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Case Report

Rare Frontal Intraparenchymal Epidermoid Cyst: Case report

Jihane Saidy1*; Said Hilmani1,2; Abdelhakim Lakhdar1,2

¹Departement of Neurosurgery, Hassan 2 University Casablanca, Morocco.

²Laboratory of Research on Nervous and Neurosensory Disease and Disability, Faculty of Medicine and Pharmacy, Hassan 2 University Casablanca, Morocco.

*Corresponding Author: Jihane Saidy

Department of Neurosurgery, Hassan 2 University

Casablanca, Morocco.

Email: jihanesaidy@gmail.com

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Abstract...

Epidermoid cysts are benign slow growing extra-axial tumours that occur most commonly in the cerebellopontine angle, parasellar region, and subarachnoid spaces of the basal cisterns while their occurrences in intra-axial locations are exceptionally rare. We present the clinical, imaging, and pathological findings in a patient with atypical epidermoid cysts.

A 36-year-old presented with a history of seizures for 7 months. The patient had a normal physical exam. CT of the brain that revealed a low-density frontal lesion in continuity with another interhemispheric lesion. MRI showed an intra-axial mass with a low T1 weighted images signal and hyper signal mass on the diffusion MRI. A left frontal craniotomy and incision into the cortex revealed a cystic lesion filled with keratinized debris. The interhemispheric mass has a yellow content. The histopathological exam of the entire mass concludes to epidermoid cyst. The pathogenesis of intraparenchymal epidermoid cyst remain unclear, may be that congenital sequestration of ectodermal elements can involve the parenchyma through vessels of cisternal areas.

Key words: Epidermoid cyst; Brain; MRI; Pearl like tumor.

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Introduction

Epidermoid cysts are benign, extra-axial rare inclusion lesions accounting for approximately 0.2%–1.8% of all brain tumors. They characteristically spread along anatomical cleavage planes, progressively filling the subarachnoid spaces [1,2]. However, intraparenchymal epidermoid cysts (IECs) are extremely rare lesions. Only few cases of IECs have been previously reported. We present the clinical, imaging, and pathological findings in a patient with atypical epidermoid cysts.

Case report

A36-year-old man, presented with a history of repeated generalized tonic-clonic seizures for 7 months. The patient had a normal physical exam, including a complete normal neurological and cranial nerve exam. CT-scan of the brain revealed a low-density frontal lesion in continuity with another interhemispheric lesion (Figure 1). Cerebral MRI showed an intra-axial mass with a low T1 weighted images signal and hyper signal mass on the diffusion MRI (Figure 2). The patient underwent a left frontal craniotomy for tumor resection. After opening the dura, no abnormalities were seen on the cortical surface, but incision into the cortex revealed a cystic lesion, filled with keratinized debris. The interhemispheric mass has a yellow content. Despite weak adhesion to the cerebral falx and pericallosal artery, the two capsules were totally removed (Figure 3). The histopathological exam concludes to epidermoid cyst. The patient recovered well from surgery and the anticonvulsant has been administrated to prevent seizures.

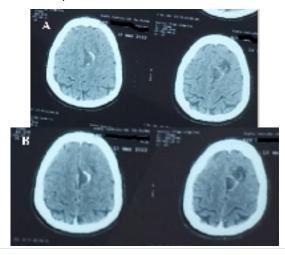


Figure 1: A: Axial CT scans showing a left frontal lobe mass in continuity with another interhemispheric mass surrounded with calcifications. **B**: No enhancement of the mass following contrast administration

Discussion

Epidermoid cysts (ECs) are rare and congenital lesions that develop from ectodermal remnants during neuroembryogenesis. They are composed of an outer cystic capsule, a keratinized stratified squamous epithelial layer, and an inner cystic content that include tissue debris, keratin, water, and solid cholesterol. Cerebellopontine angle, petrous apex, suprasellar region, and the fourth ventricle are the usual locations [3]. Epidermoid cysts that are exclusively intraparenchymal are very rare, less than 1% Clinical manifestations of the IECs are related to the volumes and locations of the tumors [2,4]. If the origin of classical loca-

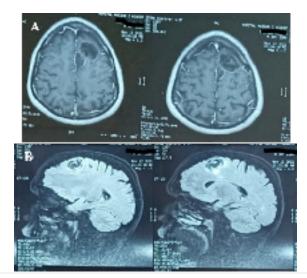


Figure 2: A: Axial T1-weighted MRI with contrast showing mixed signal with no enhancement **B**: Sagittal diffusion MRI showing a partial hyperintensity lesion.

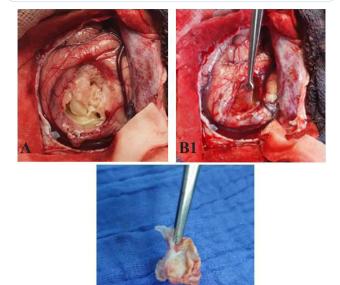


Figure 3: per operative images showing **A:** The cyst contents, **B1, B2:** Dissection and the complete removal of the capsule

tion has largely described, the pathogenesis of purely intracerebral epidermoids cyst is not clear. In our opinion, as ECs almost always grow along cisternal areas of the brain and the midline, we think that congenital sequestration of ectodermal elements can involve the parenchyma through vessels which explain adhesion to the dura and pericallosal artery in our patient.

The most commun symptoms are headaches, cranial nerve palsy, cerebellar symptoms, aseptic meningitis may also occur but not that often. The clinical particularity of intraparenchymatal epidermoid cyst is seizures and neurological deficit [5]. It can be difficult to diagnose as they often do not have classic radiologic findings, and share many similar radiologic features to other tumors [1]. CT scan usually shows a low-density area without contrast enhancement and calcification may be present as showed in our patient. MRI plays an important role in the diagnosis of intraparenchymal epidermoid cysts. They are usu-

ally hypointense on T1-weighted sequences and hyperintense on T2-weighted sequences [6,7]. Surgical excision remains the mainstay of treatment [8,9]. However, radical removal of the tumor capsule can be very difficult because of the strong adherence of the tumor capsule to surrounding brain parenchyma, particularly in eloquent areas. The tumor has a pearl-like appearance which is the result of desquamation of keratin and cholesterol within its wall [10].

Due to the cystic nature of the lesion, greater care should be taken to differential diagnosis of other intraparenchymal cystic lesions and ensure that the cyst does not rupture thereby leading to chemical meningitis [11].

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