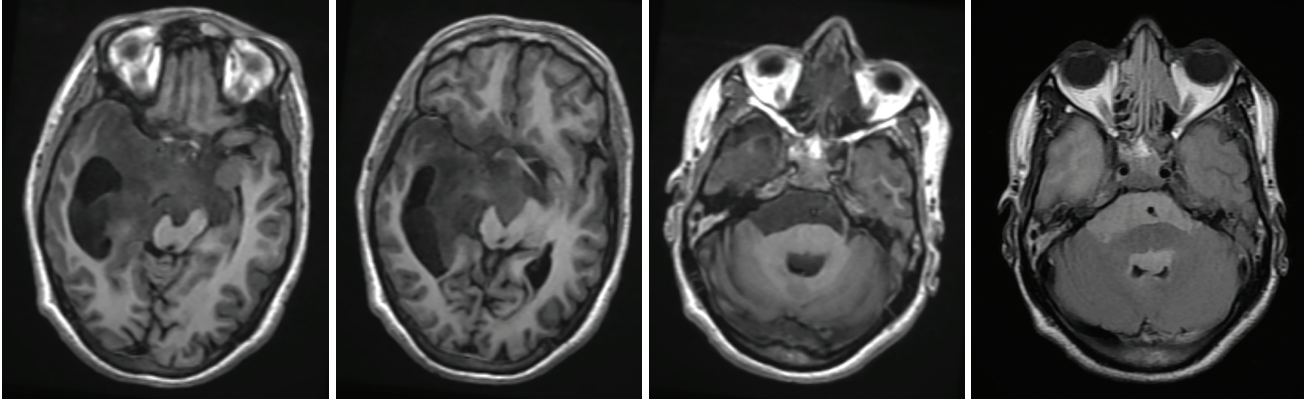


Figure 4: The lesion is seen encasing right PCA, right MCA, basilar artery. Extension into fourth ventricle is also seen on FLAIR images and is likely from foramen magendie and luschkka.

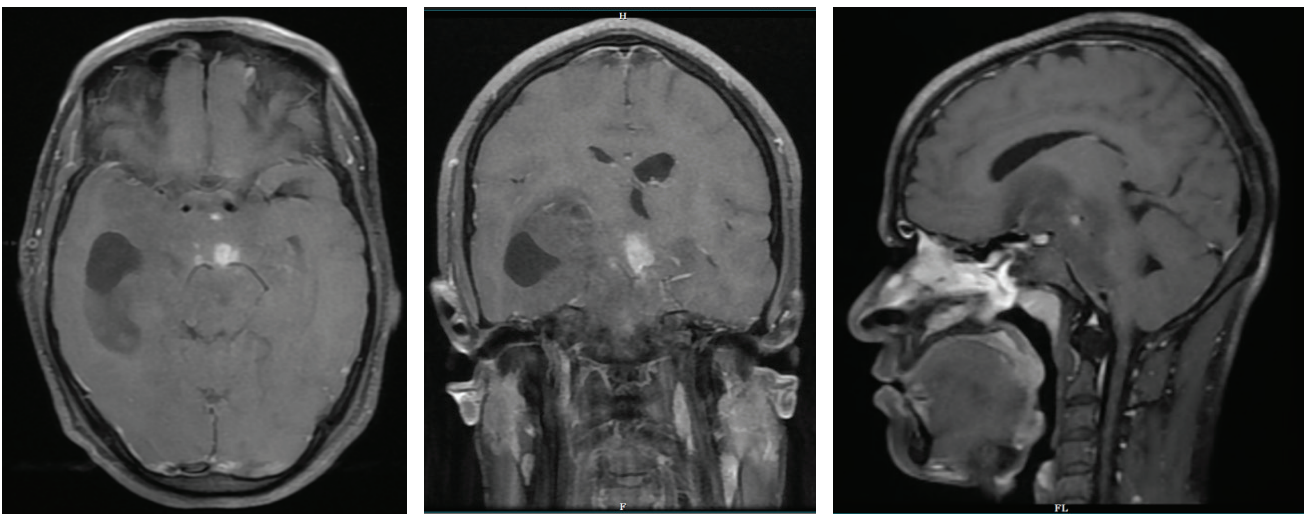


Figure 5: Few areas of ring and nodular contrast enhancement on Axial, coronal and sagittal T1 FS+ Contrast images.

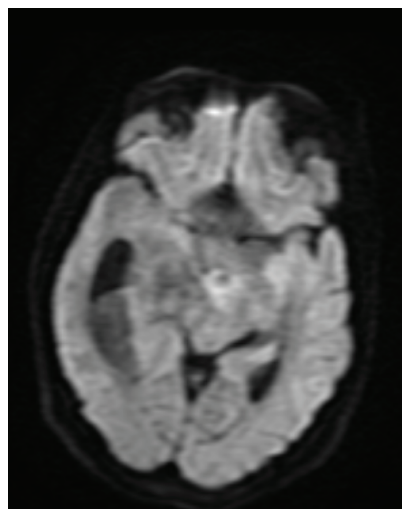


Figure 6: Patchy areas of diffusion restriction noted on DWI image.

Discussion

Majority of the epidermoid cysts are intradural and extraaxial in location and predominantly involve the

cerebellopontine angle cistern. It is thought to arise from inclusion of ectodermal elements.⁴

Grossly, epidermoid cysts appear shiny and bear resemblance to mother of pearl appearance.⁵

Microscopically, the outer fibrous layer of the cyst wall is formed by the stratified squamous epithelium. The contents of the cyst include keratinaceous debris and cholesterol.⁶

CT imaging features of epidermoid cysts are hyperdense, lobulated mass with no post contrast enhancement.⁷ Hyperdensity is attributed to high proteinaceous content, saponification of keratinized debris, lipid debris, and leukocytes.⁸

On MR imaging, the cyst usually follows CSF signal intensity and shows diffusion restriction on DWI sequence. A “white epidermoid” is a variant of epidermoid cyst which has high protein content and is viscous, due to which it appears hyperintense on T1WI and hypointense on T2WI.⁹

There can be malignant transformation of the cyst or denovo primary squamous cell carcinoma may at the same location.¹⁰ Rapid progression of the signs and symptoms holds high clinical suspicion for malignant transformation of the epidermoid cyst.¹¹ The exact pathological mechanism resulting in this transformation is not clear. However, it is proposed that chronic inflammatory response to cyst rupture and incomplete resection of the cyst might be the causative factors.¹²

Characteristic imaging findings of malignant transformation of epidermoid cyst include enhancement in some part of the lesion in nodular pattern or leptomeningeal enhancement on CT or MRI. However, it may be found spuriously if there is leakage of cyst contents into surrounding area, causing intense local inflammatory response causing foreign body giant cell reaction or if into subarachnoid space, chemical meningitis.¹³ Therefore, differentiation between these two entities is difficult due to similar appearing imaging findings.

The carcinomatous part of the cyst shows lower signal intensity owing to its central necrotic component as compared to the benign part which shows high signal intensity on DWI sequence. But the ADC value of both correspond to each other causing T2 shine through effect.¹⁴

There is poor prognosis if malignant transformation of the cyst occurs. The mortality rate is as high as 75%.¹³ Surgical removal followed by the post-operative radiotherapy is the recommended therapeutic procedure.³

This study presents the imaging features corresponding to the malignant transformation of the epidermoid cyst, which is a rare phenomenon.

Conclusion

De novo malignant epidermoid cysts or its malignant transformation is a rare occurrence we find in clinical radiology practice. The case presented to us had imaging features characteristic to the malignant epidermoid cyst. Due to its aggressively infiltrating nature, it was referred to

higher centre where it got operated and the Histopathology report turned out to be the same.

Conflict of interest: None

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