Congenital Dermal Sinus

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A congenital dermal sinus tract is an uncommon type of spinal dysraphism that results when a focal area of the ectoderm fails to separate from the underlying neuroectoderm, a process termed nondysjunction. Nondysjunction prevents the insertion and subsequent development of mesodermal tissue, which normally separates the spinal cord from the skin. Congenital dermal sinus tracts occur rarely and are usually located in the lumbar or lumbosacral regions. Skin findings include abnormal pigmentation, subcutaneous lipomas, skin tags, hypertrichosis, angiomas, infection, and sinus ostia with a cephalically oriented tract, which may prompt further investigation and subsequent diagnosis. Congenital dermal sinus tracts may also be seen with other pathologic inclusion tumors such as dermoids, epidermoids, and teratomas. Treatment is surgical excision, with successful results in more than 90% of patients. To our knowledge, neither antenatal diagnoses nor cutaneous cystic findings have been reported for congenital dermal sinus tracts to date. We report a case of a dermal sinus incorrectly identified prenatally and postnatally as myelomeningocele.

Case Report

A 20-year-old woman, gravida 2, para 1, was referred at a gestational age of 30 weeks 6 days with a sonographic diagnosis of myelomeningocele. Her pregnancy was diagnosed after episodes of nausea and a positive urinary pregnancy test result. The subsequent sonographic diagnosis was made of a 10-week 4 day pregnancy. Serum triple-screen results at 19 weeks 7 days were normal, and she had a maternal serum α-fetoprotein level of 1.03 multiples of the median. The following week, she was admitted for cholecystitis and underwent laparoscopic cholecystectomy. At 24 weeks, routine sonography showed a cystic dorsal mass. She was referred to a perinatologist, whose evaluation at 26 weeks 7 days was consistent with possible lumbar meningocele without abnormal cranial anatomy. Secondary referral to our institution occurred to facilitate access to pediatric neurosurgery after
Congenital Dermal Sinus
delivery. Conventional and 3-dimensional (3D) reconstructive views were obtained to evaluate the lumbar abnormality. Fetal biometric measurements were consistent with the patient's stated gestational age and earlier sonographic examination. With the exception of the lesion described below, the remainder of the anatomic survey was normal. Specifically, the intracranial anatomy was judged to be normal with normal cerebellar peduncles and cerebellar diameters and no abnormality seen in the posterior fossa. The antrum of the occipital horn of the ventricle measured 4 mm. There was no evidence of any ventriculomegaly or unusual contours of the fetal calvarium. Additionally, both lower extremities were observed to be mobile with no evidence of any clubfoot abnormality. On evaluation of the mass, it was seen extending from S1 to just below T12 in the lumbar region. The mass could be seen just off the midline, and it measured $5 \times 4 \times 4$ cm along its greatest dimensions. The mass was mostly cystic with a central-appearing "septum." Specifically, with the 3D reconstructive views the spinous processes of L2, L3, L4, and L5 were thought to be intact in their entirety (Figure 1). Because of the conflicting data related to the intracranial structures, sonographic findings, as well as the normal serum $\alpha$-fetoprotein level, a question was raised as to whether this was truly a myelomeningocele. The patient was scheduled for a second cesarean delivery at 39 weeks, which resulted in a vigorous female neonate weighing 4120 g with Apgar scores of 8 and 9 at 1 and 5 minutes, respectively. Her recovery was complicated by postpartum anemia but otherwise was unremarkable, and she was discharged home on postoperative day 3. After resuscitation in the delivery room, the neonate was transferred to the neonatal intensive care unit, where examination of her back revealed a pedunculated mass measuring $5 \times 6$ cm (Figure 2A). No neurologic deficit was noted on the examination. Head sonographic findings were

Figure 2. A, Neonate in the nursery on day 1 of life. B, Neonatal T2-weighted midsagittal magnetic resonance imaging with radiologic diagnosis of meningocele.

Figure 1. Anteroposterior (A) and lateral 3D (B) views show the extent of the spinal mass, intact spinous process of lumbar vertebrae, and apparent central septum.
unremarkable. Spine sonography showed a normal thoracic cord with a conus located in the lumbar spine and possible dysraphism. Magnetic resonance imaging findings of the head, cervical spine, and thoracic spine were normal. Magnetic resonance imaging of the lumbar spine showed L2 dysraphism with a small posterior midline defect and a large cyst at L2 with partial internal septations (Figure 2B), resulting in a radiologic diagnosis consistent with meningocele. On the third day of life, surgical resection was undertaken. Intraoperatively, after the lamina was exposed, a cystic tract was traced down to a stalk that exited through a bifid spinous process (Figure 3). A normal-appearing dura was exposed, and the dura was opened. Neural tissue exiting from just above the conus was tethered to the most inferior aspect of the dermal sinus and was transected as it entered the conus. The cord was then untethered, and the dura was reapproximated. A surgical diagnosis of a congenital dermal sinus tract was made. Postoperatively, the neonate recovered well. Her hospital course was complicated by neonatal jaundice with a maximum total bilirubin level of 12.2 mg/dL on the fifth day of life, which responded well to 2 days of phototherapy. She was discharged on the ninth day of life.

Discussion

To our knowledge, a congenital dermal sinus tract has not been diagnosed antenatally; however, the lesion has been noted during antenatal imaging. A single case report described the prenatal and postnatal management of an occipital dermal sinus tract accompanied by hemangioma. Of note, the antenatal diagnosis reported by Viskova et al of a subcutaneous tumor without intracranial communication did not reflect the final postnatal diagnosis.

In our case, the fluid filled lumbar cyst alerted caregivers to the presence of a malformation, diagnosed antenatally as meningocele. The confusing images also contributed to the incorrect radiographic evaluation of myelomeningocele in the neonatal period. There were several findings that were not consistent with myelomeningocele. First, the cranial anatomy was normal with a normally developed cerebellum, calvarium, and ventricles. Second, each vertebra had normal transverse and spinous processes without splaying of the lamina. Maternal serum screening showed normal α-fetoprotein levels. After neurosurgical resection and the establishment of a surgical diagnosis, we were able to review the archived image volumes and were able to reproduce the above views of the dermal sinus tract labeled antenatally as a septum within a meningocele (Figure 4).

Figure 3. Surgical view of the dermal sinus showing the origin of the sinus shown as a V-shaped structure originating from within the dura.

Figure 4. Post hoc 3D reconstructive view seen from the top of the vertebral segment looking downward. The spinal canal, intact skin, and dermal sinus extending through cystic component are shown.
In conclusion, this current case reminds us that perfect prenatal diagnosis remains elusive. In this case, the classic forms of open spinal dysraphism, meningocele, and myelomeningocele were considered. In addition to a congenital dermal sinus tract, other considerations include lipomyelomeningocele and uncommon presentations of congenital tumors of the spinal cord, including epidermoid and dermoid tumors, neuroredocrine cysts, and teratomas. Importantly, having stored 3D volumes available for post hoc evaluation can be a valuable tool. However, the precise role of 3D reconstructive views or how individualized use of them is to be applied during an examination in a prospective way is yet to be fully understood.

Confusing images and incorrect or missed diagnoses with abnormalities that are rarely seen are easy to explain. As more experience is gained using volume reconstruction in prenatal diagnosis, it is likely that the exact place for its role will also be more easily determined.

References