

Congenital Dermoid Cyst of the Anterior Fontanelle in Turkish Children

—Four Case Reports—

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Abstract

Four cases of congenital inclusion dermoid cysts located over the anterior fontanelle occurred in male Turkish children aged 6 months to 5 years. These rare cysts presented as slow-growing soft masses over the anterior fontanelle. The cysts were totally removed without recurrence. Dermoid cysts can be easily cured because of the absence of intracranial extension.

Key words: congenital dermoid cyst, inclusion cyst, anterior fontanelle

Introduction

Dermoid cysts are rare lesions, with an incidence of 0.1–0.5% of all cranial tumors.^{5,9,11)} Dermoid cysts occur along the midline of the cerebrospinal axis if the skull is involved, usually at the anterior fontanelle.^{5,6)} Based on their pathogenesis, dermoid cysts can be classified into three categories⁴⁾: Congenital dermoid cysts of a teratoma type, acquired implantation dermoid cysts, and congenital inclusion dermoid cysts. Inclusion dermoid cysts arise from sequestered epithelial rests or ectodermal inclusions along the line of growing membranous bone.^{4,12)} Such lesions can occur at various sites in the skull, but more often over the anterior fontanelle.⁶⁾ Inclusion cysts of the anterior fontanelle almost always present as slow-growing soft

masses. Computed tomography (CT) and magnetic resonance (MR) imaging are effective for the preliminary diagnosis of these lesions and total removal is possible in all cases.

Here we describe four cases of congenital inclusion dermoid cyst of the anterior fontanelle in Turkish children.

Summary of Cases

The clinical findings are summarized in Table 1. All four children presented with a slow-growing soft mass located in the midline over the vertex of the head. All masses had been noted by parents at birth, but the age at diagnosis ranged from 6 months to 5 years. All patients were Caucasian males. The masses ranged in diameter from about 3 cm to 4 cm. The lesion was always covered with intact skin with no abnormal skin changes or dimples (Fig. 1). The children had normal growth and development and no associated abnormalities of the central nervous system or body.

Skull radiography revealed a hypodense lesion surrounded by a sclerotic border at the anterior fontanelle and flattening or depression of the external table beneath the tumor mass (Fig. 2). The soft tissue shadow of the cyst was also seen. CT demonstrated an extracranial, heterogeneous

Table 1 Dermoid cysts of the anterior fontanelle

Case No.	Age	Sex	Race	Diameter (cm)
1	6 mos	M	Caucasian	3
2	5 yrs	M	Caucasian	4
3	3 yrs	M	Caucasian	3
4	5 yrs	M	Caucasian	3

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Fig. 1 Photographs of 6-month-old (A) and 5-year-old (B) boys with dermoid cyst of the anterior fontanelle showing the cyst covered with normal skin.



Fig. 2 Skull radiograph showing a hypodense lesion surrounded by a sclerotic border at the anterior fontanelle and the soft tissue shadow of the cyst.

hypodense mass, without intracranial extension (Fig. 3). MR imaging showed a low-intensity lesion on the T_1 -weighted images, and a high-intensity lesion on the T_2 -weighted images without apparent connection with the intracranial contents (Fig. 4).

All cysts were located on all aspects of the anterior fontanelle, and under the galea aponeurotica. No cyst extended into the superior sagittal sinus, but an associated erosion defect in the underlying bone was present, which may extend through the inner

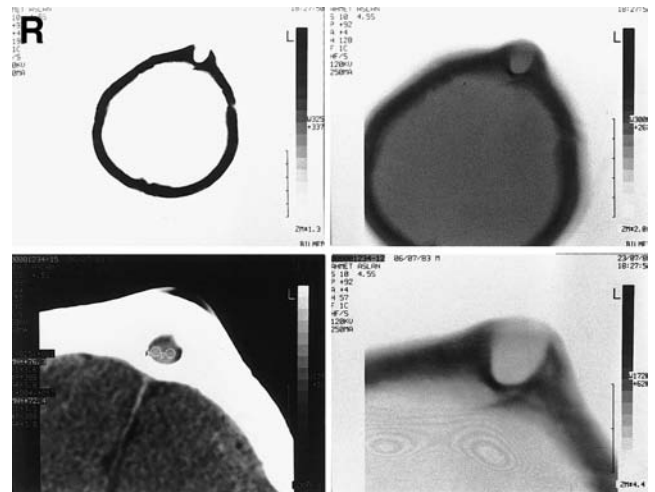


Fig. 3 Computed tomography scans demonstrating an extracranial, heterogeneous hypodense mass, without intracranial extension.

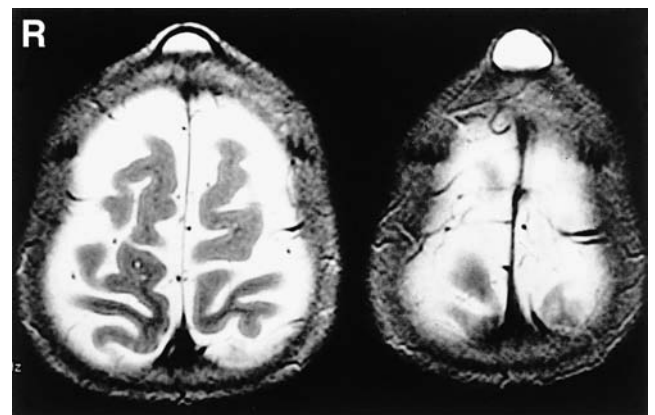


Fig. 4 T_2 -weighted magnetic resonance images showing a high-intensity lesion without apparent connection with the intracranial contents.

tube. The surgical technique of choice was a small coronal flap, elevation of the galea above the cyst, and retraction. The cyst was then dissected from the underlying tissue and removed completely. Yellowish brown fluid and some caseous material were noted. The masses could easily be dissected bluntly from the subgaleal tissue and excised intact. No tissue connection to the overlying skin or intracranial extension was found in any case. Removal was total in all cases. The patients have been followed up for 9 to 18 years with no recurrences.

Histological examination showed the cysts contained keratin debris, hair follicles, and some sweat and sebaceous glands, bordered by well-differentiated squamous epithelium. The histological diagnosis

was dermoid cyst in all cases.

Discussion

Twenty-five of the 94 cases of dermoid cyst of the skull treated at the same institution were located over the anterior fontanelle.¹⁰⁾ Dermoid cysts at the anterior fontanelle are almost always present at birth or in the first weeks of life and grow slowly and progressively. The cyst is entirely extracranial and has no communication with the intracranial cavity. The patients present with a slow-growing, non-tender, soft lump over the vertex of the head. They have normal growth and development and no associated neurological or systemic abnormalities.^{1,3,4,6-10)} Most cases occur in children and infants, with only a few scattered reports of adults.^{1,8)} The early reports gave the impression that dermoid cysts of the anterior fontanelle occurred predominantly in black people. However, more recent reports show that the occurrence is unrelated to racial factors.⁹⁻¹²⁾ The incidence of dermoid cysts is markedly higher than epidermoid cysts over the anterior fontanelle.^{10,13)} We have experienced no epidermoid cyst at this location. This situation can be explained by the different embryological origin of dermoid and epidermoid cysts. Epidermoids may occur when the secondary cerebral vesicles are formed and so usually occur laterally and rarely in the midline structures such as the anterior fontanelle. Dermoid cyst is more common in females, with a ratio of 2:1 in the literature.^{1,2,4,7,10,11)} All our patients were Caucasian males.

The current treatment of these lesions consists of complete surgical resection with removal of the cyst wall. The recommended technique is generally blunt dissection of the tumor from the underlying tissues such as the dura mater or cranium through a curvilinear skin incision along the plane of the coronal suture.⁹⁻¹²⁾ We preferred this technique in our cases and removed the cysts successfully with no recurrences.

The primary differential diagnosis for a mass over the anterior fontanelle is encephalocele. Gross examination may occasionally not differentiate encephalocele from dermoid cyst.^{3,4,12)} Other lesions of the scalp and the skull to be considered in the differential diagnosis include meningocele, sebaceous cyst, lipoma, cephalhematoma, lymphangioma or hemangioma, and sinus pericranii.^{1,3,12,14)} Invasive techniques such as puncture and aspiration of the mass, contrast agent injection into the mass, air ventriculography, pneumoencephalography, or air cisternography were previously used to determine the nature and the extent of the lesion.

However, the use of CT and MR imaging has abolished the necessity for these techniques.^{1,3,4,8)} CT and MR imaging are of great help in the diagnosis when compared with other methods, but the lesion must be verified histopathologically. Histological examination shows that dermoid cyst contains keratin debris, hair follicles, and sweat and sebaceous glands with a lining of stratified squamous epithelium.^{1,3,4,7-12)}

References

- 1) Adeloje A, Odeku EL: Congenital subgaleal cysts over the anterior fontanelle in Nigerians. *Arch Dis Child* 46: 95-98, 1971
- 2) Fermin S, Fernandez-Guerra RA, Lopez-Camacho O, Alvarez R: Congenital dermoid cyst of the anterior fontanel in mestizo-mulatto children. *Childs Nerv Syst* 17: 353-355, 2001
- 3) Gellis SG, Feingold M, Adeloje A: Congenital subgaleal cyst over the anterior fontanel. *Am J Dis Child* 129: 843-844, 1975
- 4) Glasauer FE, Levy LF, Auchterlonie WC: Congenital inclusion dermoid cyst of the anterior fontanelle. *J Neurosurg* 48: 274-278, 1978
- 5) Greenberg MS: *Handbook of Neurosurgery*, ed 3. Lakeland, Greenberg Graphics, 1994, 661 pp
- 6) Hamilton HB, Voorhies RM: Tumors of the skull, in Wilkins RH, Rengachary SS (eds): *Neurosurgery*, vol 2. New York, McGraw Hill, 1996, pp 1503-1528
- 7) Hayath S, Seetharam W, Kumari G, Dinakar I, Nightingale F: Congenital dermoid cyst over the anterior fontanelle. *Br J Clin Pract* 43: 119-120, 1989
- 8) Ojikutu NA, Mordi VPN: Congenital inclusion dermoid cyst located over the region of the anterior fontanel in adult Nigerians. *J Neurosurg* 52: 724-727, 1980
- 9) Özden B, Bilge T, Alatlı M: Congenital inclusion cyst of the anterior fontanelle. *The Islamic World Medical Journal (IWMJ)* April: 44-45, 1984
- 10) Pannell BW, Hendrick EB, Hoffman HJ, Humphreys RP: Dermoid cysts of the anterior fontanelle. *Neurosurgery* 10: 317-323, 1982
- 11) Sonntag VKH, Waggner JD: Congenital dermoid cyst of the anterior fontanel in a Mexican-American. *Surg Neurol* 13: 371-373, 1980
- 12) Stella L, Spaziante R, Maiuri F, Gangemi, de Divitiis E: Congenital dermoid cysts at the anterior fontanelle. Report of three cases in Italian children. *Neurochirurgia (Stuttg)* 27: 186-189, 1984
- 13) Tolia JU, Netski MG, Alexander E: Epithelial (epidermoid) tumors of the cranium: Their common nature and pathogenesis. *J Neurosurg* 23: 384-393, 1965

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