Ranial dermal sinus tracts are congenital malformations that involve a cutaneous pit or dimple with variable intracranial extension. These sinus tracts are virtually always located in the midline, most commonly at the occipital protuberance, the nasion and/or nasal dorsum, or the posterior parietal midline region. The associated sinus tract may remain extracranial, it may penetrate the subjacent bone to lie either in an intradiploic or an extradural location, or it may penetrate the dura mater to end within the subarachnoid space or brain parenchyma. Associated dermoid or, less commonly, epidermoid tumors can coexist anywhere along the tract. The embryogenesis of these midline dermal sinus tracts likely involves disordered separation (or dysjunction) between the cutaneous ectoderm and neuroectoderm at the time of neurulation of the cranial neural tube during the 4th embryonic week. Occipital dermal sinuses likely involve disordered closure of the rhombencephalic neural tube, whereas nasal dermal sinuses involve disordered closure of the anterior neuropore (represented postnatailly by the lamina terminalis). Nonmidline epidermoid or dermoid scalp and skull tumors have been well described; these most commonly involve the frontotemporal region along the frontosquamosal (anterior temporal region) or frontoorbital (lateral orbital region) sutures. Most are intradiploic, occasionally with extension to the extradural space, but never through the dura. Moreover, in contrast with midline lesions, these epithelial tumors are virtually never associated with a dermal sinus tract. We report on a child born with bilaterally symmetric cranial dermal sinus tracts that extended intracranially and intradurally through the asterion. To our knowledge, this malformation has never been described previously.

Case Report

History and Examination. This 16-day-old girl, the product of a 38-week gestation, was referred to our institution with a 2-day history of poor feeding and low-grade fever. There was no antenatal complication or history of maternal drug use. At birth, two symmetrical cranial dermal sinuses had been discovered, each located approximately 2 cm behind the ear on a horizontal line between the two external auditory canals (Fig. 1). The external ostia measured 5 to 7 mm in diameter, and fine hairs projected from each. Both drained a thick, creamy, white material; the right tract additionally drained cerebrospinal fluid, indicating that the tract traversed the dura. Findings on general and neurological examinations were otherwise normal.

Blood and cerebrospinal fluid cultures were sterile. Preoperative MR imaging demonstrated two prominent retromastoid dermal sinus tracts, each with intracranial extension through a defect in the asterion (Fig. 2). Each tract extended into the cerebellar parenchyma, cleaving it into two unequal parts, with the smaller portion located laterally. There were no other intracranial masses or abnormalities.

Operation and Postoperative Course. The right sinus was explored through a retromastoid approach. An elliptical skin incision was made to incorporate the external ostium. A 1-cm bone defect was encountered at the asterion,
through which the sinus extended and penetrated the underlying dura to end blindly in the subdural space with no associated intradural mass. The left tract was excised in a similar fashion. This tract similarly penetrated the skull through a bone defect in the asterion and extended through the dura; however, the left tract passed within a fold of cerebellum and ended along the petrous ridge intracranially, posterior to the otic capsule. The stalk was transected and there was no residue or visible tract within the bone. The child is currently asymptomatic and is developing normally 1 year later.

Discussion

Nonmidline epidermoid and dermoid tumors have been well described in the diploë of the skull and in the orbit, parasellar region, cerebellopontine cistern, and lateral ventricle. These lesions, however, are not associated with dermal sinus tracts and are located either extra- or intradurally, but do not traverse the dura. Nonmidline dermal sinus tracts are exceedingly rare; a literature search disclosed only two previous case reports describing such lesions. In both cases, the tracts originated in the skin of the anterior temporal region, traversed the temporalis muscle, and ended as dermoid tumors within the diploë at the region of the pterion, but had no intradural extension.

We believe that this is the first reported case of a nonmid-
line dermal sinus tract with confirmed intradural extension and the only description of retro-auricular sinus tracts involving the asterion. Unlike midline dermal sinus tracts, the embryogenesis of these lesions is not adequately explained by an abnormality of dysjunction during neurulation. We hypothesize that the developmental abnormality that led to this malformation involves the early development of the otic placode.

Otic Development

During the 3rd week of embryogenesis, the otic placode develops as a thickening in the cutaneous ectoderm overlying the rhombencephalon. The otic placode subsequently invaginates, first to form the otic pit as a depression within the cutaneous ectoderm and then the otic vesicle as the tissue buds off and eventually separates from the overlying tissue during the 4th embryonic week (Fig. 3); the intervening stalk of ectodermal tissue connecting the two structures disintegrates during the 4th embryonic week. Later development of the cranium causes the otic placode to be translocated farther caudally, to the level of the second pharyngeal arch.

The otic vesicle subsequently differentiates into the apparatus of the inner ear, including the endolymphatic sac, cochlear duct, and semicircular canals. In the 9th embryonic week, mesenchyme chondrifies around the otic vesicle to form the periotic capsule (the petrous portion of the tempo-
ral bone); the perilymphatic duct traverses the periotic capsule to connect the perilymph and subarachnoid fluid (Fig. 3). The external auditory canal develops during the 6th embryonic week as a deepening of the first pharyngeal cleft; this deepening canal meets the tubotympanic recess (derived from the oral pharynx) to form the tympanic membrane lateral to the otic vesicle and periotic capsule.  

Proposed Embryological Mechanism for the Malformation

We propose that abnormal invagination and separation of the otic vesicle from the overlying cutaneous ectoderm bilaterally in this child led to a persistent tract of tissue between the otic vesicle and the retro-auricular skin. The attachment of the left sinus tract, on its deeper end, to the petrous bone (representing the periotic capsule) is consistent with this mechanism. The subsequent explosive growth of the underlying rhombencephalon to form the cerebellum enveloped the persistent left sinus tract (Fig. 2).

The presence of any cutaneous cranial dimple indicates the potential for an underlying dermal sinus, even if the tract is not located in the midline. It is often difficult clinically to determine the depth of a sinus; in such cases MR imaging may identify the tract and/or associated intracranial abnormalities, as in our patient. After appropriate neuroimaging studies have been performed, surgical exploration of the tract is recommended, with the goal of eliminating the communication between the skin and nervous system and removing any epithelial elements. The entire tract should be excised, along with any associated dermoid or epidermoid tumors. Early operative repair will eliminate associated secondary complications such as bacterial and aseptic meningitis, brain abscess, or growth of epithelial tumors.

References


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