

Congenital Dermoid Cyst of the Anterior Fontanel

Ön Fontanelin Dermoid Kisti

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Abstract: Congenital dermoid cysts of the central nervous system are rare lesions. Congenital dermoid cysts located over the anterior fontanel are reported in two Turkish infants. These two cases are examples of such a lesion in an infant of non-African descent.

Key Words: Anterior fontanel, congenital inclusion cyst, dermoid cyst

Özet: Santral sinir sisteminin konjenital dermoid kistleri nadir lezyonlardır. İki Türk çocuğunda, anterior fontanel üzerinde yerleşimli konjenital dermoid kist bildirilmiştir. Bu iki olgu, Afrika orijinli olmayan çocukta böyle bir lezyon için örnek oluşturmaktadır.

Anahtar Sözcükler: Anterior fontanel, dermoid kist, konjenital inklüzyon kisti

INTRODUCTION

Dermoid cysts of the central nervous system are rare lesions, incidence is 0,1 - 0,7 % (8,17). First, it was only reported for black races (1,3-5,7,13-15), the cases from other races have also been reported (2,9,11,12,16,18).

Congenital inclusional dermoid and epidermoid cysts develop between 3 to 5 weeks of intrauterine life (17).

We report two cases of dermoid cysts located over the anterior fontanel in two Turkish infants.

CASE REPORT

Case 1: A 6-month-old male infant who was admitted due to the soft mass over the head, had this mass noticed by his parents shortly after birth and the mass progressively enlarged in time.

Physical examination showed no other pathological findings, except for the soft mobile mass

measuring 2 cm in diameter over the anterior fontanel (Figure 1).

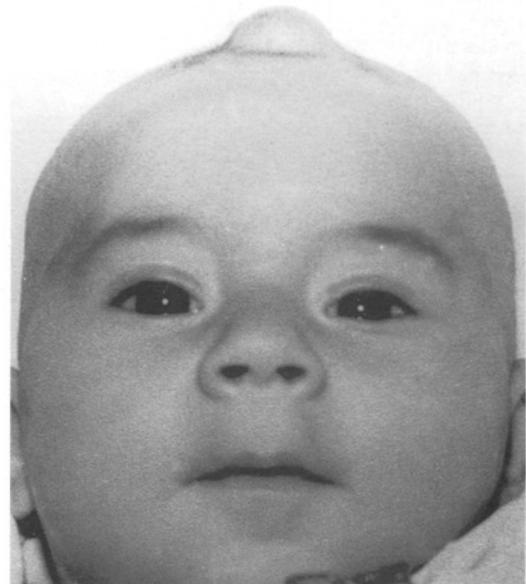


Figure 1. The first patient's photograph demonstrates a mass over the anterior fontanel.

Radiograms of the skull revealed a soft tissue shadow over anterior fontanel. Pathological findings were not detected in bone tissue. Computerized tomography (CT) showed a cystic mass measuring 2 cm in diameter located extracranially over the anterior fontanel. The lesion reflected 30 HU density (Figure 2).

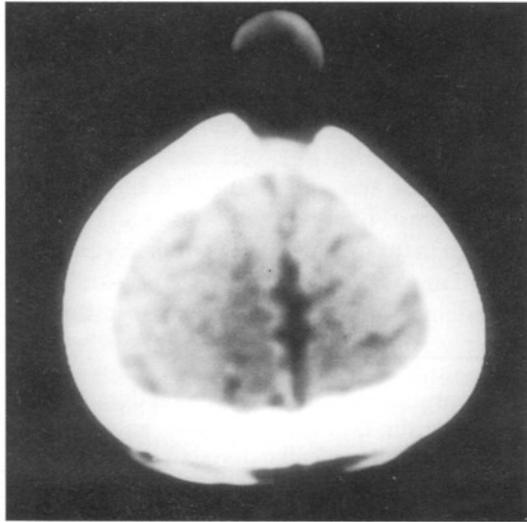


Figure 2. CT demonstrates an anterior fontanel cyst without any connection with the intracranial contents.

During surgery, over the anterior fontanel, the cyst which was attached to the underlying outer layer of the dura mater by a thin peduncle was easily dissected from the galea and scalp (Figure 3). The cyst, measuring 2 cm in diameter was excised together with its pedicle. Superior sagittal sinus or

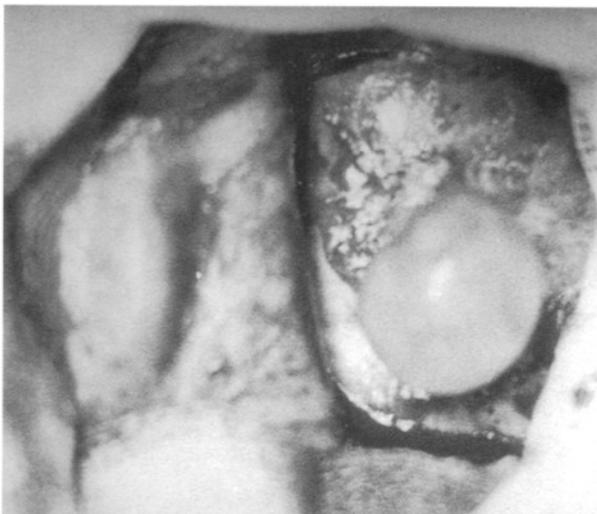


Figure 3. Operative photograph demonstrating a cystic mass over the anterior fontanel.

intracranial connection were not demonstrated. The cyst fluid was clear.

Histopathological examination revealed cystic spaces lined by stratified squamous epithelium. There were acellular eosinophilic lamellar keratinized materials in the lumen and skin appendages in the outer cystic space. Neurogenic tissue was not observed (Figure 4).

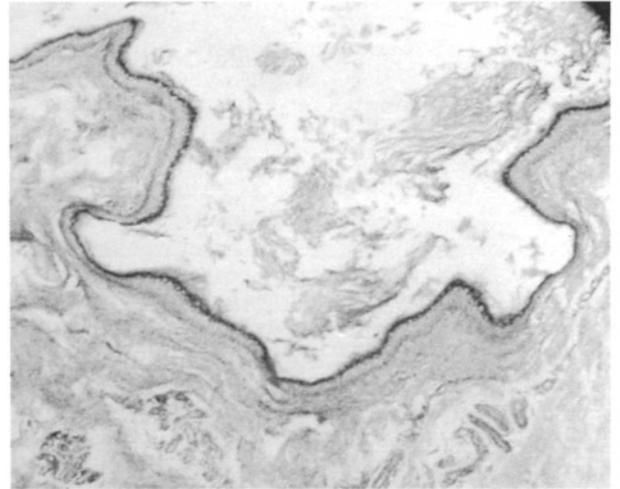


Figure 4. Histological features of the cyst, containing skin and skin appendages (H&E x 100).

The postoperative period was uneventful.

Case 2: A 8-month-old male infant who was admitted due to the soft mass over the head, had this mass noticed by his parents shortly after birth and the mass progressively enlarged in time.

Physical examination revealed no other pathological findings, except for the soft mobile mass measuring 3 cm in diameter over the anterior fontanel (Figure 5).

Radiograms of the skull revealed a soft tissue shadow over anterior fontanel. Pathological findings were not detected in bone tissue. Ultrasonography showed a cystic lesion, measuring 15,4 x 29 mm in size, localized between the scalp and the ecogenic dura over the anterior fontanel region (Figure 6). Coloured doppler duplex sonography displayed no pathological vascularization detected in the cyst wall. Intracranial parenchymal vascular structures were normal. CT showed a cystic mass measuring 3 cm in



Figure 5. The second patient's photograph demonstrates a mass over the anterior fontanel.

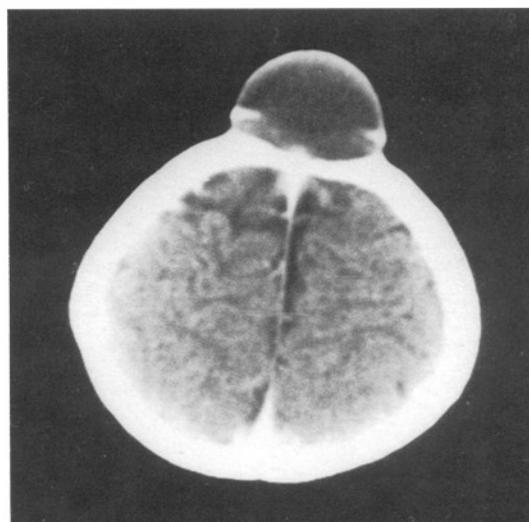


Figure 7. CT demonstrates an anterior fontanel cyst without any connection with the intracranial contents.

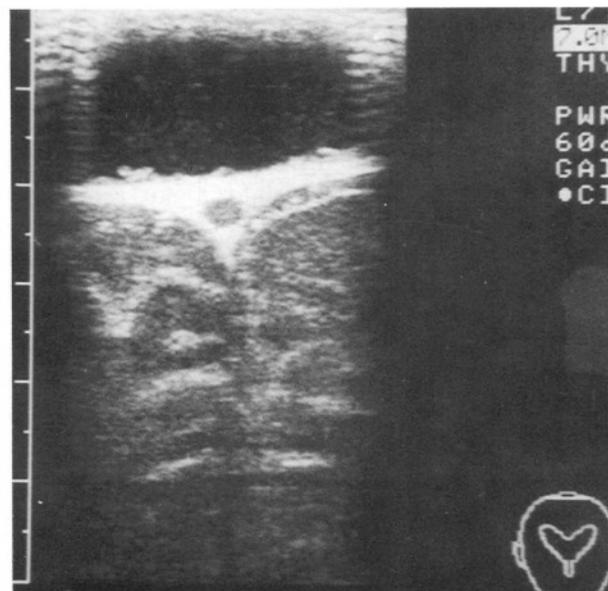


Figure 6. USG demonstrates a cystic lesion localised between the scalp and the ecogenic dura over the anterior fontanel region.

diameter located extracranially over the anterior fontanel (Figure 7). Magnetic resonance imaging (MRI) showed a cystic lesion over the anterior fontanel beneath the skin without any connection with the subarachnoid space. The cystic lesion, measuring 17,6 x 36,7 mm in size reflected isointense signal intensity close to the cerebrospinal fluid (Figure 8).

During surgery, over the anterior fontanel, the cyst which was attached to the underlying outer layer of dura mater by a thin peduncle was easily dissected

from the galea and scalp. The cyst, measuring 3 cm in diameter was excised together with its pedicle. Superior sagittal sinus or intracranial connection were not demonstrated. The cyst fluid was clear. Laboratory data of the cyst fluid showed protein 108 mg %, glucose 50 mg %, chloride 118 mEq/l, and leucocyte 10/mm³.

Histopathological examination revealed cystic peripheral tissue with keratinized materials in its lumen. There were skin appendages and scattered mononuclear leucocyte infiltration beneath the epithelium. Neurogenic tissue was not observed.

The postoperative period was uneventful.

DISCUSSION

Dermoid and epidermoid tumors of the central nervous system are rare tumors known since Cruveilhier (6) in 1829 and named as pearly tumor. The incidence of dermoid cysts is 0,1-0,7% (8, 17).

Congenital inclusional dermoid cysts develop in between 3 to 5 weeks of intrauterine life along the midline or lateral fusion lines at the time of closure of the neural groove (4, 11, 17).

The anterior fontanel is the most common site of congenital inclusional dermoid cysts (1, 2, 4, 13-15, 17) but other midline or paramedian localizations have also been reported (4,5). So far, there are 163

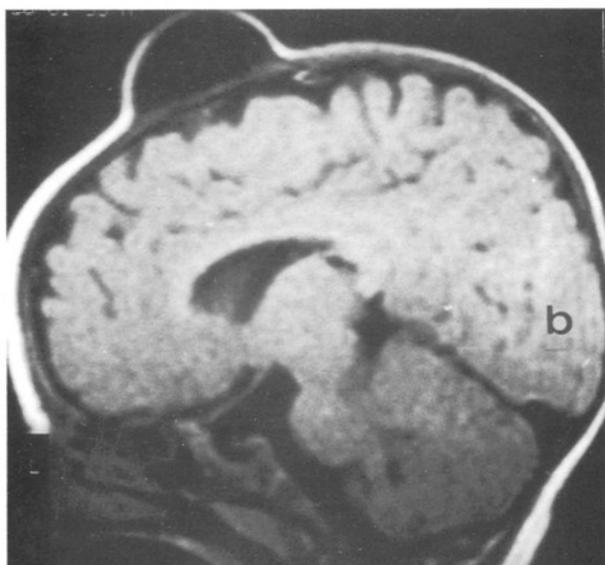
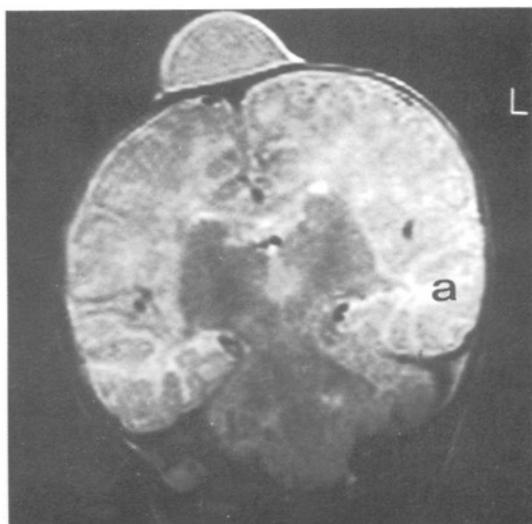


Figure 8. MRI demonstrates a lesion, reflecting isointense signal intensity close to CSF, over the anterior fontanel without any connection with the intracranial contents.

dermoid cyst and epidermoid cyst cases have been reported over the anterior fontanel in literature, though teratomas are comparatively common (5).

First, it was only reported for black cases (1, 3-5, 8, 16-19) later, cases from other races have also been reported (2, 9, 11, 12, 16, 18). Though our cases were male, there is a preponderance for females in the anterior fontanel dermoid cysts (1,4,8,13,14,16).

Clinically, these cysts don't reflect findings apart from a soft, mobile, nontender mass over the

scalp (1, 4, 5, 13-16, 18, 19). In our cases the soft mobile masses measuring 2 and 3 cm in diameter over the anterior fontanel were the only findings.

On plain films of the skull soft tissue mass over the anterior fontanel, flattening indentation or pitting of the outer table, and sometimes bone defects extending up to the inner table are noted (3-5, 7, 8, 11, 16, 18, 19). CT is valuable in showing extracranial localization of the cyst and excluding the intracranial extension (9, 11). Findings in our cases correlate well with literature.

Pathologically dermoid cysts, have been classified in three groups (10,11): a) congenital (teratoma type); b) acquired (by implantation); c) congenital inclusion cysts (these develop as a result of desquamation, proliferation, and edema of dermoid cells). Microscopic diagnosis of the dermoid cyst is very easy. Different from epidermoid cysts, they contain all the skin appendages, such as hair follicles, sebaceous glands, and sweat glands (3,15-19). The contents of the cyst are occasionally infected. Coliform B was isolated from the contents in two reported cases (4, 8). Adeloje and Odeku (1) reported that the contents of the smaller dermoid cysts were low in protein and glucose while high protein and glucose values were found in larger ones. Our second case reflects the same findings.

Though the central nervous system dermoid tumors may occasionally undergo malignant change and recurrence, there is no reported case for inclusion dermoid cysts undergoing malignant change and recurrence in the series in which total excision has been used as a method of treatment (4, 8, 10). Recurrence has only been reported in partially excised cases (4, 13).

The differential diagnosis includes encephalocele, sebaceous cyst, lymphangioma, hemangioma, melanoma, progonoma, cephal hematoma, lipoma, and sinus pericranii (4,5,9,11).

As in our cases, treatment in congenital inclusion dermoid cysts is total excision, which provides definitive diagnosis, prevents infection and serves cosmetic reasons (8, 11, 16). Diagnostic aspiration of the cyst is never recommended because it increases the risk of contamination and secondary infection (4, 8).

In conclusion, our cases confirm that congenital dermoid cysts are seen in whites as well as in blacks.

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