CASE REPORT

INTRADIPLOIC EPIDERMOID CYST OF THE SKULL

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Intradiploic epidermoid cyst is an uncommon, benign, slow growing neoplasm that occurs between two tables of cranial bones and constitute 0.4% of all cranial epidermoid. It usually occurs due to the entrapped ectodermal embyronal remnants within the skull bones or rarely secondary to trauma. Pre-operative diagnosis on the basis of radiologic investigations is difficult. Complete surgically excision is usually required in order to prevent complications like super infection, intracranial rupture with pneumocephalus and rarely malignant degeneration. We are presenting a case of incompletely resected and chronically infected intradiploic epidermoid cyst of right parietal bone operated inadvertently by a general surgeon elsewhere without doing any radiological investigations.

Keywords: Intradiploic Epidermoid cyst; Cranial epidermoid; Epidermoid; Skull tumours

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INTRODUCTION

Epidermoid cyst is a rare, benign, slow growing lesion that account for less than 1% of all cranial tumors.[1] It may arise intracranially, within diploic space or extracranially with in scalp tissues. Intradiploic epidermoid cyst occur between two tables of cranial bones and constitute nearly 25% of all cranial epidermoids.[2] They are thought to arise from defects in the separation of the neuroectoderm during the formation of the neural tube, leading to sequestration of ectodermal remnants within the cranial bones, However they can also result from introduction of epidermal elements at the time of a trauma.[3,4] Men are affected more commonly than women.[5] Most intradiploic epidermoid cysts manifest clinically within the 3rd or 4th decade of life as a small, longstanding , painless subcutaneous scalp swelling covered with normal skin.[6] Headache, focal tenderness, seizures, intracranial hypertension, focal neurological deficit, traumatic rupture with bleeding, super infection or malignant transformation may occur with large cyst.[7] Histologically epidermoid cysts are characterized by keratin-filled cyst lined by stratified squamous epithelium.[8] Differential diagnosis of intradiploic epidermoid cysts includes aneurysmal bone cyst, haemangioma, eosinophilic granuloma, dermoid cyst and, in the orbitofrontal region, cholesterol granuloma.[9] In general, epidermoid cysts typically appear on CT scans as low-density, hypo dense, non-enhancing lesions, similar to the fat density. On MRI this lesion demonstrates high signal intensity in T1- weighted and variable T2-weighted signal. Sometimes, the cyst contents can be hyper dense, mimicking a haemorrhage. Complete surgical excision is associated with permanent cure, minimal operative mortality and good long-term prognosis.[8]

CASE REPORT

A young 18 years old male patient with the history of painless slowly growing subcutaneous swelling in right parietal region from the last 15 years operated 1 year back by a general surgeon at periphery without doing any radiologic investigations. According to the patient, specimen was not sent for histopathology and no previous record including operation notes were available. The patient presented to neurosurgery out patient department with pus discharge from the surgical wound since surgery despite of using multiple antibiotics. On examination there was 5×5cm swelling over right parietal region with small surgical scar and sinus at the wound edge with pus discharge. Appropriate radiological investigation including computed tomography (CT scan), MRI and bone scan was done. On CT brain there was lytic lesion in right parietal bone with thinned intact inner table and partially eroded outer table. The interior of lesion was iso to hypodense on CT brain with marked soft tissue swelling and little air in subcutaneous tissues (Figure-1 & 2). On MRI the lesion was iso to hypointense on T1WI, hyperintense on T2WI with variable contrast enhancement and marrow oedema (Figure-3 & 4). Bone scan was done to rule out any metastatic bony lesion but findings were consistent with infective process. On the basis of radiologic investigations initial differential diagnosis of Aneurysmal bone cyst, Eosinophilic granuloma and Dermoid/epidermoid cyst was made. Surgery was planned. Under general anaesthesia incision was made over the lesion followed by subcutaneous tissues dissection. Pus collected in 5 cc disposable syringe and was sent for culture and sensitivity. The remaining of the eroded outer table was removed with bone nibbler and sent for histopathology. A white cheesy material within the cyst was removed. The cyst linings were

tightly adherent to the surrounding bone. The inner table was thinned and deformed inward having tiny hole most likely showing intracranial connections (Figure-5). Both the cyst wall and its contents were sent for histopathology. The inner table was kept intact because of the fear of spread of intracranial infection. The culture report was obtained on 4th post-operative day showing staphylococcus aureus as the offending organism sensitive to multiple drugs. Histopathology report showed squamous lined cyst with keratinization, confirming the diagnosis of epidermoid cyst.

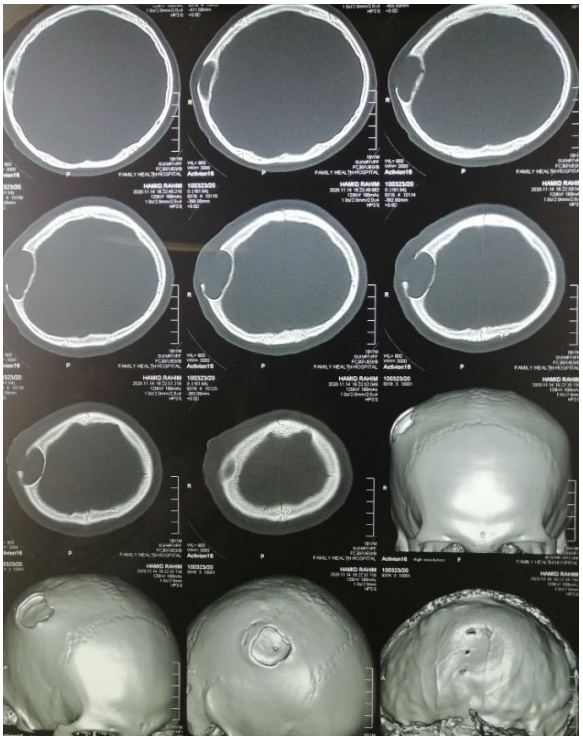

Figure 1: CT skull bone window with 3D reconstruction, Showing lytic expansile lesion in right parietal bone with thinned, intact inner table and partially eroded outer table.

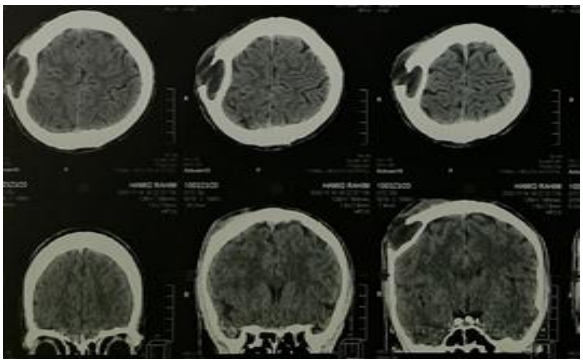

Figure-2: CT brain (axial view), showing intradiploic hypodense lesion with lesion in right parietal bone.

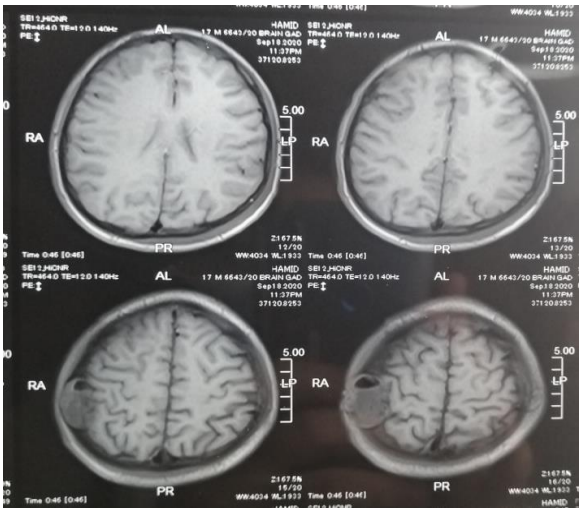

Figure-3: MRI brain (Axial T1WI) showing a hypointense lesion within right parietal bone.

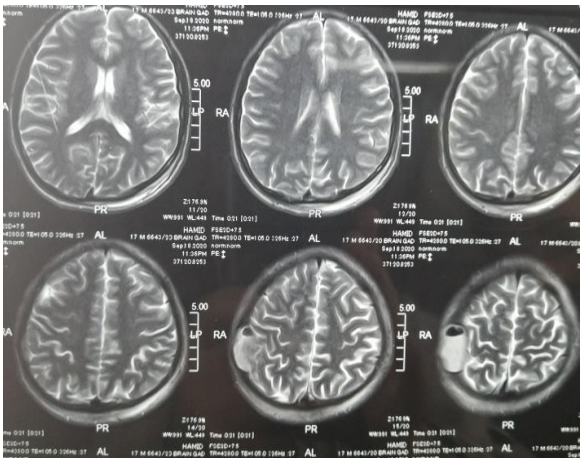

Figure-4: MRI brain (Axial T2WI), showing a hyperintense lesion with area of hypo intensity within right bone.

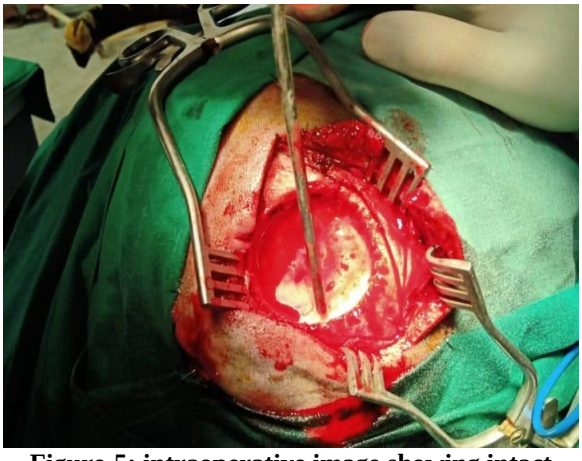

Figure-5: intraoperative image showing intact inner table.

DISCUSSION

Intradiploic epidermoid cyst is an uncommon, slow growing benign lesion that may involve any cranial bone in relation to their size.[9] The origin in this uncommon location is controversial, some believed that inclusion of epidermal nests is of developmental origin, with epidermal cells sequestrated in ectoderm structures during closure of the neural tube during the 3rd to 5th embryonic weeks. Other relates it origin to trauma.[9] In our case the patient was 18 years old having history of long standing, asymptomatic scalp swelling for the last 15 years with no past history of trauma signifies its developmental origin. The most common presentation of intradiploic epidermoid cyst is long standing asymptomatic scalp swelling as evident from our case report, however focal tender, seizures, focal neurologic deficit, super infection, traumatic rupture with bleeding or pneumocephalus and rarely malignancy may occur. Pre-operative diagnosis of intradiploic epidermoid can be challenging because of its close resemblance to other intradiploic lesion like aneurysmal bone cyst. Moreover, the cyst content may be variable in different cases which can alter signals on various MRI sequences further increasing difficulty in diagnosis. In a case report by S. Gaivas the lesion was homogenously hyperintense on both T1 and T2 weighted MR sequences, however in a case report by Gi-Young *et al* the lesion was showing heterogenous signal intensity both on T1 and T2 weighted images.[9] The MR findings of our case report is different from previously reported cases. The high signal intensity on T2 and low to intermediate signal intensity on T1 may be due to super infection with presence of pus with in cyst cavity. No contrast enhancement occurs unless the lesion is super infected. CT scan finding are similar to previously reported case. The goal of the surgery is complete surgical removal of tumour with its capsule which is the only living and growing part of the tumour. Incomplete surgical resection may be associated with complications like malignant degeneration and recurrence. A recurrence rate of 8.3–25% have been reported.[10–12] In our case report we are unable to decide whether the lesion was incompletely resected or recurrent because of lack of previous record. In previous literature the surgical procedure includes removal of inner table as well, followed by cranioplasty but in our case the inner table was thinned and intact. Also due to presence of pus within cyst, inner table was left intact to prevent intracranial spread of infection. We have planned removal of inner table followed by cranioplasty once infection settles down.

CONCLUSION

Intradiploic epidermoid cyst though rare should be included in the differential diagnosis of any scalp lesion. Appropriate radiologic investigation should be done before proceeding to surgical excision of any scalp lesion. Total surgical excision should be performed to prevent complications like super infection, malignant degeneration or recurrence.

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