Analysis of the growth pattern of a dermoid cyst

Case report

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Dermoid cysts are rare lesions of the CNS with a slow rate of growth. For this reason, they are rarely discovered during infancy. Although benign, these cysts may be associated with devastating complications due to mass effect or meningitis. The discovery of completely asymptomatic dermoid cysts in the pediatric population is exceedingly rare; however, correct and prompt diagnosis is crucial for early surgical treatment to minimize morbidity and mortality. The authors report the unique case of a posterior fossa dermoid cyst discovered in a 5-month-old girl and monitored for 2.5 years with serial imaging studies before performing a resection. The imaging characteristics of dermoid cysts are reviewed, and the challenges associated with the radiographic diagnosis of such lesions are discussed. Analyzing the growth of this particular cyst on MRI allowed comment, for the first time to the authors’ knowledge, regarding the growth rate of dermoid cysts. Unlike true tumors, which grow in an exponential pattern, the dermoid cyst in the reported case exhibited a linear growth pattern. The increase in volume followed the classic appearance of a cuboid sequence, which is also consistent with linear growth in all 3 dimensions.

Case Report

History and Examination. The patient was a white female who initially came to our attention at 5 months of age

Abbreviation used in this paper: DWI = diffusion-weighted imaging.

This article contains some figures that are displayed in color online but in black-and-white in the print edition.
A stalk of tissue was emanating from the skin and eroded, revealed evidence of hair and fat within the lesion. Gross observation of the resected specimen cyst was removed en bloc, intact and without any violation. The lesion was barely in contact with dorsal brainstem. The lesion itself extended intradurally. The lesion was minimally increased in size. The syrinxes had minimally increased in size. A decision was made to observe the patient and perform follow-up diagnostic studies. When the patient was 22 months of age, only MRI of the cervical (Fig. 3) and thoracic spine was performed, which again revealed largely stable appearances of the syrinxes (41 × 15 × 10 mm). The patient remained completely asymptomatic.

At 35 months of age, the patient returned to our office for a routine follow-up visit with MRI of the cervical and thoracic spine. She remained completely asymptomatic at that time. The syrinxes had minimally increased in size. More concerning, however, was evidence of an enlarging posterior fossa lesion noted on higher cuts of the cervical spine MRI study. Dedicated MRI of the brain was performed immediately and demonstrated an enlarging midline posterior fossa lesion abutting, but not adherent to, the inferior cerebellar vermis and the dorsal brainstem (53 × 18 × 14 mm; Fig. 4). The lesion extended superficially into the diploic space and appeared to be connected to a dermal sinus tract. There was no fat signal and no evidence of contrast enhancement within the abnormality. Diffusion-weighted imaging (DWI) demonstrated strong restricted diffusion of the lesion (Fig. 5). Because the imaging characteristics were most consistent with a dermoid cyst, prompt excision was recommended; and the patient underwent lesion resection the following day.

Operation. A suboccipital craniotomy was performed. A stalk of tissue was emanating from the skin and eroding into the bone. The lesion had been pushing the dura outward, and the lesion itself extended intradurally. The lesion was barely in contact with dorsal brainstem. The cyst was removed en bloc, intact and without any violation of the capsule. Gross observation of the resected specimen revealed evidence of hair and fat within the lesion.

Postoperative Course. Immediate postoperative MRI revealed complete resection without evidence of residual lesion. The final pathological analysis was consistent with a dermoid cyst. The total length of the resected specimen was about 70 mm, although the cyst itself, which was dark red in color, was approximately 55 mm in length (Fig. 6). These findings were consistent with our aforementioned radiographic measurements. The patient had an unremarkable postoperative course and was discharged home on the 5th day after surgery.

Discussion

Dermoid cysts are benign lesions lined by stratified squamous epithelium and may contain keratin, cholesterol, sebaceous glands, hair follicles, and cellular debris. These cysts grow by incorporating desquamated cell debris. Because of their slow rate of growth, intracranial dermoid cysts are rarely discovered during infancy. One study reported a mean age of 15 years at the time of diagnosis, whereas other studies have documented the most common presentation with symptoms in the 3rd or 4th decade of life. Despite their benign nature, dermoid cysts may result in serious CNS complications, such as progressive symptoms related to local mass effect, septic or aseptic meningitis, obstructive hydrocephalus, or intracranial hypertension. Reported, intracranial dermoid cysts do not cause symptoms until the lesion exceeds 3.0 cm in diameter. In the occasional case reports describing dermoid cysts in the pediatric population, the cysts were associated with posterior fossa abscess formation and carried a high rate of mortality and morbidity. This highlights the importance of early treatment of these lesions.

Gross-total resection whenever possible remains the mainstay of treatment for dermoid cysts. However, this may not always be safely feasible; the cyst’s capsule may be difficult to remove if it is tightly attached to surrounding parenchyma, nerves, or vasculature. This can have particularly devastating sequelae if the capsule is adherent to the brainstem. Chemotherapy and radiation have no role in the management of dermoid cysts. For this reason, dermoid cysts should be resected early at the time of diagnosis before they involve surrounding neural or vascular tissue, even in patients lacking neurological symptoms.

In this case report, the posterior fossa dermoid cyst considerably increased in size in the span of 2.5 years. The cyst was attached to a cutaneous dermoid sinus tract and...
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extended in the midline all the way to the fourth ventricle. Had the lesion not been excised when it was, it would have likely extended further to involve the dorsal aspect of the brainstem, producing neurological deficits and making surgical removal difficult and highly risky. Through the successful en bloc excision of the lesion in our case, we also avoided the risk of aseptic meningitis in our patient.

Imaging Characteristics

The inconsistent appearance of dermoid cysts on MRI makes the description of classic features difficult. Commonly, they exhibit increased signal intensity on T1-weighted sequences.17,18 The appearance of dermoid cysts on T2-weighted sequences is highly variable and ranges from decreased signal to heterogeneously increased signal.16,17 In a study of 9 patients with intraaxial dermoid cysts, Park et al.18 found 42.9% of the lesions to have decreased signal on T1-weighted sequences and 57.1% to exhibit decreased signal on T2-weighted sequences. Among the 9 patients, 28.6% of the lesions had a heterogeneous appearance on MRI, which was attributed to the presence of fluid and fat components. Dermoid cysts typically exhibit no contrast enhancement, although sporadic case reports have featured dermoid cysts that enhance after the administration of gadolinium.15,17 In our case, the lesion demonstrated decreased signal on T1-weighted sequences and increased signal on T2-weighted sequences. A heterogeneous signal within the lesion was observed on all sequences. No contrast enhancement was noted, which is consistent with the most common behavior of dermoid cysts.

Dermoid cysts are known to demonstrate restricted diffusion on DWI, which is related to the fact that these lesions are commonly filled with components of ectodermal origin, such as fat and hair, resulting in decreased water proton diffusion within the lesion.19 However, this is not a consistent finding, with some case reports describing normal to decreased signal of posterior fossa dermoid cysts on DWI.17 In our case, the posterior fossa dermoid cyst exhibited strong restricted diffusion (Fig. 5).

Growth Rate

True tumors are characterized by an exponential rate of growth.25 In contrast, benign entities such as epidermoid cysts are known to exhibit a linear rate of growth at rates comparable to those for the growth of normal human skin.1 Little is known about the rate of growth for dermoid cysts, and, to our knowledge, there have been no reports in the literature about the growth pattern of these lesions. This is probably attributable to the fact that dermoid cysts tend to be excised shortly after their diagnosis, which precludes the study of their progression over an extended period of time. Our case is unique in that it sheds light on the natural history of a dermoid cyst over the period of a few years. The dermoid cyst exhibited a linear pattern of growth in all 3 dimensions. The total volume of the cyst was also measured by calculating its surface area on MRI at different levels, followed by multiplying the sum of the areas by the thickness of the cuts. The graphic representation of the increase in volume against age is demonstrated in Fig. 7 and exhibits the classic appearance of a cuboid sequence, which is consistent with linear growth in all 3 dimensions.

Conclusions

The dermoid cyst in this case report exhibited a linear pattern of growth, similar to the growth pattern described for epidermoid cysts. Radiographic diagnosis of dermoid cysts is difficult, as they can mimic other lesions. Follow-up evaluation with MRI may be required at regular in-
tervals in the case of questionable posterior fossa lesions. Early clinical diagnosis of dermoid cysts in infants and children is also difficult given the rarity of these lesions and their initial lack of symptomatology. However, correct and prompt diagnosis is crucial for early surgical treatment to minimize morbidity and mortality. Excision should be strongly considered even in patients lacking any symptoms.

Fig. 5. Diffusion-weighted MR image obtained when the patient was 35 months of age, showing strong restricted diffusion of the lesion.

Fig. 6. Photograph of the resected dermoid cyst specimen.

Fig. 7. Graph showing increase in volume of the dermoid cyst (in mm³) over time (in months).

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Disclosure

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