Congenital inclusion dermoid cyst located over the region of the anterior fontanel in adult Nigerians

Report of two cases

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Two cases of a congenital inclusion dermoid cyst located over the region of the anterior fontanel are described in adult Nigerians, aged 28 and 32 years. In both cases, the cystic lesion was noticed at birth over the anterior fontanel as a small soft lump, and was carried on the head from infancy into adult life. The clinical, radiological, and the operative findings revealed no intracranial extension. The two lesions were histologically verified as dermoid cysts. Surgical excision was curative.

Key Words • congenital cyst • anterior fontanel • dermoid cyst

The presence at birth of a soft, fluctuant, non-tender mass which transilluminates and is located over the anterior fontanel raises the suspicion of a congenital inclusion dermoid cyst rather than any of the other extracranial masses. This is particularly true when seen in black African children. The etiological factors responsible for the frequent occurrence of this entity in the black African race more than in other races are not known. Most children born with this type of a congenital cystic tumor located over the anterior fontanel are treated surgically within the 1st year of life. No case has yet been reported in the literature in which a child has carried such a congenital lesion from infancy into adulthood. The largest recorded series of extracranial dermoid cysts located over the anterior fontanel was reported by Adeloye and Odeku; the oldest of their 18 patients was a girl of 18 years.

We are reporting on the oldest patients with this type of congenital cystic lesion in this location to appear in the literature to date. The contents of the excised tumors and the length of time these lesions were carried were unusual features of these cases.

Case Reports

Case 1

This 32-year-old Nigerian woman was born with a very small soft mass located in the midline over the vertex of the head. The mass increased gradually in size as she grew older. Her parents were fearful of hospitals, and as a result she harbored the mass on the head until after her marriage. For cosmetic reasons, she and her husband decided to have the tumor removed, and she was referred to the Neurosurgical Unit of the Lagos University Teaching Hospital on December 5, 1978, for surgical treatment.

Examination. The patient appeared healthy, and was fully conscious and alert. There was a big, soft, non-tender and non-pulsatile cystic mass situated over the region of her closed anterior fontanel, covered with intact skin (Fig. 1). The mass was not translucent. Physical examination revealed no other abnormality.

There was no neurological deficit. The skull radiographs showed no intracranial involvement, but there was a flattening or depression of the external table of the cranium beneath the tumor mass, and also a soft-tissue shadow over the region of the anterior fontanel. Routine air encephalography revealed a normal ventricular pattern with no extension of the air into the cystic swelling. Hemoglobin and electrolytes were normal.

Operation. On May 4, 1979, the patient was operated on. An elliptical skin incision over the summit of the cystic mass revealed a semisolid thin-walled tumor, which was carefully shelled out from beneath
Dermoid cysts over anterior fontanel

**Pathological Examination.** Gross examination of the excised tumor showed an egg-shaped mass measuring $6.5 \times 4.5 \times 5.5$ cm, which weighed 75 gm (Fig. 2 left). The cut surface of the tumor showed a yellowish pastry-like material with a few hairs contained within the cyst cavity (Fig. 2 right). Microscopic sections showed the cyst wall to be made up of connective tissue lined by stratified squamous epithelium, which was mostly atrophic. Beneath the epithelium could be seen some large structures containing clear spaces, very reminiscent of sebaceous glands. Inside the cyst, keratin debris was found, and in areas where stratified epithelium was not visible, some foam cells could be seen adherent to the cyst wall. There was no evidence of malignancy (Fig. 3). Histologically, the tumor was diagnosed as a dermoid cyst. The postoperative course was uneventful.

**Case 2**

This 28-year-old Nigerian man was born with a small soft mass located over the anterior fontanel. A few months after his birth, he was taken to a private hospital for treatment. When surgery was suggested as the only curative treatment, the patient's parents took him from the hospital against medical advice. The cystic mass gradually increased in size with increasing age. Shortly before attaining the age of 28 years, the patient started to feel heat and pain in the head. He consulted a private clinic, which referred him to the Neurosurgical Unit of the Lagos University Hospital for treatment on July 11, 1973.

**Examination.** Clinical examination revealed a big, rounded, soft, non-tender and non-pulsatile mass, measuring about $12 \text{ cm} \times 12.5 \text{ cm}$, located over the...
region of the anterior fontanel (Fig. 4). The mass had a typical sessile base, and was covered with intact skin. It did not transilluminate. It was slightly mobile from front to back and from side to side. There was no neurological deficit. Skull radiographs showed a big, rounded, soft-tissue shadow situated over the region of the anterior fontanel, and a flattening of the external table of the cranium directly beneath the tumor shadow (Fig. 5). All cranial sutures were closed. Air encephalography did not show any intracranial connection. Routine laboratory investigations of blood and urine were normal.

Operation. At operation on July 16, 1973, a large thin-walled cystic mass was excised radically from beneath the galea aponeurotica over the region of the anterior fontanel. The tumor had depressed the external table of the cranium, but there was no intracranial connection seen. The mass was sent for histological study.

Pathological Examination. Histologically, the tumor specimen was found to show the same structures as those described in Case 1 (Fig. 3), and was diagnosed as a dermoid cyst. There was no evidence of malignancy. As in Case 1, the postoperative course uneventful.

Discussion

Most reports on dermoid cysts of the craniospinal axis have been confined to the intracranial and intraspinal cavities. Congenital extracranial dermoid cysts located over the region of the anterior fontanel in adults are very rare. No case involving an adult has been described so far in the literature. Reported cases of congenital extracranial subgaleal dermoid cysts located over the anterior fontanel have been confined to infants and children under the age of 18 years.  

This type of inclusion dermoid cyst, which is rare in the general population, is more frequently seen in black African children than in Caucasian children. The etiological factors that may explain this difference are probably unknown. The rarity in the Caucasian adult of this congenital cyst lesion in this location may be connected with the fact that in developed countries children born with this type of lesion receive palliative surgery before adulthood; whereas in underdeveloped or developing countries the chances of carrying the cystic tumor into adulthood are greater due to a scarcity of essential facilities.

Embryologically, dermoid and epidermoid cysts
Dermoid cysts over anterior fontanel

Dermoid cysts over anterior fontanel

Fig. 5. Case 2. Radiograph showing soft-tissue shadow and a depression of the external skull plate.

develop from germ cells displaced between the 3rd and 5th week of embryonic development. These tumors are particularly apt to lie along the line of closure in the skull, and, when lying over the gap in the midline of the cranium, a pedicle may connect the dermoid cyst with the dura mater, in which case it may be mistaken for a meningoencephalocele. Other cystic lesions occurring in the same neighborhood that may also be considered in the differential diagnosis are sebaceous cysts, lipomas, hemangiomas, and subgaleal hematomas. Extracranial dermoid cysts are characteristically roundish in shape, with a typical broad sessile base, and are soft and non-reducible on pressure. The skull radiographs often show no intracranial connection, but may reveal bone changes that include flattening of the external table of the cranium (Fig. 5).

Histologically, these extracranial dermoid lesions have a cystic structure, with a lining of stratified squamous epithelium. Adnexal structures, such as sweat glands, hair follicles, and sebaceous glands, are commonly present in the cyst wall. This same histological pattern is seen in dermoid cysts over the anterior fontanel in both children and adults.

Dermoid cysts in infants and young children contain largely colorless to yellowish fluids mixed with some cheesy materials and hair. Adult dermoid cysts include mostly semisolid pastry-like material or buttery yellowish masses of sebaceous secretion with desquamated epithelial cells mixed with a matted mass of hairs accumulated over a period of years.

References

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