CASE REPORT

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CONGENITAL INTRACRANIAL FRONTOTEMPORAL DERMOID CYST PRESENTING AS A CUTANEOUS FISTULA

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Abstract: Background. Intracranial extension and a cutaneous sinus tract are rarely seen with craniofacial dermoid cysts, with few cases reported in the literature.

Methods. We report a case of a 1-year-old girl who was initially seen with a cutaneous fistula of the frontotemporal region, which revealed an intracranial dermoid cyst.

Results. The patient underwent a right lateral orbitotomy by a bicoronal approach. The cyst was seated within the lateral orbital wall, with intracranial extension through the temporal and sphenoidal bones to the dura of the temporal lobe. Histopathologic analysis confirmed the diagnosis of a dermoid cyst.

Conclusions. Craniofacial dermoid cysts may be associated with a cutaneous sinus tract and/or intracranial extension. Failure to recognize and promptly treat these lesions may lead to a progressive skeletal distortion and/or recurrent infection with a potential for meningitis or cerebral abscess. Therefore, detailed CT and MRI scans are mandatory before surgical treatment of any cutaneous fistula in the head and neck region. © 2005 Wiley Periodicals, Inc. Head Neck 27: 429–432, 2005

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Dermoid cysts are uncommon congenital or acquired developmental cystic malformations that can be found in almost every organ.1 Craniofacial dermoid cysts are typically encountered in the fronto-orbital region, especially in the upper lateral quadrant of the orbit. They are usually visible at birth or early in life as a solitary, slow-growing, subcutaneous asymptomatic mass unless inflamed or infected.1–4 They probably result from sequestration of dermal and epidermal cells along the lines of embryonic closure during fetal development and consist of epithelium-lined cysts with skin appendages such as hair follicles and sebaceous and sweat glands. Rarely, dermoid cysts of the frontotemporal region may also be associated with a cutaneous fistula and/or with an intracranial extension.5,6

We report here an unusual case of an infant with a congenital frontotemporal dermoid cyst...
presenting as a small cutaneous fistula and associated with intracranial involvement.

CASE REPORT
In November 2003, a 1-year-old girl was seen in a nearby hospital for the first time with an uninfamed cutaneous sinus of the right frontotemporal region. No mass was noticed over this region, and she was otherwise in good health. In December 2003, she underwent an exploration of the sinus under general anaesthesia without further investigation. Intraoperatively, a subcutaneous cyst was found to be eroding the lateral wall of the orbit and extending intracranially by way of a fibrous stalk. The superficial portion of the cyst was removed, and the surgeon requested a radiologic workup. The CT scan and MRI performed a few days after the operation revealed a cystic lesion within the frontal bone with a stalk extending down to the dura mater of the right middle cranial fossa (Figures 1 and 2). Microscopic examination revealed a cystic lesion of the dermoid type. The patient was then referred to our maxillofacial unit for treatment. In January 2004, we performed a second operation by means of a coronal approach. A lateral orbitotomy was used to expose the cystic tract fully as it approached the dura (Figure 3A). The stalk was completely circumscribed and excised without any dural tearing. The lateral wall of the orbit was then replaced and fixed with a resorbable plate (Polymax Resorbable Fixation System 1.5; Synthes, Oberdorf, Switzerland). The young patient fully recovered with no complications, and follow-up at 8 months showed no recurrence.

Histopathology. The submitted material consisted of eight fragments ranging from $0.2 \times 0.1 \times 0.1$ cm to $1.0 \times 0.3 \times 0.5$ cm. Some of these were fibrous, and scar tissue contained a scattered inflammatory infiltrate made of lymphocytes, plasma cells, and macrophages. Others showed an edematous stroma with a dense mixed inflammatory cell infiltrate composed mostly of neutrophils. There was no epithelial lining. Some deposits of desquamated keratin squames were found, which were compatible with remnants of a dermoid cyst content.
The previous biopsy was then reviewed. It showed a cyst formed by a rather uniform layer of stratified squamous lining epithelium, surrounded by a fibrous connective tissue layer. The lining epithelium lacked rete pegs in many areas, and the luminal surface was covered by laminated orthokeratin resting on a prominent granular cell layer. The basal layer was composed of cuboidal or cylindrical cells that showed no polarization or hyperchromatic nuclei. A few eccrine glands and ducts were present and located adjacent to, but not in contact with, the epithelium of the cyst wall (Figure 3B). The final histologic diagnosis was that of a dermoid cyst.

DISCUSSION

Described by Hirschberg as “oil cysts,” dermoid cysts represent the most common mass of the orbitofacial region in the pediatric population and, although rare, should be included in the differential diagnosis of all nodular cyst-like lesions in the head and neck region. They are benign lesions that do not contain tissue from all three germ layers, which probably represent a forme fruste of a teratoma. In 1993, Bartlett et al proposed a topographic segregation of these lesions into the following three groups: frontotemporal, orbital, and nasolabial dermoids; the authors defined a treatment algorithm based on the potential extension of the lesions to the contiguous structures (ie, meninges, orbital soft tissues, skin). They found that the frontotemporal and orbital dermoids with definable margins are superficial, slow-growing masses that can be excised, preferably by a lid crease incision, without an extensive radiologic diagnostic workup. However, the orbital dermoids with indistinct margins and nasolabial dermoid cysts required a thin-section CT scan or MRI because of their propensity to extend beyond the bone either into the orbit or intracranially. In contrast to the nasolabial dermoids, which are commonly associated with a cutaneous sinus tract and an intracranial involvement, this association rarely occurs with frontotemporal dermoids.

Cutaneous fistula as a presenting sign of a frontotemporal dermoid is very uncommon and can be deceptive. It should be promptly recognized, because it can represent the “tip of the iceberg” of a deeper and more extensive lesion. Bartlett et al, in their series of 54 cases of frontotemporal dermoids, found no similar cases. Bonavolontà et al found cutaneous fistula in only two of 145 patients analyzed in their study. Our case is unusual because of the simultaneous presence of a discharging cutaneous fistula and an intracranial extension with a fibrous stalk solidly attached to the dura of the temporal lobe. This association represents the typical scenario encountered with the nasolabial dermoid cyst, whereas it occurs exceedingly rarely with dermoid cysts of the frontotemporal region. To the best of our knowledge, only two similar cases have been reported in the literature.

There are three important considerations concerning dermoid cysts. The first consideration concerns the recurrence, which usually results from an incomplete excision, especially when the dermoid cyst extends deeply intracranially. Sessions proposed that a biopsy of the stalk be performed. If the biopsy is positive, then a combined intracranial—extracranial approach should
be used. The second consideration concerns the possible complications, including inflammation, osteomyelitis, meningitis, and cerebral abscess. Finally, the potential for malignant transformation, especially to a squamous cell carcinoma, must be considered. This may occur especially in patients with long-standing dermoid cysts, most often in cases of dermoid cysts of the ovary and rarely in cases of orbitofacial dermoids.$^{11,12}$

The orbitofacial dermoid typically presents as a soft, slow-growing, nonfixed, asymptomatic mass. However, these features may not be apparent in intracranial or deeply situated lesions. It is important to be aware that the cutaneous sinus tract of the head and neck region may represent the “tip of the iceberg” of a deep dermoid cyst. For this reason, early recognition and accurate diagnosis by means of CT scan or MRI facilitate successful treatment.

REFERENCES