Case Report

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A swelling on the fore head- the tip of the iceberg

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ABSTRACT

An epidermoid cyst is a benign cyst usually found on the skin, developing out of ectodermal tissue. These are relatively uncommon benign cysts or tumors to arise over the surface of the brain. They are mostly intra dural and few percentages are located in the diploic space. We are reporting a case of 51 years old lady presenting with a swelling over forehead which was diagnosed to be an intradiploic epidermoid cyst of the frontal bone with an intracranial extension which is an unusual presentation causing clinical dilemma. Radiological findings led to a correct diagnosis and with complete removal patient is doing well.

Keywords: Epidermoid cyst, Brain tumour, Intra diploic cyst, Frontal bone, Skull tumour

INTRODUCTION

Epidermoid cysts are benign congenital lesions constituting approximately 1% of intracranial tumours, 75% of these cysts are located intradurally and 25% within the diploic spaces. Intradiploic epidermoids are relatively rare insidious tumours that occur in all bones of the calvarium, temporal and sphenoid bones, paranasal sinuses, and maxilla. Treatment is surgical removal and is usually curative. We present a 51 year old lady with intra diploic epidermoid cyst of the frontal bone protruding from the fore head. This unique and rare site presentation of this cyst, as a simple looking forehead swelling causing clinical dilemma is put forward in this case report.

CASE REPORT

A 50 years old lady presented with a swelling on the forehead of five months duration which gradually increased in size with minimal discharge from it (Figure 1). Patient complained of spontaneous bleeding from right nostril with frequent headache. No history of vomiting, neck stiffness, diplopia, hyperacusis or gait ataxia was present. Physical examination elicited a tender soft swelling measuring 2x2cm in fore head midline, 1cm

below the hair-line discharging pus. Margins were well felt with bony defect on the left side. It was a nonpulsatile, nonreducible, noncompressible swelling with no expansile cough impulse.



Figure 1: Swelling on the fore head.

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Routine blood investigations were within normal limits. Computed tomography revealed a well circumscribed extracerebral hypodense intradiploic tumour with sclerotic rim involving the frontal bone, and compressing bilateral frontal lobes (Figure 2a). On MRI study, the mass demonstrated iso-hypointense signal on T1W images, homogenously high signal intensity on T2-weighted images, and mild peripheral post-contrast enhancement (Figure 2b). Patient underwent craniotomy and exploration of the invasive lesion under general anesthesia.



Figure 2a: Axial CT showing a well circumscribed extracerebral hypodense intradiploic tumor with sclerotic rim involving the frontal bone, and compressing bilateral frontal lobes.



Figure 2b: Coronal T2W image showing predominantly hyperintense signal (isointense to CSF) within the mass. Cortical discontinuity is noted along its right infero-lateral margin.

Intra operative finding showed numerous defects in frontal bone, with pearly material lesion present extradurally but adherent to dura lesion invaginating between the two frontal lobes and compressing and separating them (Figure 3a). The capsule had a pearly sheen (Figure 3b).



Figure 3a: Defects in frontal bone with pearly material lesion present extradural.

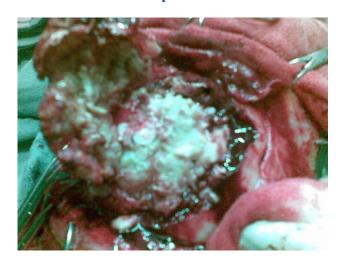


Figure 3b: The capsule with a pearly sheen.

Sections studied from both the specimens bone and dura showed abundant keratin flakes and cholesterol clefts surrounded by epitheloid macrophages and multinucleated foreign bodies type of giant cells and infiltrated by abundant neutrophils and few lymphocytes suggestive of infected epidermoid cyst (Figure 4a & 4b). Post-operative period was uneventful and patient recovered well. On six months follow up patient is doing well.

DISCUSSION

An epidermoid cyst is typically a benign brain tumour that arises from abnormal cells being left in the nervous system during development, accounting for

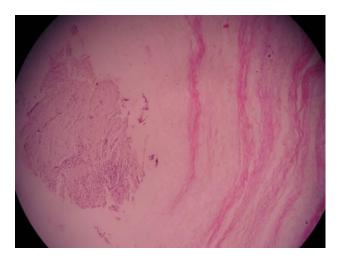


Figure 4a: Microphotograph showing laminated keratinous material, areas of necrosis and dystrophic calcification (Haematoxylin & Eosin x10X).

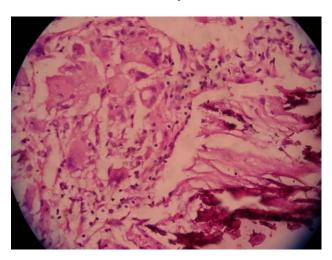


Figure 4b: Higher power magnification of the same case showing foreign body giant cells, mixed inflammatory cells and dystrophic calcification (Haematoxylin & Eosin x45X).

approximately 1% of intra cranial tumour. Ectodermal cells that normally form skin are trapped in the developing nervous system or stay within the cranial bones during embryonic development, creating a welldefined cyst with a pearly coloured capsule which is lined inside with ectodermal cells.1 The cysts consists of desquamated epithelial cells with concentric layers of keratin and cholesterols and do not contain hair or other dermal elements which differentiate it from the dermoid tumor.² Although these lesions are congenital, patients are usually not symptomatic until they are aged 20-40 years, because they take a long time to expand to a size that causes symptoms.^{3,4} Bony epidermoid cysts which nearly form 25 percent of epidermoid cyst are mainly located in the skull, although other locations, including the mandible, maxilla, temporomandibular joint, distal phalanges, tibia and femur, have been described.⁵

The presentation and clinical symptoms of these tumours vary depending on size and location. These are rare, slow growing tumours that can present in many different ways, including a painless lump, tenderness, headache and rarely with focal neurological signs. These symptoms and others will vary from patient to patient. Malignant transformation, intracranial hypertension, infection, seizures and focal neurologic signs have been described in patients with large cysts. ⁶

A CT scan or MRI scan is needed to detect the tumor. Characteristically in CT scan, there is a well-defined lytic intradiploic lesion with decreased attenuation and areas of calcification, expanding and eroding the inner and outer tables of the skulls with or without sclerotic margins. MRI findings are reliable in diagnosis. They are usually lesions displaying a hypointense signal on T1weighted sequences and a heterogeneous hyperintense signal on T2-weighted sequences, as was it in our case too. Other sequences Proton density-weighted, fluidattenuated inversion recovery (FLAIR) images, magnetic resonance diffusion-weighted imaging (MR DWI) sequences have been described to better characterize these tumours and thereby eliminate diagnostic uncertainty.^{7,8}

Treatment usually consists of a surgical resection of the tumor. The cyst lining can be very adherent to the brain structures hence a complete cyst removal may not be possible in many patients. If any of the cyst wall was not removed, they can continue to enlarge slowly and recur after surgery hence follow-up MRIs is necessary for many years to monitor for recurrence. Chemotherapy and radiotherapy are generally not used for treating epidermoid cysts. 10

CONCLUSION

Intradiploic epidermoid cysts of the skull are rare, slow-growing, benign tumours of the skull which are composed of epidermoid cell debris. This case illustrates a common presentation of a relatively uncommon tumour of the cranium. Radiology imaging of CT scan and MRI are needed to diagnose these lesions. The prognosis is generally good with appropriate surgical intervention.

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