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RESEARCH ARTICLE

INTRADIPLOIC EPIDERMOID CYST– A RARE LOCATION

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ABSTRACT

Epidermoid cysts are among the rare benign tumors contributing to only 0.3% to 1.8% of all intracranial tumors. This tumour is located most commonly in the region of cerebellopontine angle cistern, the parasellar region, and diploic space being the rarest location. In this case report of a4yr old female child who had history of epidermal cyst located in left parietal bone for which she was operated twice and recurrence of the cyst occurred again with sinus tract formation between two layer of the skull. We present pre-operative and postoperative radiological presentation, clinic-pathological correlation and surgical outcome. During 8 months of follow-up, the patient remained in good condition without recurrence and complications.

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INTRODUCTION

Epidermoid cyst originates from ectodermal cell present within the cranial bone during embroyogenesis. Bony epidermoid cysts are mainly develops in the skull, other location includes mandible, maxilla, temporomandibularjoint, distal phalanges, tibia and femur have been reported .In a skull, this tumour is located most commonly in the region of cerebellopontine angle cistern, the parasellar region, and diploic space being the rarest location (1) Epidermoid cysts are among the rare slow growing benign tumors contributing to only 0.3% to 1.8% of all intracranial tumors.

Case History: A 4 year old female child presented to our departmenthad past history of the excision of epidermoid cyst done 3 yrs back at the age of one year. Later cyst recurred after 6 month of surgery for which the patient was operated again .Now the patient has presented with bony defect in the left parietal bone with watery discharging sinus. There was no associated history convulsion and fever and no focal neurological sign on examination

Pre-operative presentation: Patient had underwent CT BRAIN scan which was done by Siemens Somatom Emo 6 machine.

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CT scan revealed bony calvarial defect in left parietal bonewith destruction of the outer table of the skull and thinning of inner table of the skull (Figure No1). There was no evidence of intracranial extension of the cyst. The surgical planning was done and patient was operated again for the recurrent cyst and specimen containing two soft tissue bits with whitish coloured track were identified, largest measuring 1.5cm and smallest measuring 0.5cm in length which were sent for histopathological confirmation. The histopathological report of the specimen revealed cyst wall lined by stratified squamous epithelium and fibrous tissue showing a dense inflammation by lymphocytes, plasma cell and neutrophils. Subepithelial tissue showed presence of granulation tissue thus confirming the diagnosis of epidermoid cyst of the skull (Figure No:-2).

Post-operative presentation: Patient was followed up after 8 months of surgery and again underwent CT BRAIN which showed no defect in the left parietal bone with mild bony calvarial thickening was seen.

DISCUSSION

Epidermoid tumour are remnant of ectodermal cell which are divided into two types –primary /congenital lesion which is due to the incomplete cleavage of ectoderm in neural groove and secondary /acquired lesion having post traumatic etiology resulting invagination and implantation of epidermalcells on the bones (2). In skull congenital inclusion cyst are

histologically differentiated into dermoid and epidermoid. Epidermoid cyst have epidermal cell rich in cholesterol and kernatised stratified squamous cell as compared dermoid cyst which contains element of dermal cell layer (1). Epidermoid tumour account for 0.3 to 1.8 % of all intracranial lesions out of which 75 % are present intradurally and rest 25% arelocated intradiploic.

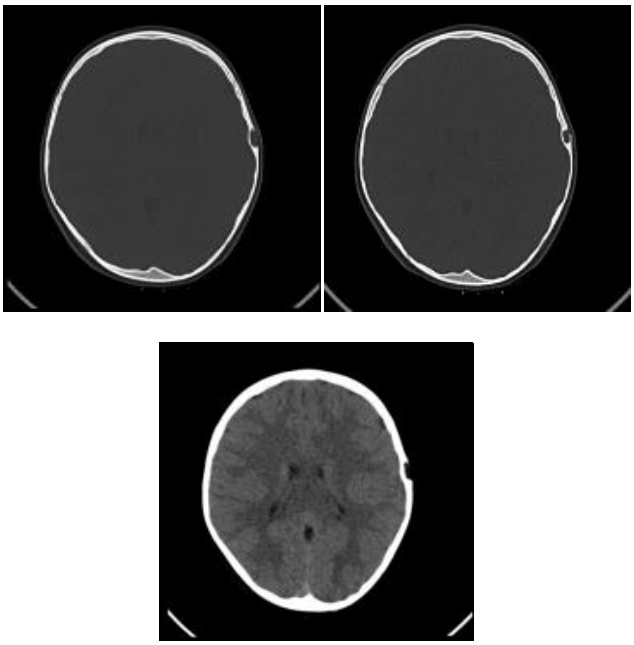

Figure 1. CT Brain shows defect in left parietal bone with complete destruction of outer table and thinning of inner table of skull

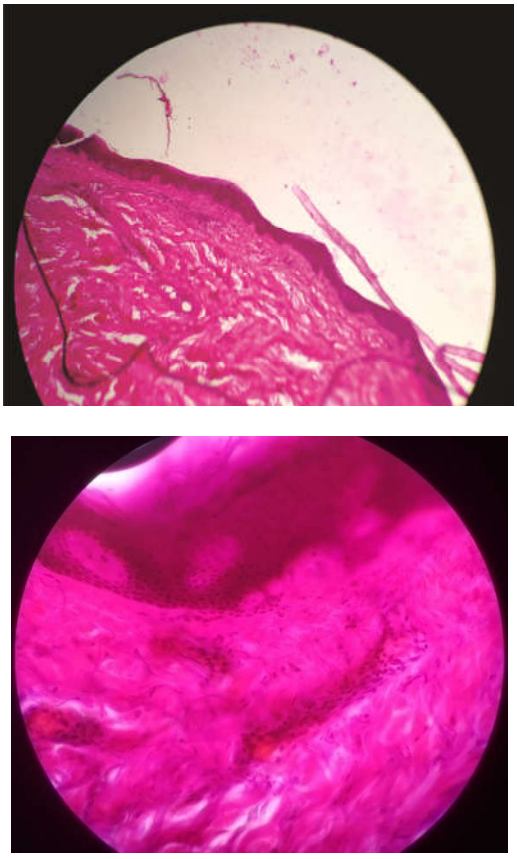

Figure 2. Histopathological specimen showing stratified squamous epithelium and fibrous tissue

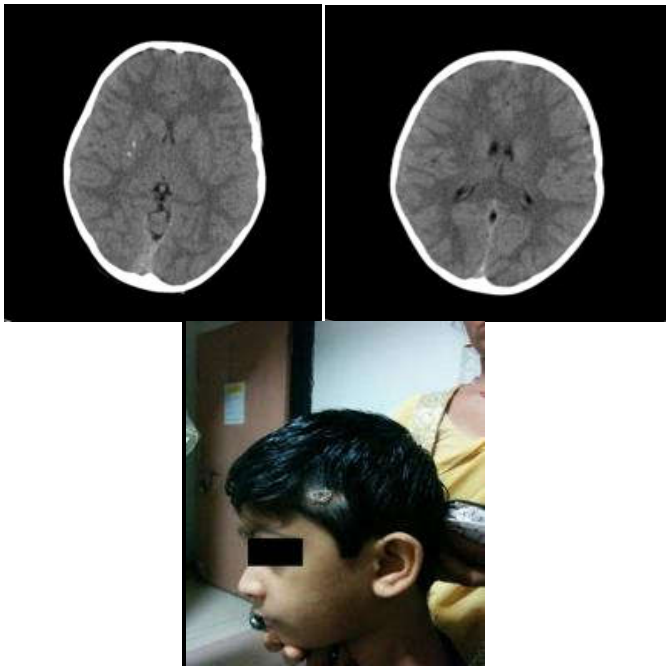

Figure 3. CT Brain follow up shows no defect in left parietal bone and no signs of recurrence after surgery

Intradiploic tumour arise due to embryological accident during early stage of development, between 3rd and 5th week resulting into incomplete cleavage of neural ectoderm from cutaneous ectoderm at the time of closure in the neural groove (1,3,5,6). Intradiploic epidermoid cyst mostly arise from frontal, parietal and occipital bone and involving both table (45%), outer table (31%), both table and duramater (10%), inner table (7%), inner table and duramater (3%) and inner table duramater and brain(3%).(3).

The patient will give history of chronic asymptomatic swelling on the scalp with occasional headache and focal tenderness (4). Malignant transformation is extremely rare and usually occur with larger lesion represented in chronic state (5). The radiologically intradiploic tumour features as an area of bone destruction of lower density than the surrounding bone, with smooth sclerotic margins as first described by Cushing in 1922 (5). CT is excellent source of investigation as compared to plain radiography since it is of greater value in describing bone details. (6), CT scanning provides better visual in defining the size, nature and density of lesion, involvement of outer and inner table and its extension, thus helping to recognize the zones of thinning of bone and status of cerebral parenchyma. CT scan finding will mostly show predominately hypodense lesion with destruction of outer table of skull and thinning of inner table as in our case and attenuation varying from -20 to 20 HU depending upon the ratio of keratin (protein) to cholesterol(lipid) in the lesion. Epidermoid shows characteristic crenated margin on CT and it is devoid of enhancement on contrast administration with occasional calcification noted in the lesion (6). MRI is not a primary modality for evaluation of intradiploic epidermoid since it involves bone destruction and remodeling of the bone for which CT scan has a greater value. MRI may be used as adjuvant to CT which shows low signal with T1 weighting and a high signal with T2 weighting.MRI may be useful in cases of intracranial extension or petrous apex masses such as neurinomas, meningiomas, cholesterol granulomas lipomas, and epidermoids to rule out differential diagnosis (7). The radiological differentiation of intradiploic epidermoid cyst

involves dermoid cyst, eosinophilic granuloma, cavernous hemangioma, giant cell reparative granuloma,arachnoid cyst and cholesterol granuloma. Dermoid cyst is found in orbital region and midline associated with suture line often containing fat mostly seen in childhood, Eosinophilic granulomahave beveled edge appearance and are less heterogeneous as compared to epidermoid. Cavernous hemangioma shows honey combing or radiating sunburst appearance. Giant cell reparative granulomas shows lytic lesion in the absence of sclerosis with MRI scanrevealing isointense to gray matter showing intense enhancement after gadolinium. (8) The mainstay of the treatment is surgical dissection of intradiploic epidermoid cyst for identification of disease, to relieve symptoms, for cosmetic purpose and avoiding possible complication such as intracranial masses, abscess formation, bleeding and malignant transformation (9). Sometime spontaneous ruptureoccur into the subarachnoid space causing aseptic meningitis due to discharge of cystic component like keratin, cholesterol and cellular debris, finally leading to chronic granulomatous archnoiditis (10). The goal of surgey is to remove cyst with complete capsule to lessen the chances of recurrence and malignant transformation. Due to malignant potential, capsule must be carefully dissected from the bone and duramater (9).

Conclusion

At the end we conclude intradiploic epidermoid cyst are slow growing benign tumour arising from ectoderm which should be excised completely along with its cyst lining and capsule, as incomplete removal of cyst is susceptible to recurrence, inflammation and malignant transformation. Meticulous radiological assessment of the cyst should be done by CT and MRI with complete removal of cyst along with its capsule for excellent surgical outcome and better long time prognosis.

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