—Case Reports—

Congenital Dermoid Cyst at the Anterior Fontanelle: Neuroimaging before and after Fontanelle Closure

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Abstract

We report on a 2-month-old boy with a dermoid cyst arising at the anterior fontanelle, with observation during both the open and closed stages of the fontanelle. The etiology of this benign, curable tumor is discussed.
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Key words: dermoid cyst, anterior fontanelle, neuroimaging

Introduction

Congenital dermoid cyst of the anterior fontanelle is an uncommon cystic lesion. We report herein a dermoid cyst arising at the anterior fontanelle. We were able to observe the case radiologically from the open to the closed stages of the anterior fontanelle.

Case Report

A 2-month-old boy was referred by a plastic surgeon for evaluation of a tumor at the anterior fontanelle. A solid spherical mass was identified, 5 mm in diameter and covered by normal scalp. The mass was soft and mobile and did not collapse on compression. Neither discomfort nor neurological symptoms were associated with the lesion. Computed tomography (CT) revealed a well-demarcated low-density tumor with no enhancement after contrast media infusion (**Fig. 1A**). With contrast enhancement, CT showed an open anterior fontanelle with the tumor at its center (**Fig. 1B**). Reconstructed CT showed that the tumor was located on the dura mater, with no communication with the superior sagittal sinus (**Fig. 1C**). Magnetic resonance imaging (MRI) showed a low-intensity mass on both T1-weighted imaging (T1WI) and diffusion-weighted imaging (DWI) (**Fig. 2**). Given its location on the superior sagittal sinus, the tumor was first merely observed. By 18 months after birth the anterior fontanelle had closed, and the location of the tumor was confirmed as being either subperiosteal or subgaleal. Three-dimensional CT revealed a depressed outer table beneath the lesion (**Fig. 3**). The size of the tumor remained unchanged. When the patient was 4 years old, the tumor was removed for cosmetic reasons. The tumor was located subgaleally, with no adhesion to adjacent structures, and complete removal was easily achieved (**Fig. 4**). The tumor consisted of yellowish fatty tissue. The histological diagnosis was dermoid cyst (**Fig. 5**). The

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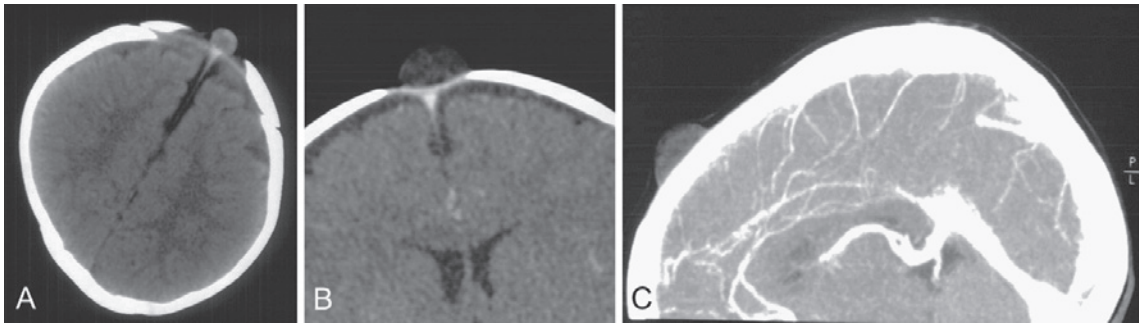

Fig. 1 A) CT demonstrating a low-density to isodense round tumor at the center of the anterior fontanelle, which is open. B) On contrast-enhanced CT, the tumor does not show enhancement and is located at the center of the fontanelle. C) On reconstructed enhanced CT, no connection is apparent between the tumor and the superior sagittal sinus.

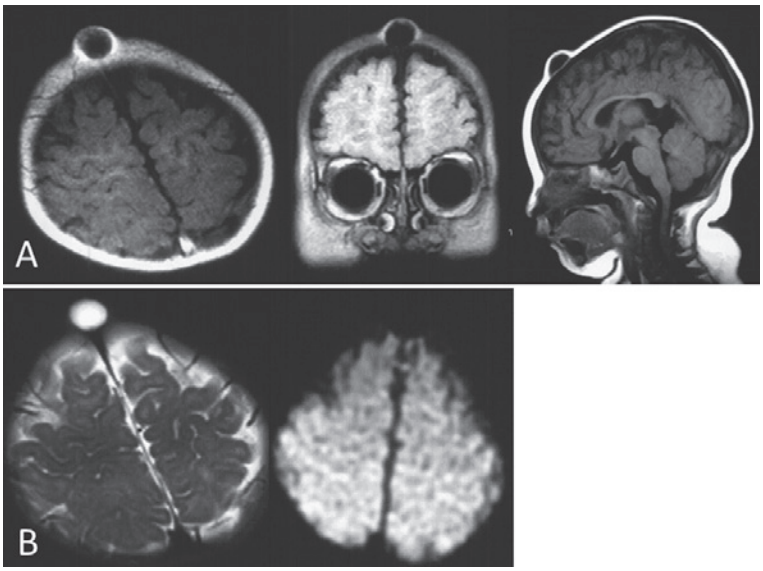

Fig. 2 A) T1-weighted imaging showing a low-intensity tumor on the superior sagittal sinus. B) T2-weighted imaging and DWI showing high- and low-intensity lesions, respectively. The tumor borders on the dura, which is continuous under the tumor.

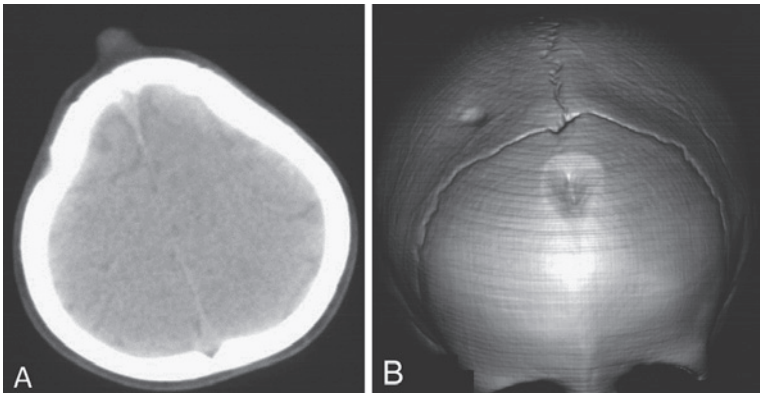

Fig. 3 A) CT after fontanelle closure, showing the tumor with unchanged size. B) Three-dimensional CT showing depression of the frontal bone where the tumor is located.

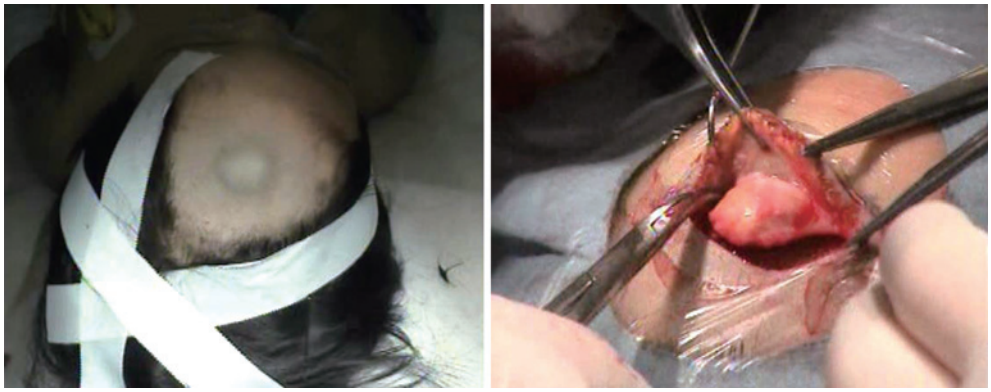

Fig. 4 Intraoperative photograph showing the well-demarcated tumor under the scalp.

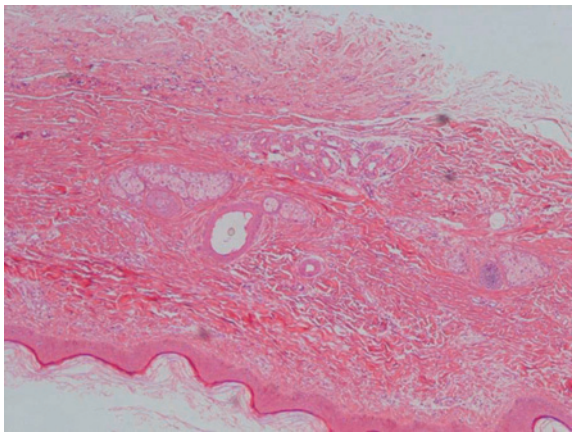

Fig. 5 Histopathological study of the anterior fontanelle tumor showing proliferation of stratified squamous epithelium with hair follicles and sebaceous and sweat glands. No malignant features are apparent. Hematoxylin and eosin, ×200.

tumor has not recurred.

Discussion

Adeloye and Odeku were the first to publish a clear and full description of anterior fontanelle tumors[1]. The majority of anterior fontanelle tumors represent congenital inclusion cysts, including dermoid and epidermoid cysts. Dermoid cysts are more frequently seen than are epidermoid cysts[2–5]. About two-thirds of anterior fontanelle tumors are dermoid cysts. A female preponderance is seen, and both dermoid cysts and epidermoid cysts are more prevalent among populations of African descent[6]. However, both types of cyst are rare among children in Asia and Europe. A total of 17 cases of anterior fontanelle dermoid cyst have been reported in Japan[5,7–13]. More cases may have gone unreported, as dermoid cysts are considered simple lesions.

Dermoid cysts and epidermoid cysts are closely related developmental tumors resulting from the inclusion of dermal elements within the neuraxis between the third and fifth weeks of embryogenesis, when the ectoderm folds into the neural tube[14,15].

Dermoid cysts are classified into 3 categories on the basis of etiology: 1) congenital dermoid cysts of the teratoma type, derived from the embryogenic epithelium, and confined to the ovary or testis; 2) acquired implantation dermoid cysts formed by cells implanted traumatically into deeper structures; and 3) congenital dermoid inclusion cysts resulting from the inclusion of displaced dermal cells along the embryonic fusion line[14,16]. In our case, the most likely diagnosis was congenital dermoid inclusion cyst, although the true pathogenesis is unclear.

Most of the time, dermoid cysts can be diagnosed at birth. CT will show the shadow of the swelling in the region of the anterior fontanelle with no change after contrast enhancement. On MRI-DWI, epidermoid cysts appear as high-intensity lesions in most cases[17,18]. Conversely, dermoid cysts usually appear as low-intensity lesions on MRI-DWI[19,20]. MRI is essential for visualizing the relationship between the tumor and intracranial structures, such as the superior sagittal sinus, dura mater, and brain parenchyma. Important differential diagnoses include encephalocele, meningocele, hemangioma, lipoma, cephalohematoma, sebaceous cyst, pilonidal cyst, and sinus pericranii[21,22], but differentiating

epidermoid cysts from dermoid cysts can be difficult, even with careful MRI observation. A final diagnosis might be established only with histological evaluation.

Dermoid cysts are known to arise intracranially as well as in the diploic space and subcutaneously. When a fontanelle tumor is discovered, the final tumor location should be predicted so that treatment options can be clarified. In the case of dermoid cyst, the location of the lesion shows a left-sided predominance, with most lesions located at the lateral angle of the anterior fontanelle and subgaleally[23]. Dermoid cysts tend to be more often located laterally and anteriorally and to be less often of epidural／intraperiosteal origin than are epidermoid cysts[24–32]. If the tumor is located subcutaneously, complete removal is easier. Subcutaneous tumors are removed for cosmetic reasons, whereas intracranial and cranial tumors do not usually need to be treated. The location of the tumor is estimated on the basis of MRI characteristics before the anterior fontanelle closes. In the case of surgical intervention, operation after fontanelle closure is safe, as in the present case.

Conclusion

We have reported a case of dermoid cyst of the anterior fontanelle. This tumor is a benign lesion that is easily treated with surgery and is free of significant complications when situated subcutaneously.

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