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Subgaleal Dermoid Cyst of the Anterior Fontanelle: Diagnosis with Sonography

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We report the plain film and sonographic findings in six children (2½ months to 5 years old) with a subgaleal dermoid cyst at the anterior fontanelle. Plain films of the skull made in all patients showed a soft-tissue mass superficial to the anterior fontanelle with a normal subcutaneous fat layer above it. In three children, the margins of the anterior fontanelle were beveled to match the contour of the mass. Sonography in the five children with open fontanelles showed the mass to have a well-defined sonolucent interior with a clear wall separating it from the superior sagittal sinus and other intracranial contents.

The combination of clinical evaluation, plain films, and sonography appears to be sufficient to make the diagnosis of subgaleal dermoid cyst of the anterior fontanelle.

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Congenital dermoid cyst of the anterior fontanelle is an uncommon condition that has received little attention in the radiologic literature. In this article we report the imaging results in six children with this lesion and present a brief review of the literature. Real-time sonography in five instances yielded useful diagnostic information.

Case Material

The six children varied in age at presentation from 2½ months to 5 years. Two were black, two Latin American, and two white. Four were female and two male. All had a mass over the anterior fontanelle noticed by parents in the first few months of life. At clinical presentation, the mass in all instances was nontender, broad-based, slightly movable, and fluctuant without being compressible. All patients had skull films that showed a soft-tissue mass (Fig. 1A) with smooth margins. It was located directly over the fontanelle in three patients and over the anterior corner of the fontanelle in the other three. The bony margins of the fontanelle were normal in two patients and were beveled to match the contours of the mass in another three (Fig. 2A). The remaining patient had films of the skull at 2½ months, at 4 months, and again at 5 years. At 2½ months, the soft-tissue mass was minimal. At 4 months the mass had enlarged considerably but the margins of the anterior fontanelle remained normal; at 5 years, after closure of the fontanelle, there was only slight flattening of the vault underlying the mass, without scalloping. Lateral views of the skull with low kilovolts (45 kV) to show the soft tissues (Fig. 1A) were obtained in four patients and showed that the normal subcutaneous fat layer was superficial to the dermoid. Axial CT scans of the head (Fig. 1B) in two patients showed the masses to have an interior of low radiodensity. Although beveling of the bony margins of the anterior fontanelle was shown, the relation of the cysts to underlying structures was not evaluated optimally owing to the plane of scan used.

Sonographic examination in five patients revealed a well-defined lenticular mass with sonolucent contents and posterior wall enhancement strongly suggestive of a cyst (Fig. 2B). In the four patients with an open fontanelle there was a well-defined echogenic plane between the base of the cyst and the intracranial contents, without any signs of connection between the two compartments. Doppler sonography, used in one of these patients, readily identified venous blood flow in the underlying superior sagittal sinus (Fig. 2C).

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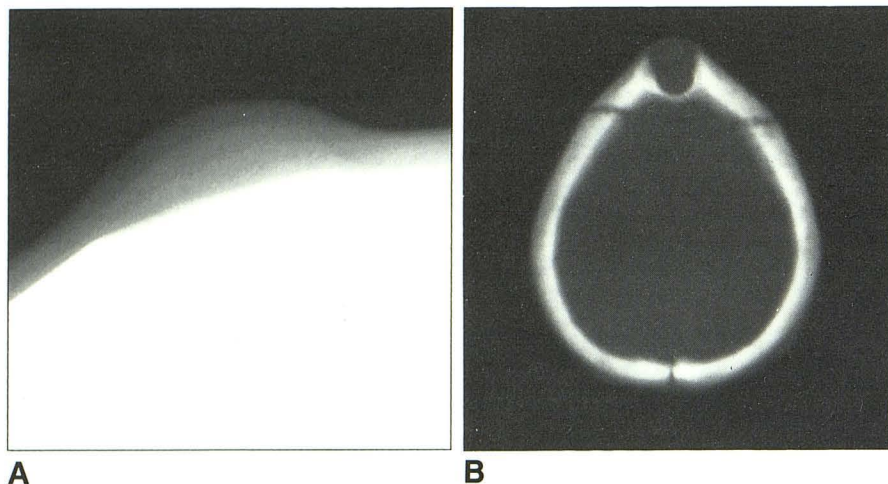


Fig. 1.—13-month-old child with dermoid cyst of the anterior fontanelle.

A, Lateral view of skull, soft-tissue technique (45 kV), shows mass located beneath normal layer of subcutaneous fat at anterior angle of fontanelle. Bony margins of fontanelle are normal.

B, Axial CT scan shows low attenuation of the cyst.

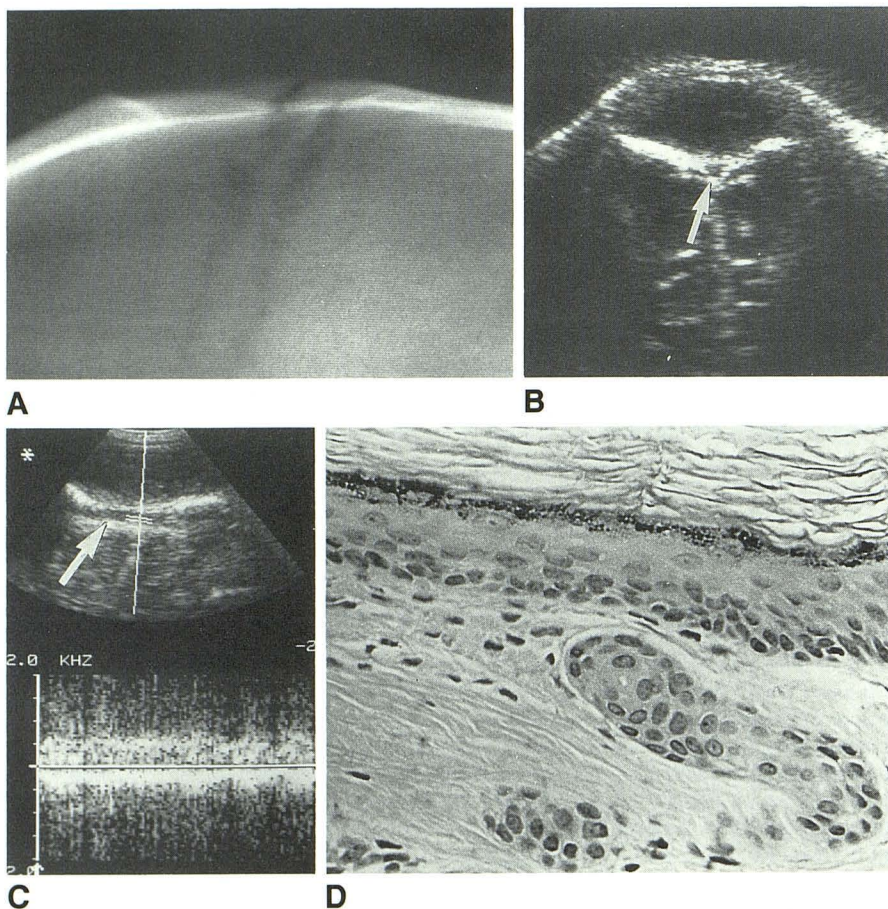


Fig. 2.—15-month-old child with dermoid cyst of the anterior fontanelle.

A, Lateral view of skull shows beveling of outer table of calvarium at level of fontanelle.

B, Coronal sonogram (5 MHz) using a standoff pad shows a lens-shaped cyst with echo enhancement of inferior wall. Deep to it lie the superior sagittal sinus (arrow) and the sulci of the medial surfaces of the cerebral hemispheres.

C, Sagittal pulsed Doppler (7.5 MHz) shows superior sagittal sinus (arrow) and continuous Doppler recording typical of venous flow.

D, Microscopic section of dermoid wall shows keratinizing squamous epithelium associated with dermal appendages.

All cysts were removed surgically and examined histologically. In some of the surgical reports, incision of the galea and separation of a small flap of underlying pericranium from the skull were described, together with the immediate proximity of the cysts to the superior sagittal sinus. All cysts had a fibrous capsule and were lined by stratified squamous epithelium. The cysts contained sebaceous and serous fluid together with keratin and dermal appendages (Fig. 2D). One specimen contained hair.

Discussion

Dermoid cysts of the anterior fontanelle were first described with histologic confirmation in 13 Nigerian patients by Adeyoye and Odeku in 1971 [1]. Since then, reports have appeared in the neurosurgical literature [2–10]. The only radiologic paper that we have been able to find is by Nicolau et al. in a French journal [7].

Dermoid cysts of the anterior fontanelle have a fibrous capsule that is lined by stratified squamous epithelium and contains skin appendages such as hair follicles, sweat glands, and subcutaneous glands. They usually contain clear fluid with some desquamated epithelial cells and keratin debris. The cysts are located beneath the galea aponeurotica and are attached by a broad base to the pericranium (Figs. 3A and 3C). Beneath the pericranium, when the fontanelle is open, is the outer or endosteal layer of the dura, which lines the calvarium and forms the roof of the superior sagittal sinus. Deep to this, in turn, the meningeal dura forms the lateral walls of the sagittal sinus. Pericranium and endosteal dura are closely applied to each other so that a dermoid may be "stubbornly adherent to the roof of the superior sagittal sinus" [8], posing difficulties in surgical dissection. Following closure of the fontanelle, bone is interposed between pericranium and endosteum (Figs. 3B and 3D). We find no record of intracranial extension of a fontanelle dermoid.

Dermoids of the anterior fontanelle are the most common dermoids of the calvarium after those of the eyebrow. They are more common in females than males (2:1). The earliest reports [1, 2, 5] suggested a predilection for black Africans but subsequent papers have recorded occurrences in patients of other races [3, 4, 7-10]. Dermoids may be noted at birth or in the first months of life. They enlarge slowly as a result of the accumulation of secretions within them. They are nontender, fluctuant, and slightly mobile but are not decompressible. They are usually 1-3 cm in diameter, and the

overlying scalp is normal. Rarely, dermoids may lie in the midline anterior to the anterior fontanelle [11].

The differential diagnosis of a mass lesion at the anterior fontanelle is limited. Meningocele and encephalocele are rare in this location [12], and should be identifiable clinically. Of the 14 histologically verified cysts reported by Adeloye and Odeku [1], 13 were typical dermoids. The remaining example had "a thick wall lined by fibrocollagenous tissue without dermal elements but with an edematous fibrocellular mass with islands of rudimentary brain tissue." This lesion was interpreted as a form of encephalocele and may be related to glial rests described in other parts of the skull [13]. At least 11 instances of melanotic progonoma at the anterior fontanelle have been reported [14-16]. There is a single case report of sinus pericranii at the anterior fontanelle [17]. Other scalp lesions such as lipoma, hemangioma, lymphangioma, and sebaceous cyst are quoted in the differential diagnosis, but no examples have been seen by us or reported in the articles we reviewed.

The diagnosis can usually be made from clinical history, physical examination, plain films, and sonography. Plain films of the skull show a water-density mass with a broad base and smooth borders without calcification. The subcutaneous fat layer at the level of the cyst is well preserved and is displaced outwardly by the cyst, a sign that is considered to be characteristic of a subgaleal lesion [7]. Lateral radiographs show the margins of the fontanelle to be either normal or beveled. In one of our cases there was no bony changes at 2½ months and only mild local flattening of the outer table at 5 years.

The value of sonography has not been evaluated previously as far as we could determine. We find this technique to be useful because it demonstrates the probable cystic nature of the lesion and, when the anterior fontanelle is patent, sonography shows the relationship of the posterior wall of the cyst to the superior sagittal sinus (especially if Doppler sonography is used). Sonography can confirm the absence of an intracranial extension of the cyst.

CT has been used in several patients in the past [5, 7-10] and was used in two of our cases. The optimal projection is the coronal plane. The interior of the cyst is generally hypodense relative to brain. The value of MR has not been fully established. In the 2-year-old girl reported by Naidich [6], the cyst was found to be isointense with brain in T1-weighted images and had a higher intensity signal in the T2-weighted images. Other investigation methods that have been used in the past such as air ventriculography, pneumoencephalography, arteriography, air cystography, and cyst aspiration are no longer considered appropriate. In the five recorded cases of infection of a dermoid cyst, three followed diagnostic air cystography.

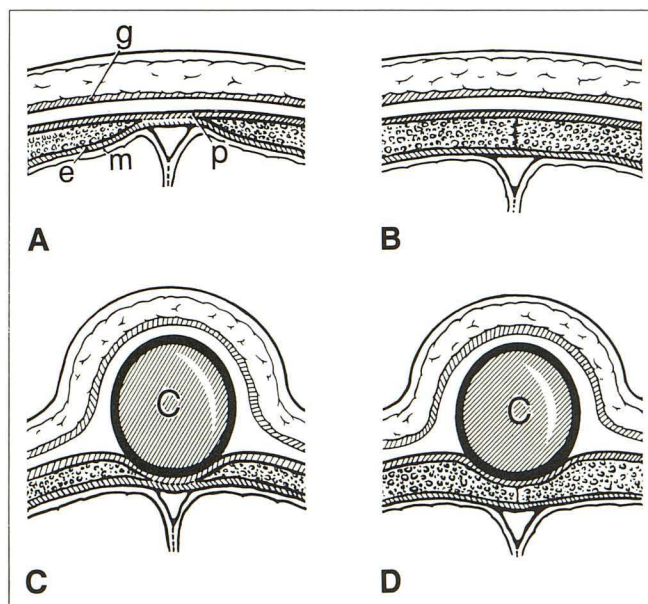


Fig. 3.—A and B, Diagrammatic representation of normal anatomy before (A) and after (B) closure of fontanelle.

C and D, Relation of dermoid cyst of the anterior fontanelle to adjacent structures before (C) and after (D) closure of the anterior fontanelle. c = cyst, e = endosteal dura, g = galea aponeurotica, m = meningeal dura, p = pericranium.

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