Spinal congenital dermal sinus associated with upper thoracic meningocele

Case Report

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The congenital dermal sinus is an abnormal epithelium-lined sinus tract between the skin surface and deeper tissues. It occurs during neurulation when the neural groove closes to form the neural tube on Day 26 of gestation and results from a failure of neuroectoderm to separate from the cutaneous ectoderm. The most frequent location is the lumbosacral area; an upper thoracic location is quite rare.

This 37-year-old man presented with headache and numbness in both arms. No specific neurological findings were observed. Physical examination revealed a dimple at T-2. Radiography and magnetic resonance imaging of the thoracic spine revealed spina bifida at T1–3, a meningocele, and a dermal sinus tract complex. The treatment approach and outcome in this unusual case are presented.

KEY WORDS • congenital dermal sinus • meningocele • thoracic spine

Congenital dermal sinus is a channel lined between the skin and deeper tissues.^{4,6} It may be caused by the failure of cutaneous and neural ectoderm to separate during the closure of neural groove. Congenital dermal sinus generally occurs at one end of the neural tube and is localized most frequently in the lumbosacral area (41%), followed by the thoracic (10%) and cervical (1%) areas.^{3,4,6,10}

The incidence of dermal sinus in the population is one in 2500. Bacterial infections passing through this sinus cause symptoms. Aseptic meningitis may be observed because of pressure or rupture of a coexisting dermoid cyst and/or epidermoid tumors.^{4,5} Adults, generally considered to be asymptomatic, are rarely reported to have a complex of CDS, meningocele, and spina bifida.^{1,3}

CASE REPORT

This 37-year-old man was admitted with numbness and muscle spasms in his right arm; he also experienced headaches. A converse disorder was diagnosed and treated in intervals for approximately 2 years. The results of neurological examination were completely normal, and

there was only a dimple, existing since birth, at the T₂-weighted level. There was no history of trauma. He did have a history of headache and seizures for 3 or 4 days when he was 18 years old.

Examination. A spine radiograph revealed a spina bifida at the T1–3 level (Fig. 1). A preoperative axial T₁-weighted MR image demonstrated the presence of a hypointense meningocele at T1–3 and a T₂-weighted MR image revealed a hyperintense dermal sinus tract at T-2 and disc protrusion at C5–6 and T5–6 (Figs. 2 and 3). The results of an electromyography study were normal. No other pathological entities were observed using spinal and cranial MR imaging.

Operation. The patient was placed prone and a general anesthetic was administered. An elliptical skin incision was made around the dermal sinus and the meningocele. No sign of infection was observed. The meningocele and dermal sinus tract inside the dura were resected by microsurgery and repaired primarily.

Cerebrospinal fluid samples obtained during surgery showed no microbiological or pathological abnormalities. Histopathological examination of the surgical specimen revealed the coexistence of meningocele and dermoid sinus. Dermoid and/or epidermoid tumor did not exist in the intradural area.

Abbreviations used in this paper: CDS = congenital spinal dermal sinus; MR = magnetic resonance.

Fig. 1. Anteroposterior thoracic radiograph revealing spina bifida at the T1–3 level.

Postoperative Course. The patient was well 1 month postsurgery. He underwent control MR imaging, which revealed that the dermal sinus and meningocele had been completely resected (Figs. 4).

At the 6th postoperative month, he complained of seizures followed by weakness of the right arm and neck. The results of neurological examination and cranial fluid-attenuated inversion-recovery MR imaging were normal. Electroencephalography was performed, revealing an epileptiform disorder with a left frontal focus. He received 30 mg/kg carbamazepine daily and his seizures resolved.

DISCUSSION

Congenital spinal dermal sinus is a type of spinal dysraphism^{3,6} and a rare condition resulting from the failure of cutaneous ectoderm to separate from the neuroepithelial ectoderm to separate.^{4,6,8} Dorsal CDS is rarely localized at the thoracic level. According to the order of frequency, dorsal CDS is seen predominantly in the lumbar region (41%), lumbosacral region (23%), sacrococcygeal junction (13%), thoracic region (10%), and cervical region (1%).^{3,5,6} Wang, et al.,¹⁰ stated that they observed thoracic CDS quite rarely. To the best of our knowledge,

Fig. 2. Axial T₁-weighted MR image revealing the presence of an isointense meningocele at the level of T1–3.

Fig. 3. Sagittal T₂-weighted MR image demonstrating the presence of a dermal sinus tract at the of T-2 level as well as the presence of intervertebral disc herniation at C5–6 and T5–6.

there is no known case of an adult with CDS in the thoracic region.

Dermal sinus can be classified into five subgroups based on embryological development: 1) dermal channels in the sacrococcygeal region, extending up to the fascia, which should not be excised unless infection occurs; 2) channels frequently localized at the lumbosacral region, passing through all layers of subcutaneous tissue and involve 10 to 20% of all dermal sinuses; 3) those beginning from the skin, passing through dura, and ending at the conus medullaris, which reach intradural region; 4) those with a dermal sinus tract, passing through dura, and forming a dermoid structure where it meets the conus medullaris; and 5) dermal sinus developing from a dermoid or epidermoid structure just where it enters the intradural area.⁶ The case presented here involves a very rare complex: a dermal sinus beginning from the skin, passing through the tissue layers, and entering the intradural area with meningocele sac and spina bifida located at the upper thoracic region.

Verebely first described dermal sinus in 1913. Walker and Bucy first used the term “congenital dermal sinus” in 1934.² Congenital spinal dermal sinus presents clinically with recurrent meningitis, tethered cord syndrome, and neural compression. The most common agents involved in meningitis are *Staphylococcus aureus* and *Escherichia coli*. Morandi, et al.,⁷ presented 17 cases of CDS, 13 of the patients had meningitis and abscesses. Proteus and anaerobic bacteria were the common agents in this series. When cells proliferating in the dermal channel contain excess cholesterol crystals, aseptic meningitis occurs. Cholesterol crystals may cause severe irritation in the subarachnoid space. Therefore, aseptic meningitis may play an important role in recurrent meningitis in patients with dermal sinus.⁶ Our patient showed no signs of infection; the history of seizures and headache may have been symptoms of aseptic meningitis.

Diagnosis in cases of dermal sinus is made based on

Fig. 4. Magnetic resonance images obtained 1 month postoperatively, demonstrating total excision of the dermal sinus and meningocele.

physical and neuroimaging examinations. Physical manifestations include typical skin lesions such as lipoma, hemangioma, hair follicles, dermal sinus mount, dimple, and meningocele manqué.⁸ In patients with CDS, neurological disorders are commonly observed.⁹ Neuroimaging examinations include direct x-ray films, computerized tomography, and MR imaging. The sinus tract is best demonstrated by MR imaging.^{1,3,8}

There is no conservative treatment for dermal sinus, except as indicated in Subgroup 1 patients. Dermal sinus lesions should be surgically excised after the diagnosis is made.^{3–8} The aim of surgery must be a complete excision of dermal sinus. Broad spectrum antibiotic prophylaxis should be used even when no infection is detected. Special attention should be given to avoiding tethered cord syndrome. Cultures should be obtained when symptoms of infection occur.^{1,6}

The complex of upper thoracic CDS, meningocele, and spina bifida observed in this adult patient is a rare. This condition may be missed as a result of an asymptomatic clinical presentation. Physical examination and MR imaging are helpful in the diagnosis.

References

- Alafaci C, Salpietro FM, Grasso G, et al: Lumbosacral congenital dermal sinus presenting in a 52-year-old man. Case report. *J Neurosurg Sci* 44:238–242, 2000
- Amador LV, Hankinson J, Bigler JA: Congenital spinal dermal sinuses. *J Pediatr* 47:300–310, 1955
- Aydin K, Sencer S, Minareci O: Thoracocervical dorsal dermal sinus associated with multiple vertebral body anomalies. *Neuroradiology* 43:1084–1086, 2001
- Greenberg MS (ed): *Handbook of Neurosurgery*, Greenberg Graphics, Inc. 1994, pp 170–173
- Hattori H, Higuchi Y, Tashiro Y: Dorsal dermal sinus and dermoid cyst in occult spinal dysraphism. *J Pediatr* 134:793, 1999
- McComb JG: Congenital dermal sinus, in Wilkins RH, Rengachary SS (eds): *Neurosurgery*, ed 2. New York: McGraw-Hill, 1996, Vol 3 A, pp 3561–3564
- Morandi X, Mercier P, Fournier HD, et al: Dermal sinus and intramedullary spinal cord abscess. Report of two cases and review of the literature. *Childs Nerv Syst* 15:202–206, 1999
- Shen WC, Chiou TL, Lin TY: Dermal sinus with dermoid cyst in the upper cervical spine: case note. *Neuroradiology* 42:51–53, 2000
- Takahashi M, Murata H, Ohmura T, et al: A congenital dermal sinus presenting the muscle fasciculation and hypertrophy. *Acta Neurol Scand* 103:323–326, 2001
- Wang KC, Yang HJ, Oh CW, et al: Spinal congenital dermal sinus—experience of 5 cases over a period of 10 years. *J Korean Med Sci* 8:341–347, 1993

Manuscript received March 17, 2003.

Accepted in final form May 7, 2003.

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