The Neuro Image Quiz, Answers

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This patient has a typical anterior fontanelle der-**m**oid. On T**1**-weighted images (fig. 1a) subcutaneous fat (1) has high signal intensity, so it appears as a bright crescent that defines the scalp. Bone marrow (2) is slightly less bright, and forms a crescent that defines the diploic space of the calvarium. This stripe is interrupted at the anterior fontanelle, the site of the lesion. No marrow stripe is observed in neonates, because their thin calvar-ia lack a diploic space. The outer table (3) and the inner table are cortical bone with few mobile protons. Cortical bone gives no signal, so they appear black. Such absence of signal is termed a signal void. The outer table may be identified specifically, because it lies between the bright fat and marrow stripes. Unfortunately the inner table cannot be identified as easily. On T1-weighted images, cisternal CSF (4), ventricular CSF and flowing blood also appear black. On a single T1-weighted image then, one cannot differentiate among bone, CSF and flowing blood; one cannot distinguish the precise contribution of the inner table, superior sagittal sinus (5) and interhemispheric fissure-parasagittal cisterns to the crescentic stripe of low signal that circumscribes the brain. The mass (6) is isodense with brain, so one cannot rule out an encephalocele by signal intensity alone. However, no midline pedicle of brain traverses the low signal stripe between mass and brain, making this diagnosis less likely. Nasal gli**o**ma and cysts of other etiology also cannot be ruled out, but they are far less likely choices than dermoid for a lesion situated precisely at the anterior fontanelle. The mass appears to lie within the fontanelle and adjacent bone, deep to the scalp. Posteriorly, the mass bulges over the external table. The thin stripes of high signal marrow are separated from the anterior and posterior poles of the mass by low signal cortical bone. This finding suggests that the mass sits within a well corticated – therefore chronic or congenital – defect in the bone. The deep surface of the mass lies flat against a low signal structure, suggesting that it is tightly applied to either the inner table or the subjacent dural **r**oof of the superior sagittal sinus. With increasing T2 weighting, the signal intensity of fat falls off rapidly, so the scalp appears far less bright (fig. 1b). The signal intensity of fluid spaces such as ven-

tricles and cysts increases with T2 weighting, exactly as seen here, raising the possibility that the mass is a fluid-filled cyst. However, solid dermoids may also show increased intensity with T2 weighting. The signal void of flowing blood persists, so the triangular superior sagittal sinus (6) remains black. The beveled walls of the bone fossa indent the inferior contour of the dermoid.

Figure 2 a-c are intraoperative photographs. Figure 2a: Following transverse skin incision and retraction, the periosteum (P) was denuded to expose a 2×2 cm smooth, well-encapsulated brownish-yellow midline cyst (arrow). Approximately half of the cyst lay within a deep, sharply marginated bone fossa (arrowheads).

Figure 2b: The mass was shelled out with a periosteal elevator to expose a small vessel (arrow) that penetrated the inner table, passing toward the superior sagittal sinus. The inner bone layer was otherwise intact. The bony walls of the fossa were smooth, well corticated and beveled.

Figure 2c: The pedicle was coagulated and severed, permitting the cyst to be delivered intact. Bisection of the mass revealed a smooth thin-walled cyst filled with homogeneous cheesy yellow-brown debris. At histology, the cyst lining was keratinizing stratified squamous epithelium with surrounding adnexal structures. The cyst content was keratinaceous debris.

Dermoids of the Anterior Fontanelle

Dermoids of the anterior fontanelle are the single most frequent type of dermoid seen in neurosurgical practice, representing 27% of all head and neck dermoids [5]. They usually present at birth or within the first few months of life. Females predominate 2:1 [1,2,5, 6]. Nearly all are sporadic. No family history has been reported [1]. Early literature suggested increased incidence of these dermoids in black Africans [1,3,4], but increasing experience indicates that they occur in all ethnic groups [5, 6].

Any part of the anterior fontanelle may be affected [5], but the anterior angle of the fontanelle is the most common site [5]. Identical cysts may also occur just anterior to, posterior to or at a distance from the anterior fontanelle, near to the vertex or occiput [2].

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Clinically, anterior fontanelle dermoids present as solitary, round, nontender, slightly mobile or sessile masses situated at the anterior fontanelle [1]. The overlying skin is normal with no dimple or sinus [3, 5]. Some of the lesions are fluid-filled and fluctuant. These trans-illuminate. Others are solid and opaque. The lesions may become tender with crying [1, 5]. A few (11%) are pulsatile with bruits.

Cyst size varies from 1 cm to 10 × 15 × 6 cm [17]. The lesions may enlarge very slowly over time but there is no good correlation between lesion size and patient age [5]. Affected children are otherwise normal, with normal growth and development, and suffer no concurrent malformations of brain or body. Surgical exploration documents that the mass lies in the subgaleal or subperiosteal space. It is well encapsulated and separate from the overlying skin. Typically it rests within a well corticated bone depression and has a small pedicle that attaches to the bony base or to the roof of the superior sagittal sinus. This must be coagulated and severed prior to delivery of the cyst. The inferior pole of the mass may compress the subjacent tissue slightly. It can be stubbornly adherent to the superior sagittal sinus leading to sinus damage at surgery. However, the cyst has never been reported to extend into the sinus or to extend intracranially. The mass is usually excised in toto and does not recur. Malignant degeneration has never been observed, but remains a potential. Infection of the cyst is rare and, if present, is usually iatrogenic after diagnostic cyst puncture [5].

Histologically, the lesion is a thin-walled dermoid cyst lined by keratinizing squamous epithelium containing skin appendages such as hair follicles, sebaceous glands and sweat glands. There are variable amounts of collagen in the capsule [2]. Giant cell foreign body reaction typical of cholesterol-induced inflammation may be present [4]. The cyst contents vary from clear liquid to thick white cheesy material and may evolve with age. Adeloye and Odeku [1] reported that the small cysts contained uniformly clear colorless fluid with low pH, low protein, low sodium, low chloride and high potassium. Larger cysts are yellow to yellow-brown with high pH, high sodium and chloride and low potassium, protein and glucose. The color may result from chromidrosis -

the secretion of pigment-containing sweat by apocrine sweat glands [2].

Plain radiographic findings vary with patient age and precise lesion location. In infants with open fontanelles and small cysts, radiographs may show only the soft tissue mass. In older children with closed fontanelles and larger masses, radiographs usually demonstrate a scalloped bone defect beneath the mass. The defect may be shallow, moderately deep or full thickness. The depth of the defect does not correlate with the size of the cyst and may not increase over time [2]. The shape of the depression has been postulated to be congenital [2]. Calcification is not reported, but can be expected, rarely, as a consequence of inflammation or formation of calcium soaps.

CT scans through the mass perpendicular to the skull demonstrate the intact skin, separate round well-encapsulated lesion, associated bone erosion and the exact relationship of the mass to the superior sagittal sinus.

References

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