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Anterior Fontanelle Congenital Inclusion Dermoid Cyst

KEYWORDS

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INTRODUCTION

A dermoid cyst is the most common congenital lesion found around the anterior fontenelle. However, a congenital inclusion dermoid cyst of the anterior fontenelle is a rare lesion accounting for mere 0.2% of all scalp lesions and 0.1% of all cranial tumours1. So far only 163 cases are reported in the world literature 2.

The following case report describes the rare lesion of a congenital inclusion dermoid cyst found at the anterior fontenelle, which was successfully treated by surgery.

CASE REPORT

A 13-month old female child was brought to the hospital with the history of a progressively increasing globular mass over the anterior half of the midline of the head since the time of birth. It was very small measuring about a pea at the time birth but has been progressively increasing in size.

Physical examination showed no other finding except for the presence of a soft mobile non tender mass measuring 4cm in diameter over the anterior fontenelle (Fig:1). Fluctuation was positive. Clinically there was no evidence of intracranial extension of the cyst. The child was normal neurologically.

Routine investigations were normal. Fine needle aspiration cytology of the mass was done by a private practitioner before coming to our hospital, which revealed clear fluid- ?cerebrospinal fluid, without any cells. CT scan demonstrated non calcified cystic lesion over the anterior fontenelle just beneath the galeaaponeurotica. There was no evidence of any intra cranial extension (Fig: 2).

Clinically, a case of congenital dermoid cyst over the anterior fontenelle was diagnosed with no intra cranial extension and the child was prepared for surgery.

Under endotracheal general anaesthesia, the child was placed in supine position. The part was cleaned and draped. An antero-posterior elliptical incision was made directly over the mass. The incision was deepened. A cyst with a pearly white in appearance was encountered (Fig: 3).The entire was easily dissected from the surrounding tissue. The excised cystic lesion was found to contain clear fluid (Fig: 4). There was no intracranial extension nor any attachment to the underlying superior sagittal sinus. After ensuring haemostasis, the skin margins were approximated with the sutures. The immediate postoperative period was uneventful. During follow-up there was no recurrence after six-months. The histo-pathological examination revealed a cyst lined by stratified squamous epithelium. The stratum corneum showed keratinization in basket weave pattern. Skin appendages were found in the cyst wall. No neurological tissue was found. The histological diagnosis was "inclusion dermoid cyst".

DISCUSSION

Dermoid cyst is a pathological cyst lined by squamous epithelium containing skin appendages like hair follicles, sebaceous glands and sweat glands. Dermoid cyst is classified into three categories: (1). Teratoma type: True congenital dermoid cysts. (2). Acquired implantation dermoid cysts and (3) Congenital inclusion dermoid cysts, resulting from the inclusion of displaced dermoid cells along the embryonic fusion lines.

Congenital inclusion dermoid cysts of the scalp region are very rare lesions, but known since Cruveilhier days of early nineteenth century 3. Earlier it was named as 'pearly tumours'. The incidence of congenital inclusion dermoid cysts as per the available medical literature is very rare 1. So far only 163 cases are reported in the world literature 4. A total of only 17 cases of anterior fontenelle inclusion dermoid cysts have been reported in Japan till 1993 5. More number of cases may have gone unreported as extra cranial dermoid cysts without intracranial extension are considered as simple lesions.

The congenital inclusion dermoid cysts develop as a result of desquamation, proliferation and accumulation of dermal cells. These develop in between 3 and 5 weeks of intrauterine life along the midline or at the lateral fusion lines. The anterior fontenelle is the most common site of congenital inclusion dermoid cyst 4. Initially it was exclusively reported in the black races.6, however later on these lesions have also been reported from the other races also 7. There is a slight preponderance for females as in our case. Most of these cysts are located at the lateral angles of the anterior fontenelle and found to be sub galeal in location. These are usually observed at the time of birth and gradually increase in size through the accumulation of secretions and internal desquamation of cells.

Physical examination reveals a soft mobile non tender mass over the anterior fontenelle covered with normal intact skin as in our case. Any congenital midline lesion of the scalp should inevitably arouse in the clinician`s mind the possibility of the presence of intracranial extension. However most of the inclusion dermoid cysts over the anterior fontenelle do not have any intracranial extension as in our case.

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On plain radiograph of the skull, a soft tissue mass over the anterior fontenelle is seen. It may show either flattening or indentation or pitting of the outer table and some times a bone defect extending up to the inner table. On neuro-imaging, the CT scan shows an extra cranial heterogenous hypo dense mass without any intracranial extension.as in our case.

Diagnostic aspiration of the cyst is not recommended because of increased incidence of contamination and secondary infection. Though, in our case, fine needle aspiration cytology was done, there was no contamination nor any secondary infection.

The differential diagnosis of congenital inclusion dermoid cyst at the anterior fontenelle includeencephalocele, sebaceous cyst, haemangioma, lymphangioma, melanoma and lipoma.

The inclusion dermoid cysts are excised mainly for cosmetic reasons, to prevent infection, to obtain histological diagnosis and to rule out malignancy. The best procedure for treatment is a complete excision of the cyst by careful blunt dissection from the underlying structures through an ellipsoidal incision bordering the mass. Care should be taken while excising the mass overlying an open anterior fontenelle or a lytic bone to avoid injury to its underlying dura and / or superior sagittal sinus. Local anaesthesia under sedation is preferable to general anaesthesia in most cases especially when no lytic bone exists or the size of the cyst is not very large. Though the central nervous system dermoidtumours are occasionally undergo malignant change, there is no reported evidence of malignant transformation of inclusion dermoid cysts. Hence simple complete excision is the best method of treatment. Recurrence may occur if partial excision is done.

Histology of congenital inclusion dermoid cyst shows a cyst wall lined by a thick stratified squamous epithelium containing skin appendages like hair follicles, sebaceous glands and sweat glands.

CONCLUSION

Congenital inclusion dermoid cyst is a rare type of dermoid cyst presenting at the anterior fontenelle in a new born. It requires a careful examination to rule out intracranial extension. It requires a simple surgical excision as the treatment of choice.

Fig: 1 Mass over anterior fontanelle

Fig: 2 CT Scan showing non calcified cystic lesion

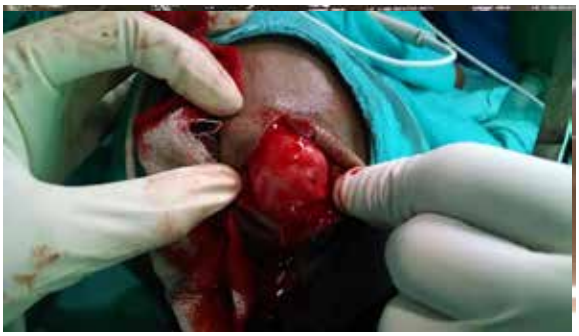
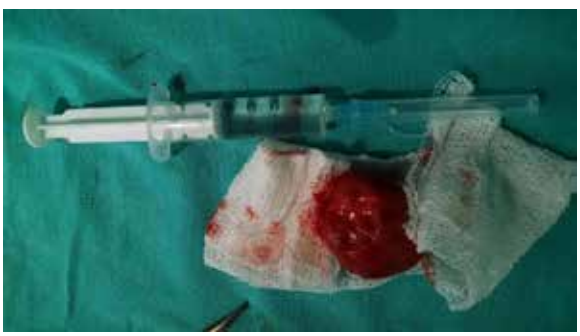


Fig: 3 removal of the cyst
Fig: 4 excised cystic lesion containing clear fluid



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