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Congenital Giant Dermoid Cyst of the Orbit Associated with Microphthalmia and Cataract

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Summary

Baiyeroju-Agbeja AM and Thomas J . Congenital Giant Dermoid Cyst of the Orbit Associated with Microphthalmia and Cataract. *Nigerian Journal of Paediatrics 1995; 22: 94.* A case of a giant dermoid cyst of the orbit associated with microphthalmia in a two-year old female child is reported. Clinical diagnosis of the condition was made when the child was two months old, although it had been observed at birth by the parents. Surgical excision of the tumour and the microphthalmic eye was undertaken when the child was two years old. Findings at the operation included a giant cyst that was partially intraorbital with a portion of it extending into the temporal fossa, a microphthalmic eye with cataract and non-closure of the zygomatico-frontal suture. Six months after surgery, there was no evidence of recurrence of the tumour.

Introduction

DERMOID cysts of the orbit are usually congenital, although most of them do not manifest until later years. The cyst is thought to arise from the sequestrated surface epithelium within the deeper orbital tissues during embryonic development. A typical cyst usually presents as a painless subcutaneous mass which is regular in configuration, non-mobile to palpation and not attached to the overlying skin. It is most often located in the upper temporal quadrant, where it manifests near the region of the lacrimal gland.[1] This benign tumour does not usually threaten sight but very rarely, can be associated with microphthalmos.[2] Complete surgical excision of the cyst is the treatment of choice, avoiding rupture of the fibrous wall as the contents could incite a mild, but smouldering granulomatous inflammation.[1,3] Prognosis, following removal of the cyst, is good and the lesion rarely recurs. The present communication reports a rare case of congenital giant dermoid cyst of the orbit associated with microphthalmia and cataract.

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Case Report

A two-year old girl (UCH No 883629) first attended the Eye clinic, University College Hospital (UCH), Ibadan, at the age of two

months. At that initial visit, the main complaint given by the parents was that the left eye was smaller than the right. This was noted at birth and was associated with a small swelling lateral to the small eyeball. Examination under anaesthesia and ultrasonography, revealed a left microphthalmic eye, measuring 9mm anteroposterior (AP) diameter; the right eye measured 20mm (AP). There were also corneal opacity and a small pupil with a densely cataractous lens. The eye was compressed by a 25 x 20mm cystic mass in the superotemporal aspect of the orbit, extending into the left temporal area and causing a complete ptosis. On the basis of the above findings, a clinical diagnosis of congenital microphthalmia with cataract and a dermoid cyst was made. It was then decided that the mass be left alone until the child was older, as the eye was already blind. As the child grew the extraorbital portion of the cyst increased in size (Fig) until at the age of two years when the cyst was found on ultrasonography to measure 50 x 35mm with irregular solid areas in it, occupying mainly the temporal area, while a smaller part of it was still intraorbital. A bone defect was also noticed on the orbital rim with non-closure of the zygomatico-frontal suture and an extra growth of bone in a fingerlike fashion on the lateral aspect of the frontal bone. The microphthalmic eye had also reduced in size to 8mm AP. Ultrasound and skull radiograph showed no communicaiton with the intracranial cavity.

Under general anaesthesia, the patient underwent surgery at which the cyst was found to be partly intraorbital with a portion extending into the temporal fossa, causing muscle atrophy and pressure erosion of the frontal bone with non-closure of the zygomatico-frontal suture; it was also firmly attached to the finger-like bony growth from the frontal bone. The cyst and the microphthalmic eye were excised. The immediate post-operative condition of the child was satisfactory. At six months follow-up of the patient, no recurrence was noted, but the bony growth was still present although it had regressed slightly in size.

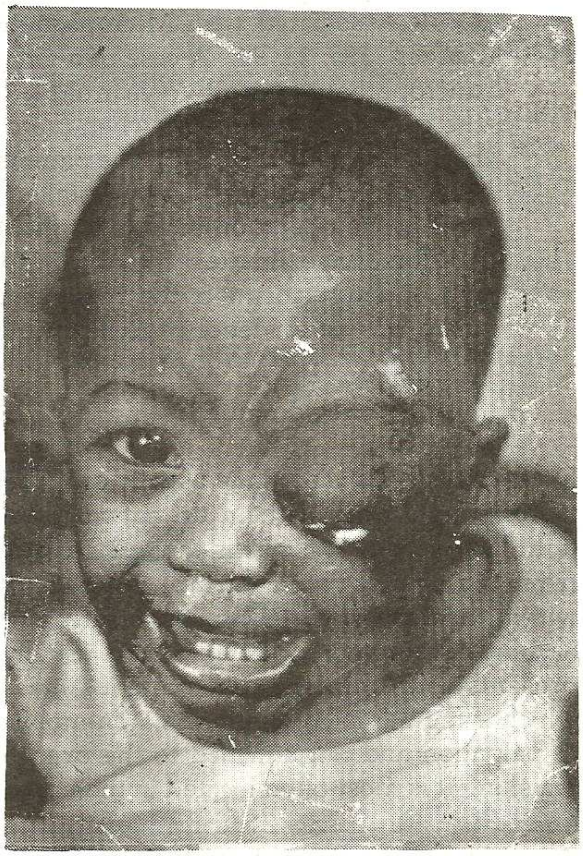

Fig: Photograph of the patient showing marked swelling of the left eyelids and ptosis caused by a giant dermoid cyst.

Discussion

Dermoid cysts are common periocular childhood tumours that constitute about seven percent of all orbital tumours.[4] By contrast, giant dermoid cysts of the orbit, as far as can be ascertained, has never been reported in childhood, and only three cases have been reported in adults during the last 20 years. [5 6] Dermoid cyst associated with microphthalmos is much rarer still.[2] The pressure effect of the cyst presumably caused maldevelopment of the foetal eye *in-utero*. Dermoid cysts have occasionally caused non-closure of the zygomatico-frontal suture as occurred in our patient. Samuelson *et al*[7] have reported a case of zygomatico-frontal suture defect associated with a dermoid cyst, which communicated with the temporal fossa. The zygomatico-frontal suture is a strong joint that is required for protection of the orbit. During infancy, its development can be altered by local mechanical factors which prevent its complete union. Sathananthan *et al* [8] also reported significant bony abnormalities in the lateral wall and supero-temporal angle of the orbit with dermoid cysts. These abnormalities included pressure erosion, pit or tunnel formation, abnormal orbital shapes and abnormal bone texture. These findings are extremely important for planning adequate surgery; they also indicate that bony involvement is much more frequent than previously appreciated. Prognosis following excision of the cysts is good and the lesion rarely recurs. In our patient, prognosis is expected to be good as the cyst was removed in all its entirety. At six months follow-up, no recurrence was noted and the fingerlike bony growth had regressed slightly.

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