Intradiploic Dermoid Cyst of the Lateral Skull

Naresh Kumar G.*, Vamshi Krishna**, A.K. Purohit***

Abstract

Background: Intradiploic dermoid cysts represent 0.04– 0.7% of cranial tumours. Very few cases of dermoid cysts occurring in the lateral front temporal region with a sinus tract up to dura are described in literature. We hereby report a case with intradiploic dermoid cyst with intracranial (but extra parenchymal) extension with complaint of a discharging sinus since birth. Case Report: A 2 year old male presented with history of serous discharge from the sinus located in right lateral frontal region at the level of coronal suture noticed since the age of two months. On CT and MRI imaging of brain, patient was found to have intradiploic expansile lytic lesion in the region corresponding to the sinus and was extending intracranially. Differential diagnosis as per imaging studies were Dermoid cyst, Epidermoid cyst and Eosinophilic granuloma. Gross total excision of the cyst along with involved sinus tract done and histopathology of the cyst confirmed it to be Dermoid cyst. Conclusion: Intradiploic dermoid cysts are relatively rare especially with discharging sinus and intracranial extension. We hereby add one such rare case to the literature. Surgical excision with careful excision of tract forms the adequate treatment of choice.

Keywords: Dermoid; Sinus.

Introduction

Dermoid cysts are rare congenital non-neoplastic lesions composed of dermal and epidermal derivatives. They typically occur in childhood, during the first or second decade of life. Dermoids represent approximately 1% of all craniofacial space-occupying lesions and are commonly found in the periorbital, nasoglabellar, and frontotemporal regions [1]. Despite their slow growing nature , these cysts are associated with wide variety of complications including secondary infection, fistula in to cranial cavity etc. Lesions in the midline of the cranium are more concerning for intracranial extension. Intradiploic dermoid cysts are a rare subtype of the frontotemporal dermoids accounting for 0.04–0.7% of all cranial tumors, occurring most commonly at

Author's Affiliation: *Senior Resident, **Assistant Professor, ***Professor and Head, Dept. of Neurosurgery, Nizams Institute of Medical Sciences, Panjagutta, Hyderabad, Telangana State.

Reprint Request: Dr. Naresh Kumar G., H.No 11-15-33/4, Road 1, Plot 5, Doctors Colony, Kothapet, Saroornagar, Hyderabad, Telangana State, Pin No -500035.

E-mail: drnareshgajjala@Gmail.Com

the bregma. They may appear as a lytic defect in the skull, giving a wide differential diagnosis by imaging alone. Frontotemporal dermoid cysts associated with sinus tracts or bony invasion are extremely rare, with only 20 case reports described in the literature [2-9].

We report a case of 2 year old male who presented with discharging sinus from right pterional region since childhood and on evaluation found to have intradiploic lesion.

Case Details

A 2 year old male presented with discharging sinus from lesion in right pterional bone since birth. On examination revealed an abnormal hard bony mass of size 2x2 cm over the right pterion with no discoluration or tenderness. Medical, developmental, prenatal and birth histories were unremarkable. CT brain plain revealed an expansile lytic bony lesion in right intra diploic space with breach in the inner table and MRI of the corresponding section showed intracranial extension with thick peripheral enhancement and central fluid levels in favour of a dermoid or epidermoid cystic lesion.

In view of persistent sinus discharge, patient was operated.



Fig. 1:

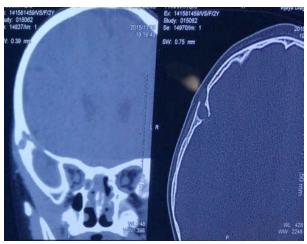


Fig. 2:

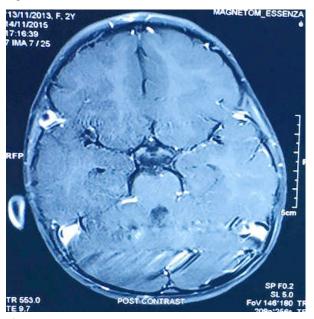


Fig. 3:

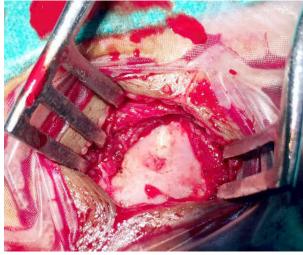


Fig. 4a:

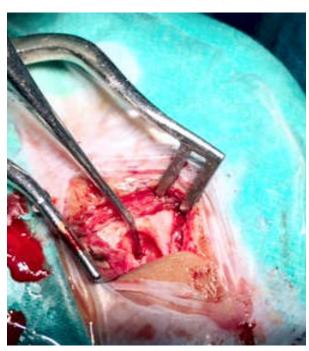


Fig. 4b:

A curvilinear skin incision was made with a temporal base centered on the soft tissue mass. The skin flap was turned and the dermal sinus tract transected. The lesion was found to be 0.7 cm in diameter and fully enclosed by bony matrix. The roof of the growth was drilled through and drained of a sebaceous material which was then sent to pathology. Probing the now open lesion, confirmed partial extension into the cranial vault. The dura was intact and there was small extradural mass which was curetted. The cutaneous portion of the sinus tract was then excised and a primary closure accomplished. Histopathology of the lesion confirmed it to be a dermoid cyst.

Discussion

Dermoid cysts of the skull represent 1% of all craniofacial tumors and Intradiploic dermoids represent between 0.04 and 0.7% of all cranial tumors. They tend to be found along planes of embryonic closure. Majority of periorbital and frontotemporal dermoids are superficially located and amenable to treatment with a simple excision. Histologically, they have a lining of squamous epithelium with dermal elements such as hair follicles, sebaceous and sweat glands. Within the cyst, keratin, hair, smooth muscle, and lipid debris may be seen. There are fewer than 20 case reports which describe lateral frontotemporal dermoids with associated sinus tracts and bony invasion, of which 7 have extended intracranially. We are adding one more case to literature of this relatively rare disease especially with intracranial extension. As discussed there are wide variety of differential diagnosis to intra diploic expansile lytic lesions including Dermoid cyst, Epidermoid cyst, Eosinophilic granuloma etc. Differentiating them imageologically is difficult [10].

These cysts need to be operated in view of its complications [11,12] like secondary infection ,pressure effects, risk of intracranial extension, formation of sinus or a fistula, cosmetic deformity etc.

Considering these complications, they need to be operated at the earliest. Principles of surgery would be complete excision of lesion along with capsule and removing intracranial part in toto if its present. Main focus should be on excising the whole of the sinus tract to prevent recurrence. Entire lesion is to be curetted out and bony reconstruction is advised if bony defect is larger. Histopathologial examination is confirmatory for final diagnosis.

Conclusion

Intradiploic dermoid cysts of skull are rare and are prone to complications if untreated. Early recognition and surgical excision is the treatment of choice. Complete excision has good results and less chances of recurrence.

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