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Congenital Dermal Sinus Tract of the Spine: Experience of 16 Patients

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Abstract

Congenital dermal sinus tract is a rare entity which lined by epithelial cells and can end anywhere between subcutaneous planes to thecal sac. These tracts may be accompanied with other pathologies such as lipomyelomeningocele, myelomeningocele, split cord malformation, tethered cord, filum abnormality and inclusion tumors and treatment includes resection of tract with intradural exploration. The authors review their experience with 16 cases. Clinical, radiological appearance and treatment of these lesions discussed with literature review.

Keywords

Spinal dysraphism, spinal skin lesion, tethered cord

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Congenital dermal sinus tract is a rare condition, with an incidence of 1 in 2500 live births.[1,2,4] The dermal sinus includes a tract that is lined by epithelial cells and can terminate anywhere between the subcutaneous planes and the thecal sac.[1,3-5] This pathology is thought to occur when the neural ectoderm fails to separate completely from the cutaneous surface ectoderm between the third and eighth weeks of gestation.[6-8] The major clinical presentations are skin abnormalities, infections, and neurologic deficits.[1,3-5] Congenital dermal sinus tract is commonly associated with other spinal abnormalities such as a tethered cord, inclusion tumors, and split cord malformations,[1,3,6] and treatment includes resection of tract using intradural exploration.[6] This pathology should not be confused with pilonidal sinus. The congenital dermal sinus tract is located above the gluteal crease, is cephalically oriented, and is often associated with intradural pathology. In contrast, pilonidal sinuses are located in the gluteal crease, are oriented caudally, and are not associated with intradural pathology.[7]

Methods

This study included 16 patients (10 females and 6 males) who were operated on by one of authors in the Neurosurgery Department of the Celal Bayar University School of Medicine from 1994 to 2013. Patients who were operated before 1994 by senior author Mehmet Selcuki were also included in the study. Ethics committee approval was obtained from the Celal Bayar University School of Medicine.

Results

Epidemiologic and clinical data; imaging, surgical, and pathologic descriptions; and follow-up data were examined retrospectively. The ages of the patients ranged from 10 days to 36 years, and the mean age at referral was 10 years 5 months 2 days. The mean follow-up time was 3 years (range, 6 mo to 5 y). Each patient underwent detailed neurologic and radiologic examination to determine spinal abnormalities. Four patients were older than age 16 years. Congenital dermal sinus tracts were located most frequently in the lumbar region (10 cases), followed by the lumbosacral region (5 cases; Table 1). Most patients presented with skin abnormalities with concurrent leg and/or back pain (Table 1).

Lumbar dimple and hypertrichosis were the most frequently seen skin abnormalities, each in 5 patients (Table 1). All patients had at least 1 skin abnormality, and 5 patients had 2 (Figure 1). Neurologic examinations can reveal neurologic deficits including motor weakness, sensory changes, reflex changes, and urologic and gait complaints. Four patients in our

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Table 1. Patient Parameters.

	Number (%) of Patients
Dermal sinus tract level	
Lumbar	10 (62.5)
Lumbosacral	5 (31.2)
Cervical	1 (6.2)
Thoracic	0 (0)
Sacral	0 (0)
Main presentation	
Skin abnormalities	7 (43.7)
Pain (leg and/or back)	4 (25)
Orthopedic anomalies	2 (12.5)
O leg deformity	1 (6.25)
Thinning and shortening of leg	1 (6.25)
Motor weakness	2 (12.5)
Urologic complaints	2 (12.5)
Skin abnormalities	
Lumbar dimple	5 (31.2)
Hypertrichosis	5 (31.2)
Sinus ostium	4 (25)
Abnormal pigmentation	3 (18.7)
Subcutaneous lipoma	2 (12.5)
Telangiectasia	1 (6.2)
Cerebrospinal fluid leakage	1 (6.2)
Neurologic examination	
Urinary retention/incontinence	2 (12.5)
Motor weakness	2 (12.5)
Sensory change	0 (0)
Reflex change	0 (0)
Normal	12 (80)
MRI findings	
Tethered cord	10 (62.5)
Thick filum	3 (18.7)
Lipoma	1 (6.2)
Split cord malformation type I	1 (6.2)
type II	1 (6.2)
Syrinx	1 (6.2)
Cervical myelomeningocele	1 (6.2)
Tract end points	
Dural surface	5 (31.2)
Intradural space	10 (56.2)
Attached to filum	7 (43.7)
Lipoma	1 (6.2)
Intradural abscess	1 (6.2)
Spinal cord	1 (6.2)
Epidural abscess	1 (6.2)
Intraoperative findings	
Tethered cord	12 (75)
Thick filum	4 (25)
Lipoma	2 (12.5)
Split cord malformation type 1	1 (6.2)
Split cord malformation type 2	1 (6.2)
Intradural abscess	1 (6.2)
Epidural abscess	1 (6.2)
Arachnoiditis	1 (6.2)
Myelomeningocele	1 (6.2)

study exhibited neurologic deficits: 2 had motor weakness in the lower extremities and 2 had urinary retention or incontinence (hyperreflexic and atrophic bladders were detected during urodynamic studies). During physical examination, 2 orthopedic anomalies were noted (O leg deformity and thinning and shortening of the leg; Table 1). The patient with O leg deformity also had hypertrichosis, whereas the patient with the thin and short leg had a dimple on her lumbar region.

Magnetic resonance imaging (MRI) scans were performed in all patients (Figure 2). A tethered cord and thick filum terminale were the most common spinal abnormalities, which were seen in 10 (62.5%) and 3 (18.7%) patients, respectively (Table 1). None of the patients had been operated on previously. All operations were carried out under an operating microscope (Figure 3). A midline incision was made, and the sinus tract was dissected and excised completely. Ten sinus tracts had intradural continuity and were attached to filum terminale (7 patients), lipoma (1 patient), intradural abscess (1 patient), or spinal cord (1 patient). Five sinus tracts ended on the dural surface and 1 in the epidural space (Table 1). During operative observation, tethered cord and thick filum terminale were the most frequently seen spinal abnormalities (Table 1). Twelve patients had tethered cord on intraoperative inspection, and in all cases the filum terminale was cut.

None of the 12 patients with normal neurologic examination developed additional neurologic deficits, and those with existing motor weakness or urologic complaints were unchanged. Leg and/or back pain (complaints of 4 patients) improved during the postoperative period. Two patients suffered a cerebrospinal fluid leakage, and revisions were performed. A comparison of the characteristics of the patients in this study with those in previous reports is shown in Table 2.

Discussion

The incidence of congenital dermal sinus tract is 1 in 2500 live births,[1,2,4,7] and few published reports describe affected patients.[1,3-7] The condition is thought to result from the failure of the neuroectoderm to separate from the cutaneous ectoderm at the end of the neurulation and can found anywhere along the midline of the neural axis.[3,6,7] Selcuki et al reported that inhibition of apoptosis could cause embryologic errors that accounted for maldevelopment of the congenital dermal sinus tract.[2] Although congenital dermal sinus tracts are usually seen in the midline, they can also be detected in off-midline locations.[8,9] Dermal sinuses occur frequently at the lumbar or lumbosacral regions and account for 40% and 45% of all cases, respectively.[1,4,5] In contrast, cervical and thoracic regions are reported in <1% and 10% of cases, respectively.[3] Dermal sinus tracts reach the dura mater and thecal sac in 60% of patients. However, they can also end in the epidural space and soft tissue in 10% to 20% and 6% to 7% of patients, respectively.[1,3] Skin abnormalities such as dimples, hypertrichosis, ostium, abnormal pigmentations, subcutaneous lipomas and erythemas are common complaints at presentation[1,3-6] (Figure 1). Although none of these are diagnostic alone, a combination of 2 or more midline skin lesions strongly suggests a dermal sinus tract.[4] Neurologic deficits, orthopedic anomalies, urologic complaints, and infections are other complaints at presentation.[1,3-5] Martinez-Lage et al[5] reported that 8 of 12 patients presented with skin abnormalities in their study.

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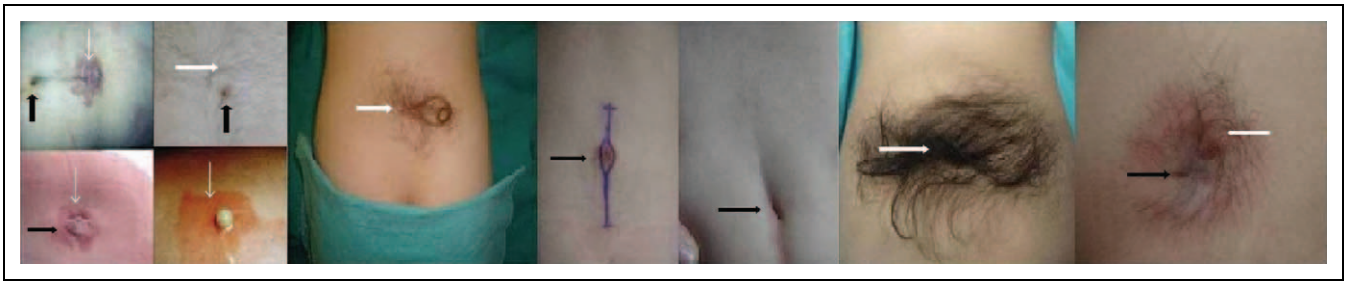

Figure 1. Skin abnormalities resulting from congenital dermal sinus tract. Thick white arrows show hypertrichosis, black arrows show sinus ostiums, and thin white arrows demonstrate telangiectasia.

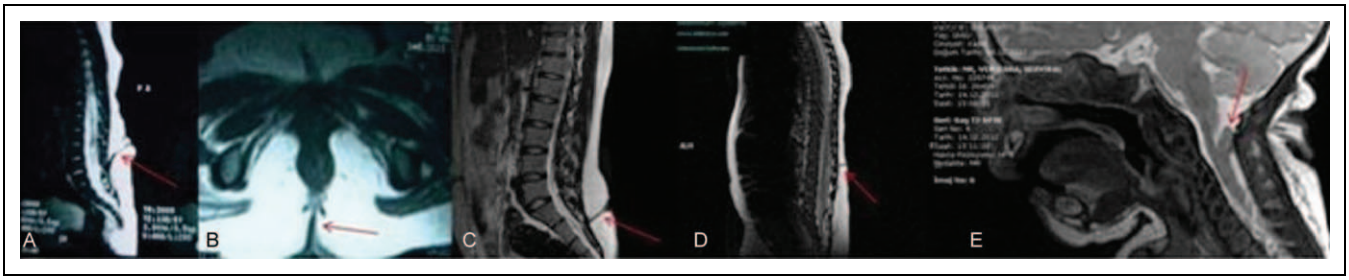

Figure 2. Sagittal and axial lumbar magnetic resonance images (MRIs) demonstrated (red arrow) an apparent lateral sinus tract originating from the midline skin surface and extending down to the spinal canal (A-D). The red arrow (E) shows a cervical myelomeningocele in which we noted dermal sinus tract during surgery that was attached to the spinal cord.

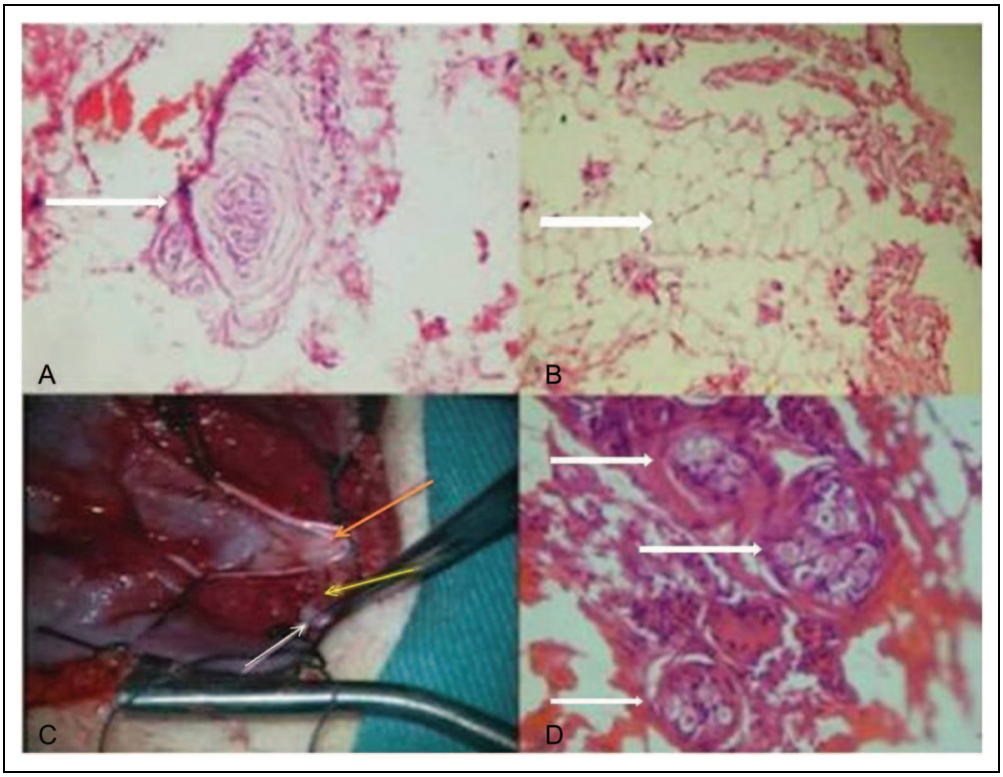

Figure 3. Dermal sinus tract ending intradurally, and attached to the thick fatty filum terminale. The white arrow shows the starting point of the tract in the skin, the yellow arrow shows the internal dural section, and the orange arrow shows the attachment point of the dermal sinus tract (end point) (C). During histologic examination, the white arrow shows a Meissner body (as evidence of dermal tissue) at the starting point of the tract (A). The white arrow shows intense fat tissue in the sections of thick fatty filum terminale (B). White arrows show the axial cut view of peripheral nerve fibers in sections of the dermal sinus tract in the dural region (D).

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Table 2. Comparison of the Characteristics of Patients in the Present Study and Those in Previous Reports.

	Jindal (2001)	Elton (2001)	Ackerman (2003)	Radmanesh (2009)	Martinez-Lage (2011)	De Vloo (2012)	Present Series
No. of patients	23	23	28	35	20	14	16
No. of patients aged >16 y	5	–[a]	2	–	–[a]	–	4
Male/female	8/15	13/10	11/17	16/19	13/7	6/8	6/10
Mean age at referral	10 y 2 mo	6.6 y	9 mo	1 y 1 mo	2 y 11 mo	2 mo	10 y 5 mo 2 d
Duration time	7	19	30	8	30	26	10
Primary reason for presentation							
Skin abnormality	3	–[a]	15	20	15	10	7
Motor weakness	20	–[a]	8	2	9	–	2
Urologic complaints	–	–[a]	–	1	–	2	2
Low-back pain	–	–[a]	2	–	–	–	4
Orthopedic abnormality	–	–[a]	5	1	–	–	2
Infection	–	–[a]	3	11	5	2	–
Clinical examination							
Cutaneous findings	–[a]	–[a]	27	20	15	10	16
Neurologic deficit	–[a]	–[a]	19	13	9	2	4
Motor weakness	23	–[a]	11	8	6	–	2
Sensory change	20	–[a]	7	–	7	–	–
Reflex change	11	–[a]	15	–	12	2	–
Urologic-gait abnormality		–[a]	15	6	8	–	2
Orthopedic abnormality	12	–[a]	5	4	–	3	2
Pain	6	–[a]	3	5	–	1	4
Operative findings							
Tethered cord	–[a]	8	22	22	13	3	12
Dermoid-epidermoid	–	6	5	15	8	3	–
Conus angioma	9	–	–	–	1	–	–
Thick filum	–	–	–	14	8	–	4
Lipoma	5	2	8	5	–	7	2
Split cord malformation	–	3	3	5	–	2	2
Abscess	6	–	–	9	2	2	2
Arachnoiditis	–	–	6	13	–	–	1
Myelomeningocele	–	2	–	–	–	–	1
Cerebrospinal fluid leakage from sinus tract	–	–	3	–	–	–	–
Nerve root herniation from tract	–	–	2	–	–	–	–
Syrinx	–	–	–	1	–	–	–

[a]Authors did not describe this information clearly.

Consistent with this, De Vloo et al[4] also reported 10 of 12 patients presented with cutaneous findings. Skin complaints were also the predominant features at presentation in studies by Ackerman et al[7] and Radmanesh et al.[1] In contrast, Jindal et al[3] reported that most patients (20 of 23) presented with neurologic deficits. In the same study, most patients (10 cases) had lumbar congenital dermal sinus tract, followed by deficits in the lumbosacral and cervical region (5 and 1 cases, respectively). In our study, most patients (7 cases, 43%) presented with skin abnormalities (Figure 1) and intradural ending (10 patients, 56%).

Dermal sinus tract can be accompanied by other pathologies such as lipomyelomeningocele, myelomeningocele, split cord malformation, tethered cord, filum abnormalities, and inclusion tumors.[1,3,7] Gupta et al[10] showed an association of 11.34% between dermal sinus and other spinal malformations. In our study, only 1 patient (6.2%) with cervical dermal sinus tract had myelomeningocele. The coexistence of congenital dermal sinus tract with other pathologies was reported at different rates. Ackerman et al[7] described the occurrence of congenital dermal sinus tract together with tethered cord and intradural masses in 79% and 50% of patients, respectively. Jindal et al[3] reported the comorbidity of filum abnormality in 22% and split cord malformation in 25% of patients. In contrast, these respective ratios were 40% and 15% in a study by Radmanesh et al.[1] In the same study, the coexistence of tethered cord and intradural masses with congenital dermal sinus tract was reported in 63% and 42% of patients, respectively.[1] In our study, tethered cord occurred in 12 (75%), thick filum in 4 (25%), and split cord malformation in 2 (12.5%) patients during intraoperative observation.

Although Jindal et al[3] reported 5 dermal sinus tract patients older than 16 years of age, Ackerman et al[7] reported only 2 (Table 2). In our study, there were 4 dermal sinus tract patients older than 16 years of age; all were female (ages 20, 20, 26, and 36 y). Three of the 4 presented with back and leg pain, with a pain duration ranging from 3 to 7 months. The remaining patient was referred to us due to a skin abnormality. In our physical examination, 2 of these 4 patients had hypertrichosis, 1 had telangiectasia, and 1 had a lumbar dimple. However, all 4 neurologic examinations

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were normal. The symptoms of tethered cord syndrome in 3 of the 4 patients could be related to the dermal sinus tract exerting traction on the spinal cord and cauda equina. The spinal column grows more rapidly than does the spinal cord, which starts on the 55th day of gestation soon after the completion of primary and secondary neurulation. If a process during either primary or secondary neurulation failed, which causes neural tissue to become attached to surrounding tissues, the tethering of the cord is already established by the time of birth. This explains why surgical interventions should be carried out as prophylactic measures before the appearance of tethered cord-related symptoms. Pool[11] theorized that a tethered cord eventually became symptomatic in 1 older patient because of local stretching and ischaemia from the repetitive and insidious lower back trauma, resulting in delayed diagnosis. However, an important question is whether the diagnosis was delayed, or surgery was not considered. Three of 4 patients in our study were referred to a neurosurgeon by other physicians because of a dermal sinus tract on MRI, but surgical intervention was not considered. In addition, some neurosurgeons do not use surgery in the presence of a dermal sinus tract if the patient has no complaints. This is likely because the importance of pediatric neurosurgery and the concept of tethered cord, which exists at the time of birth, are not fully understood. Rajpal et al[12] reported that patients who never undergo treatment for tethered cord syndrome have an elevated risk of developing symptoms with advancing age. The final patient in this group in our study was from a low socioeconomic background, and she was not admitted to a neurosurgery department until her primary care physician referred her to us because of hypertrichosis on her back. We suggest that untethering intervention should be performed as soon as possible, and in particular as a prophylactic procedure before the tethered cord and urinary incontinence symptoms appear. Urinary incontinence, one of the most challenging and inconvenient symptoms of a patient with tethered spinal cord, is unfortunately also one of the most difficult to treat.

In radiologic examinations, the radiograph of the spine has a limited role in the evaluation of a congenital dermal sinus tract,[1,3] and so MRI is the first-choice modality for diagnosis. The sinus tract appears typically as a low-intensity tract that ascends into the subcutaneous tissue with a high intensity on both T1- and T2-weighted images (Figure 2). MRI allows physicians to identify any associated anomalies, such as low-lying conus medullaris, and also assist in preoperative planning.[1,3,7] However, a normal MRI does not exclude the diagnosis of a sinus tract, because only 40% of tracts were detected in preoperative MRI.[7] Therefore, if the MRI is normal and the tract is above the gluteal crease, surgical exploration is recommended.[6,7]

Treatment of dermal sinus above the sacrococcygeal region requires complete excision of the tract. If the MRI reveals normal findings and the dermal sinus tract is located above the gluteal crease, the tract should be explored to its terminus because sinus tracts above the gluteal crease (above the sacral 4 vertebra level) are more likely to be contiguous with the dura. If intradural lesions such as a tethered cord or intradural masses are present, exploration must proceed into the intradural place, otherwise reexploration will be necessary.[1,6]

Conclusion

If a cutaneous lesion is identified over the midline neural axis, radiologic evaluation should be performed for diagnosis. If a dermal sinus tract is detected, surgical treatment should be applied as a prophylactic measure as soon as possible to prevent complications secondary to infection, tethered cord, or other pathologic conditions, such as an inclusion tumor mass. At surgery, the tract should be followed intradurally and all accompanying malformations with possible tethering effects should be corrected.

Author Contributions

MM, along with MS, wrote the first draft of the manuscript and made corrections according to the reviewers' comments. MM and ASU analyzed the data. YKD prepared the figures and MB, the tables. NU and SGG performed histopathologic evaluation.

Authors' Note

The study was done in Celal Bayar University School of Medicine Department of Neurosurgery, Manisa, Turkey.

Declaration of Conflicting Interests

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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Ethical Approval

Local ethics committee approval (No. 20478486-248) was obtained from the Celal Bayar University School of Medicine.

References

1. Radmanesh F, Nejat F, Khashub ME. Dermal sinus tract of the spine. *Childs Nerv Syst*. 2010;26:349-357.
2. Selcuki M, Vatansever S, Umur AS, et al. Apoptosis seems to be the major process while surface and neural ectodermal layers detach during neurolation. *Childs Nerv Syst*. 2008;24:577-580.
3. Jindal A, Mahapatra AK. Spinal congenital dermal sinus: an experience of 23 cases over 23 years. *Neurol India*. 2001;49:243-246.
4. De Vloo P, Lagae L, Sciot R, et al. Spinal dermal sinuses and dermal sinus-like stalks analysis of 14 cases with suggestions for embryologic mechanisms resulting in dermal sinus-like stalks. *Eur J Paediatr Neurol*. 2013;17:575-584.
5. Martínez-Lage JF, Almagro MJ, Ferri-Ñiguez B, et al. Spinal dermal sinus and pseudo-dermal sinus tracts: two different entities. *Childs Nerv Syst*. 2011;27:609-616.
6. Elton S, Oakes WJ. Dermal sinus tracts of the spine. *Neurosurg Focus*. 2001;10:e4.
7. Ackerman LL, Menezes AH. Spinal congenital dermal sinuses: a 30-year experience. *Pediatrics*. 2003;112(3 Pt 1):641-647.
8. Yamaguchi S, Takeda M, Kihara H, et al. Lateral buttock congenital dermal sinus tract. *Neurol Med Chir (Tokyo)*. 2011;51:460-462.

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9. Ikwueke I, Bandara S, Fishman SJ, Vargas SO. Congenital dermal sinus tract in the lateral buttock: unusual presentation of a typically midline lesion. *J Pediatr Surg*. 2008;43:1200-1202.
10. Gupta DK, Shastank RR, Mahapatra AK. An unusual presentation of lumbosacral dermal sinus with CSF leak and meningitis. A case report and review of the literature. *Pediatr Neurosurg*. 2005;41:98-101.
11. Pool JL. Spinal cord and local signs secondary to occult sacral meningoceles in adults. *Bull N Y Acad Med*. 1952;28: 655-663.
12. Rajpal S, Tubbs RS, George T, et al. Tethered cord due to spina bifida occulta presenting in adulthood: a tricenter review of 61 patients. *J Neurosurg Spine*. 2007;6:210-215.

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