Journal of Neuro-Oncology **47:** 271–281, 2000.

Intramedullary spinal tumors of disordered embryogenesis

Karin Muraszko[1,2] and Andrew Youkilis[1]
[1]*Departments of Pediatric Neurosurgery and* [2]*Neurosurgery, University of Michigan, Ann Arbor, MI, USA*

Key words: embryogenesis, intramedullary, spinal tumors

Summary

Abnormal spinal embryogenesis is quite commonplace. While greater than 90 percent of these errors of embryogenesis leads to occult spinal dysraphism with minimal neurologic or orthopedic sequelae, there is a significant minority of these anomalies which leads to the formation of the so-called 'congentital tumors of disordered embryogenesis'. The purpose of this article is to discuss the embryology, presentation, diagnosis and management of the spinal dysraphic states with particular emphasis on those errors which lead to mass lesions in the spinal canal such as dermoids, epidermoids, lipoma/lipomyelomeningocoele and neurenteric cysts. We also include lesions such as dermal sinus tracts and thickened filum terminale in our discussion with particular emphasis on their relationship to the tethered cord syndrome. Proper surgical management of these various conditions necessitates a thorough understanding of their embryologic etiology and the anatomic/physiologic ramifications that such lesions have on the developing spinal cord.

Spina bifida

Spina bifida refers to any developmental abnormality of the vertebrae or spinal cord. It can include such minor abnormalities as an absent spinous process or incomplete fusion of a lamina, to the more extreme abnormality of myelomeningocele and its associated abnormalities of the Chiari II malformation, hydrocephalus and abnormal brain development. Spina bifida can be divided between those forms which are considered open (myelomeningocele) versus those which are considered closed (lipomas of the spinal cord, diastematomyelia, etc.). We will discuss various subcategories of spina bifida which can create so called 'congenital tumors' such as lipomas, dermoids/epidermoids and neurenteric cysts.

Occult spinal dysraphism

Occult spinal dysraphism is comprised of a variety of malformations from such simple spina bifida occulta to more complex spinal cord abnormalities. Failure of complete formation of the lamina and spinous processes without orthopedic or neurological deficit is not uncommon. The radiological incidence of spina bifida occulta in children less than 8 years of age is as high as 49 percent at the S1 area, 13.5 percent at L5 and 9.1 percent at both L5 and S1. Studies of the general population suggest that 20–30 percent have such incidental findings on x-ray. In such cases, the overlying skin is intact and the neurological examination is normal.

Embryology

Approximately 2 percent of live births have some form of major congenital anomaly and of those involving the central nervous system, 64 percent involve some aspect of abnormal closure of the neural tube [1–3]. After fertilization of the egg, there is rapid cellular division creating a hollow sphere of cells called a blastocyst. Located eccentrically within the blastocyst is an inner layer of cells called the embryonic cell proper and out layer of cells, the trophoblast. Following this there is a period of differentiation and specialization which produces an oval embryonic disc which contained three clearly differentiated germ layers of endoderm, ectoderm and mesoderm. The outer layer of the embryonic disc is composed of ectoderm and the inner layer of endoderm. Cells begin to migrate inward from each side of the primitive streak, between the

layers of ectoderm and endoderm to form the paraxial mesoderm.

The notochord formation begins at approximately 16–17 days of gestation, as the primitive streak begins to regress, and occurs when cells from Hensen's node migrate between the sheets of mesoderm, and the ectoderm and endoderm to form a midline longitudinal cord of cells. This notochord then hollows into a cylinder of cells by day 18 and becomes incorporated into the underlying endoderm.

Formation of the neural tube occurs in 3 distinct phases. *Neurulation* occurs at days 18–28, *canalization* of the tail bud occurs at 28–40 days and *regression* or dedifferentiation occurs from 41 days onward [4,5]. The neural groove is shallow midline fold and is adherent to the notochord throughout the neural tube formation. Once the neural tube is closed, the notochord separates from it. The neural plate is formed as the midline ectoderm thickens cephalad to Hensen's node. It is this ectoderm which will eventually form the neural plate. At the same time that the neural plate is developing, the paraxial mesoderm thickens and begin to form paired segments of mesoderm called somites, along either side of the notochord. Approximately 30 pairs of somites will form by day 28 of gestation and each of these will eventually form a cavity called a myocoele which will be a sphere of mesodermal cells. The dorsomedial portion of each myocoele will go on to be a myotome and will form the skeletal muscle. The ventromedial portion is called the sclerotome and these will condense around the notochord/neural tube to form the vertebral body and posterior spinal elements. The ventrolateral portion of the myocoele is called the dermatome and will form the connective tissue and muscles of the body wall [6].

Around 22–24 days of gestation, the neural groove deepens and at somites 1–7 begins to develop prominent neural folds [7]. These folds meet in the midline to form the future lower rhombencephalon. It is this fusion of neural groove folds which transforms the flat neural plate into the hollow neural tube. Fusion of these folds is called neurulation. The fusing neural folds induce the superficial ectoderm at the lateral edges of the neural plate to come towards the midline and then fuse to the neural tube. It is at this point that the surface ectoderm separates from the neural tube and forms the epidermis. The mesoderm now interposes itself between the epidermis and the neural tube. This fusion of tissue occurs in a stepwise fashion. It begins in the mid-dorsal region of the embryo and proceeds simultaneously in the cephalad and caudal directions. The entire process of neurulation occurs between 22 and 28 days [8,9].

There are two areas in which there is a delay of closure of the neural tube. At the cephalic end at the level of the lamina terminalis, also called the rostral neuropore, the closure occurs at 24–25 days. At the caudal end of the neuropore the final closure occurs at 26–28 days [6,9]. An important date in the formation of the neural tube is day 28. By day 28 the neural tube is now formed and the brain is now visible. The various somites have arranged themselves along the neural tube and notochord in preparation for the formation of the vertebral bodies, the intervertebral discs and the paraspinal muscles. Closure or occlusion of the neural tube occurs adjacent to somites, suggesting that the somites are important in initiation of this closure. The caudal neuropore is thought to be at approximately the L1/L2 level. Thus neurulation appears not to involve the lowest portion of the spinal canal. This distal canal is thought to form via process of canalization.

Ectoderm covers the neural tube as the caudal neuropore closes. Caudal to the neuropore an undifferentiated cell mass arises. This cell mass then begins to hollow or canalize beneath an intact surface layer of ectoderm. Unlike neurulation which is precise, canalization seems to have a variety of variations resulting in a variety of distal or caudal spinal abnormalities.

Up until 9 weeks of development, the vertebral column and the developing spinal cord match segment for segment. After that time, the neural tube begins to ascend with respect to the vertebral column thus promoting the development of the cauda equina and the filum terminale [10]. The filum terminale develops as a result of cellular death of the distal neural tube and is formed at around 40–48 days of gestation. A terminal ventricle forms just cephalad to the filum terminale and is at the caudal limit of the spinal cord or conus medullaris. The coccygeal medullary vestige is a rudimentary ependymal cavity which is located within the distal end of the filum terminale. There is disproportionate growth of the vertebral column with respect to the spinal cord such that at 9 weeks the conus is located at the first to third coccygeal segments but by 17 weeks is not at the fourth lumbar vertebral body. It continues ascend more slowly after this and is located at birth at the third lumbar vertebral body and in adulthood at the L1 to L2 intervertebral disc space.

The neural crest cells originate from the neural ectoderm located at the apex of the neural folds. The form bilateral cell columns along the dorsolateral aspect of the neural tube. These neural crest cells will eventually

segment and give rise to the dorsal root ganglia of the cranial and spinal nerves, the sympathetic ganglia of the autonomic nervous system as well as the chromaffin cells of the adrenal glands, the carotid bodies and the pigment layers of the retina. These cells also contribute to the pia, the arachnoid and the sheath of Schwann cell of the peripheral nerves. It is these neural crest cells that are believed to be associated with the various cutaneous lesions seen in association with spinal dysraphism such as hemangiomas or skin discoloration.

Tethered cord syndrome

The tethered cord syndrome refers to a variety of lesions which can cause the conus medullaris to be low lying or incapable of oscillating within the spinal canal [11–13]. More recently a form of tethered cord has been described in which there is no radiographic evidence of tethered cord (no lipoma or thickened filum, no low lying conus) but only symptoms of tethered cord. This latter problem of a tethered cord with a normal positioned conus remains controversial, especially as to the need for surgical intervention [14–16]. The more common problem is one in which the conus is low lying and there may be an associated thickened filum terminale, fibrous bands or intradural lipoma [17].

The embryological origin of the various forms of tethered cord syndrome remain controversial. In the case of lipoma, it is assumed that there may be inclusion of adipose cells early in the development of the neural tube. A thickened filum terminale may result as an abnormality of the normal caudal canalization and regression which occurs in the terminal bud. Studies by Yamada have shown that, in a adult cat model, traction on the spinal cord produces elongation of the spinal cord and filum and was accompanied by significant reduction in spinal cord blood flow and evoked potential activities, as well as reduced mitochondrial oxidation of cytochrome a,a_3 [12,18–21].

Cutaneous abnormalities may be seen in as many as of these [22]. Clinical symptoms in such patients include gait manifestations, urological dysfunction including recurrent urinary tract infections, scoliosis, pes cavus deformity, pain and sensory changes. Increasingly such patients are found before symptoms present because of radiological investigation of a cutaneous abnormalities such as a dimple, hemangioma, hypertrichosis, skin appendage, subcutaneous lipoma or associated abnormalities such as imperforate anus (Figure 1A,B) [17,23–25]. The onset of symptoms may be quite gradual and require careful neurological

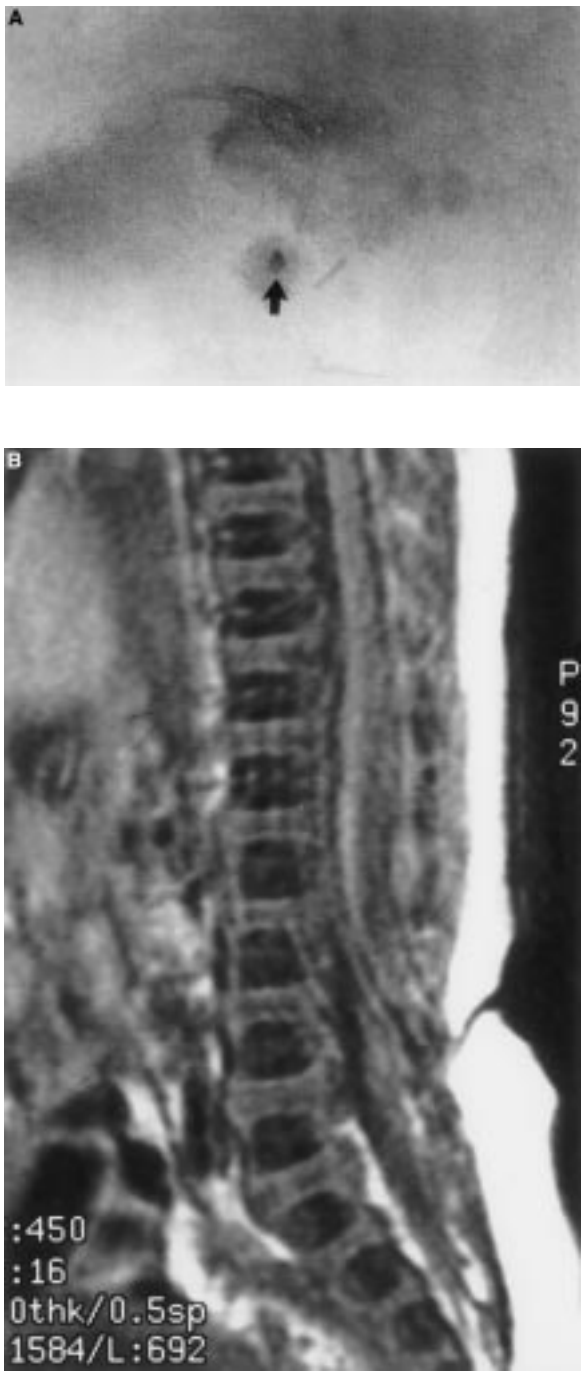

Figure 1. A: Dimple with associated hemangioma and tuft of hair. Arrow points to pit in skin. B: Sagittal MRI image of dermal sinus tract extending intradurally and attaching to filum terminale.

examination [26]. There is evidence that such patients may present during growth spurts or in the adult population after minor trauma or during lithotomy positioning during labor [27–29].

Motor weakness is the most common presenting symptom and is seen in 76 percent of these patients [26,28]. It may be asymmetrical with one leg being smaller and weaker. Sensory loss is relatively uncommon though pain is the initial complaint in 42 percent. Urological problems prompt evaluation in 35 percent of thesie patients [11,30–32]. Orthopedic deformities are varied and include pes cavovarus deformity, gait disturbance and scoliosis [33].

Surgical treatment of these lesions is recommended even if symptoms have not occurred to prevent neurological deterioration [34]. The complexity of the operation depends upon the pathology encountered. A thickened filum can be approached via a limited laminectomy. Stimulation of the filum to assure that there are no adherent nerve roots or functional neural elements is important before dividing the filum (Figure 2A,B). Monitoring of rectal tone via balloon, or rectal sphincter activity via electrodes can be helpful. Meticulous dissection of the neural elements is necessary to separate them from the dura, fibrous bands, thickened filum or intradural lipoma [35]. A lipoma in these situations should not be viewed as tumor, rather as a part of the spinal cord. The lipoma should be separated and freed from the dura. A laser is particularly helpful in decreasing the bulk of the lipoma. Separation of the lipoma from the spinal cord is dangerous and not necessary. The key to successful untethering is to free the spinal cord from the dura.

Early diagnosis and prompt surgical attention provide the best results in patients with tethered cord [36]. Some 85 percent of patients have improvement in motor function, slightly less have improvement in bladder function, and greater than 95 percent have stabilization of their neurological exam [37–39]. Hoffman has found that of the 30 percent of patients who had

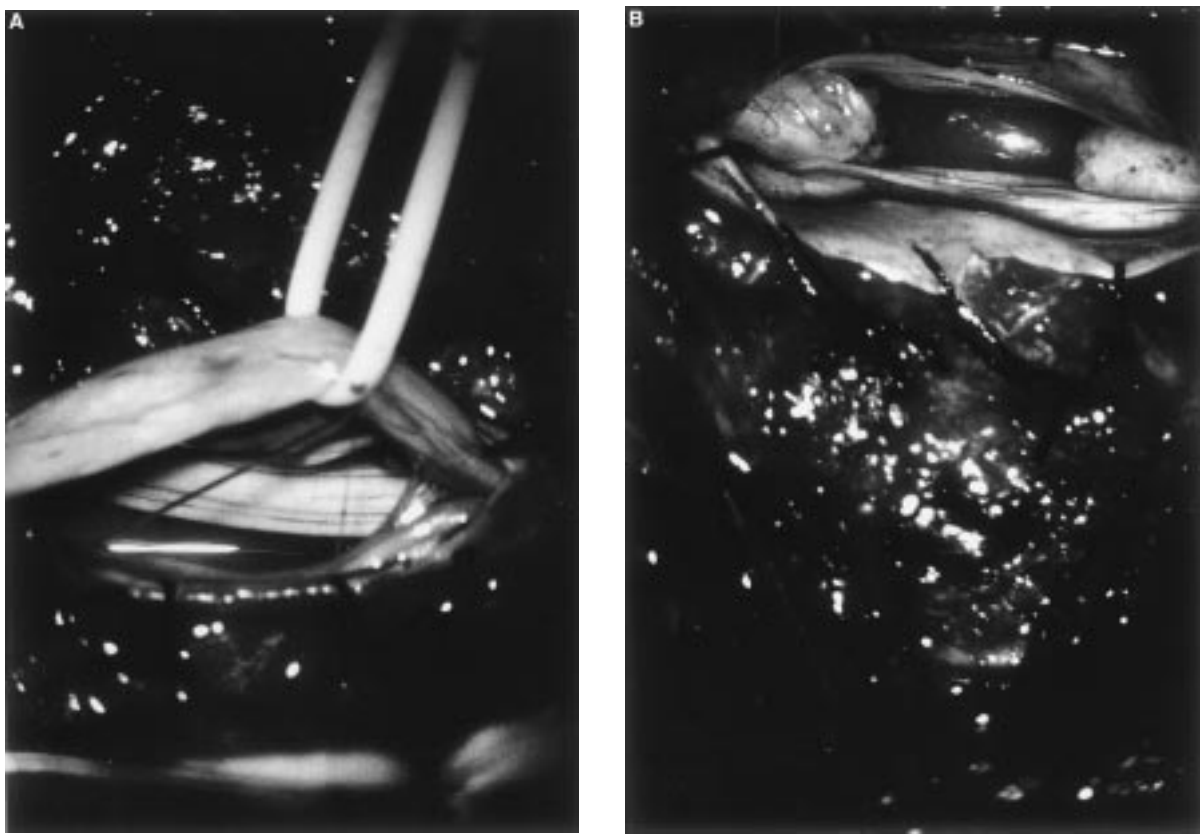

Figure 2. A: Intraoperative photograph of thickened filum terminale or lipoma of filum terminale prior to sectioning. Vascular loop is around the filum. B: After sectioning the two ends separate by over 2.5 cm.

scoliosis as a primary or secondary complaint, one third had arrest or improvement in their scoliosis such that spinal fusion was not needed [28].

Dermal sinus tracts, dermoids and epidermoids

A congenital dermal sinus tract is an epithelial (cutaneous ectoderm) lined tract that forms a potential avenue of communication between the skin surface and the deeper tissues including the spinal cord [40,41]. They occur along the midline and may terminate at the fascia, along the posterior elements of the spine, in the epidural space, at the dura or extend intradurally to the dorsal surface of the spinal cord or even into the spinal cord itself. An inclusion tumor or dermoid may develop anywhere along this tract and is found in 60 percent of dermal sinuses [42,43]. Because of the potential communication with the skin surface these lesions may present with meningitis or intraspinal abscess, or with spinal cord or cauda equina compression secondary to the development of an inclusion tumor [44–46]. They are thought to develop if there is defective separation of the epithelial ectoderm from the neural ectoderm. As the conus ascends dramatically during embryological development, if the tract terminates within the neural tube, there may be a significant distance from the terminal portion of the tract and the skin opening.

Sinus tracts in the cervical area occur in about 1 percent, in the thoracic area in about 10 percent, in the lumbar area in about 41 percent, in the lumbosacral area in 12 percent, at the sacrum in 23 percent and at the sacrococcygeal junction in 13 percent of patients. As many as 27 percent extend intradurally and are attached to either the filum terminale, nerve roots or spinal cord. Inclusion tumors from these lesions may be either dermoids or epidermoids depending on the elements found [47].

Dimples or sinus tracts may occur with many forms of spinal dysraphism and are not pathognomonic. Simple dimples in the low sacral area, below the top of the intergluteal crease, can be seen in 2–4 percent of newboms and do not always imply spinal dysraphism [22,48]. In most cases the sacrococcygeal sinus attaches to the coccyx without intradural extension. Those located higher along the spinal axis, above the top of the gluteal crease, are of greater clinical importance. There is a much higher association with these lesions of intraspinal abnormalities.

The ostium of these tracts may be quite small and easily overlooked on physical examination. It is only when the child has had meningitis or has developed a deficit that the significance of the dimple is appreciated. The most common organisms found when meningitis develops are *Staphylococcus aureus* and *Escherichia coli*, followed by *Proteus* and anaerobic organisms. Recurrent meningitis should alert the physician to the possibility of such a dermal sinus tract.

Neurological examination in such children is usually normal initially. This may change as an inclusion tumor such as a dermoid or epidermoid develops. Rapid neurological deterioration may suggest that an infection of an inclusion tumor has occurred or the development of an abscess. In some patients, the attachment of the dermal sinus tract to the underlying neural elements, creates tract on the spinal cord. In these circumstances, the presentation is as a result of a tethered cord syndrome.

Evaluation of spinal dermal sinuses can be with a spinal ultrasound in the younger child and with MRI imaging in the older child or adult [3]. Ultrasonography readily demonstrates the subcutaneous tract, intraspinal inclusion tumors and may even show diminished spinal cord pulsations. Extension of the dermal sinus tract through the dura can sometimes be difficult to assess even with very good MRI imaging [49,50]. It must therefore be appreciated that there may be a significant intradural extension of a dermal sinus tract without significant MRI findings. Scanning of the intracranial contents is not indicated. Rarely do patients with dermal sinus tracts have associated intracranial anomalies or hydrocephalus.

Removal of the dermal sinus tract, particularly those located above the sacrococcygeal region is indicated even in asymptomatic patients. The goal of the operation is total removal of the lesion and the surgeon must be prepared to perform a laminectomy and intradural exploration of the sinus tract. The mouth of the sinus tract is excised as an ellipse and the fibrous tract is kept intact. This tract is then followed down to its termination. If the tract is seen to penetrate the dura, the dura should be explored and if necessary a laminectomy performed to allow adequate intradural exposure. An aggressive attempt must be made to remove the entire tract to prevent the delayed development of a dermoid or epidermoid. If such an inclusion tumor is encountered it should be excised in its entirety, especially the capsule of the tumor. Such tumors often have a dense glial scar associated with the capsule and microscope assisted dissection of the spinal cord is often necessary. Numerous examples can be found in which inadequate removal of the epidermal elements led to future development of an inclusion tumor (Figure 3).

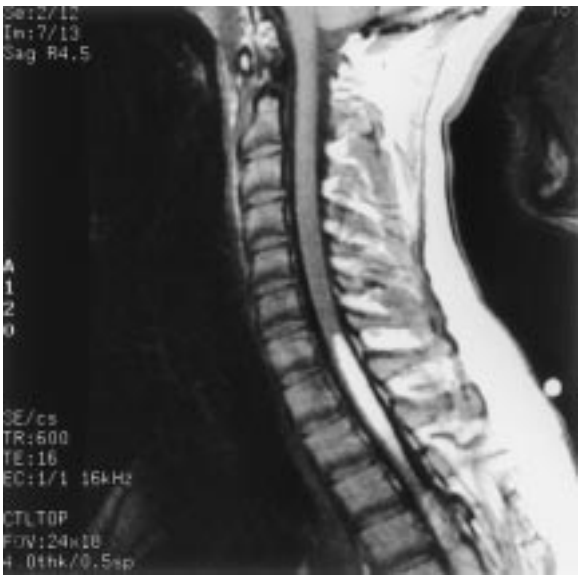

Figure 3. Sagittal, unenhanced T1 weighted MRI image of an intramedullary dermoid in 18 year old man. As a child he had a dermal sinus tract resected by a general surgeon, who had followed the tract only to the dorsal fascia.

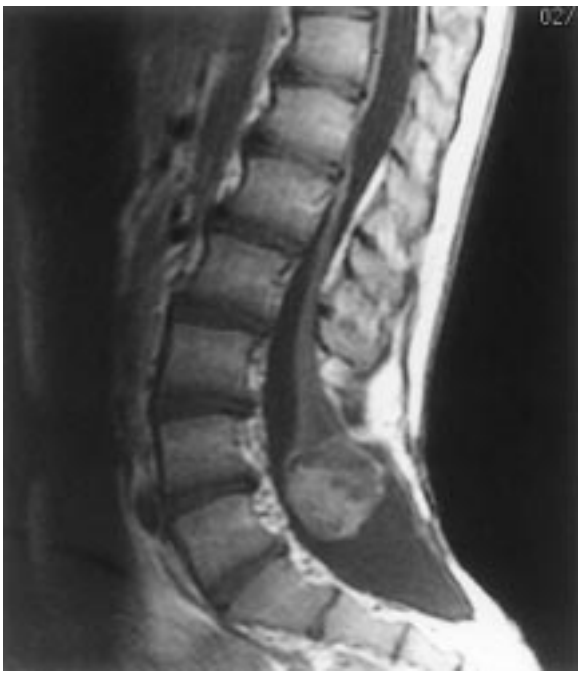

Figure 4. Sagittal, enhanced Tl weighted MRI image of an inclusion dermoid in 16 year old with previously repaired myelomeningocele.

Another example of inclusion tumors can be seen in myelomeningocele patients. If dermal elements are included in the initial repair, inclusion tumors may develop later in life (Figure 4). These tumors can result in late deterioration in myelomeningocele patients. There management is identical to other dermoids or epidermoids. Complete excision of the tumor with capsule should be attempted to prevent recurrence.

The outcome in patients with dermal sinus tracts and inclusion tumors is generally excellent. Postoperative complications can include neurological deficits and CSF leak. The contents of the cyst must not be spilled into the CSF spaces as they can result in a significant inflammatory response.

Lipomyelomeningocele

Subcutaneous lipomas located at the lumbosacral area may extend through an area of spina bifida through the dura and attach to the conus medullaris, the cauda equina or the filum terminale. Lipomas are thought to arise when the cutaneous ectoderm prematurely separates from the neuroepithelium prior to neural fold fusion. The mesenchyme can then insinuate itself between the neural tube and the overlying cutaneous ectoderm. The term lipomyelomeningocele is actually a misnomer, implying the herniation of neural elements into the subcutaneous mass. The fatty tissue generally insinuates itself within the spinal cord or filum.

Terminal or filum terminale lipomas are thought to result from disorders of secondary neurulation (Figure 5A,B). The presentation of these patients is that of a patient with a tethered spinal cord, thus patients may have scoliosis, foot deformities, weakness, sensory deficits and bladder/bowel difficulty. There is an increased association of lipomyelomeningocele in children with imperforate anus. Associated skin anomalies are common and include hairy patches/hypertrichosis, hemangiomas, dimples, sinus tracts, skin tags or even caudal appendages. Unilateral or bilateral cavovarus deformity of the foot, leg length discrepancy and claw toes may be seen. Scoliosis, especially if in a male, with a left-side curve and with rapid progression or accompanying pain should alert the physician to the possibility of a tethered cord. New onset fecal or urinary incontinence or abnormal voiding pattern may signal a tethered cord.

Such children are largely intact at birth and develop symptoms later in life [51,52]. At Children's Memorial Hospital in Chicago, 42 percent of 197 patients

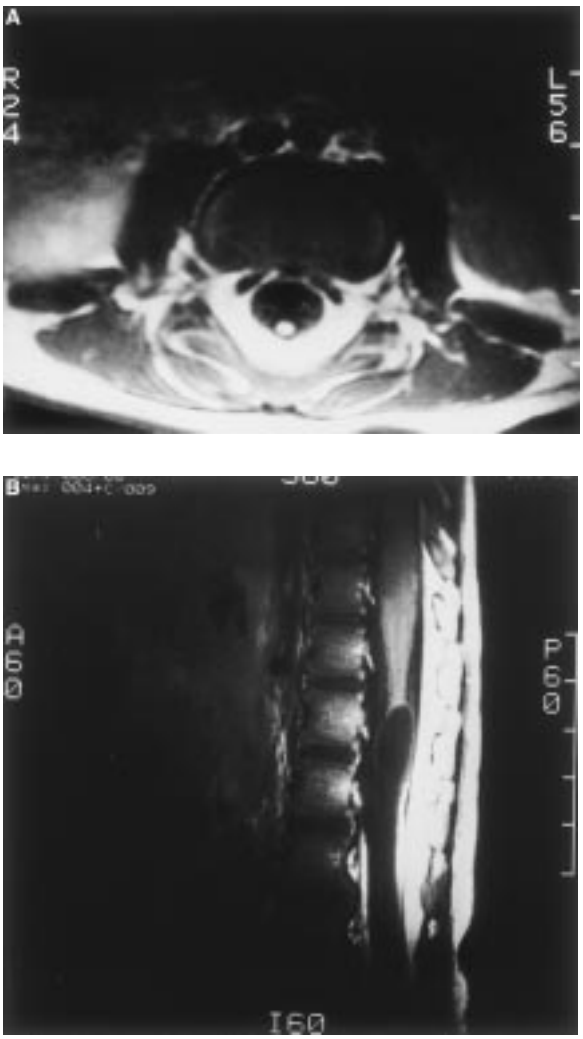

Figure 5. A: Axial, unenhanced T1 weighted MRI image of filum terminale lipoma or thickened filum in 6 year old with recurrent urinary tract infections. B: Sagittal unenhanced T1 weighted MRI images of child with tethered cord, thickened filum and cyst within the distal conus and filum.

with lipomyelomeningocele, had neurogenic bladders at presentation. Often such bladder dysfunction is apparent only on urodynamic testing, particularly in young infants [53,54]. Older patients often presented with pain, scoliosis, gait abnormalities and progressive bladder dysfunction. Approximately 75 percent of children less than 6 months of age, who are diagnosed with lipomyelomeningocele are intact, where as only a very small number are intact when diagnosed greater than 4 years of age. The natural history appears to be one of decline and worsening function, thus it is recommended that repair and release of the spinal cord occur before neurological deficits occur [55].

Ninety percent of lipomas are located in the lumbosacral area and the subcutaneous portions are evident since birth. Most patients are asymptomatic at birth but develop deficits gradually [56]. The development of symptoms appears to be related to traction on the spinal cord. The exact number of patients without symptoms who do not require surgical intervention is not known but is believed to be less than 20 percent [57,58]. Symptoms can occur throughout life and even adult patients previously asymptomatic can present with symptoms [36]. Such symptoms include pain, neurological dysfunction, urologic dysfunction and delayed orthopedic problems. Rarely symptoms can be precipitated by rapid weight gain or minor trauma.

Approximately 32 percent of patients with occult spinal dysraphism have associated lipomas and about one in 4000 births will have a lipomyelomeningocele [18]. The subcutaneous portions of these lipomas are composed of normal fat and do not have distinctive capsule. The intradural extension of these lipomas to the neural elements, acts to tether these neural elements to the subcutaneous tissue and dura [52]. The neural elements remain within the canal in the case of simple lipomas. In the case of lipomyelomeningocele, neural elements can extend through the deficient dura into the subcutaneous tissues. The site of attachment of the lipoma to neural elements consists of an intermingling of fat tissue, connective tissue and occasionally associated islands of bone, muscle and cartilage [59].

MRI has markedly improved our understanding of these lesions [50]. It has become the primary diagnostic tool in evaluating the extent of the lipoma and any associated abnormalities such as a dermoid or diastematomyelia [60,61]. CT scanning can be helpful is assessing complex bony abnormalities [3,62].

The goal of surgical intervention in such lipomas is untethering of the spinal cord [63,64]. The margins of the subcutaneous can be indistinct from normal subcutaneous fat but a plane can usually be developed to identify the lumbodorsal fascia. There is usually a defect at the level of the lumbodorsal fascia through which the lipoma penetrates into the spinal canal [65]. Laminectomy is done to expose normal dural sac and the dissection carried out caudally to expose the incompetent dura and lipoma. The dura is opened proximally to identify the normal neural elements and examine the extension of the lipoma intradurally. There may be anywhere from a small stalk of lipoma adherent to the neural elements to extensive involvement of the neural

elements and the lipoma. The lateral dura may also be deficient and the fat may fuse to it. Careful dissection of the dural fat plane is necessary on either side. Care must be taken in assessing whether any neural elements are incorporated into the lipoma. Freeing up of the distal lipoma can be accomplished if no such elements are present. The lipoma can be diminished in size with the help of a CO_2 laser [35,66]. The lipoma should not be removed in its entirety, particularly if it is attached to the spinal cord. The interface with the spinal cord can be quite fibrous and significant daniage to the spinal cord can occur [57,58,63,67].

Lipomas can be classified as dorsal, caudal, intermediate or filar base on their anatomic relationship with the neural elements [67,68]. The dorsal type fuses to the conus posteriorly and thus can be trimmed flush with the spinal cord. The caudal type fuses with the terminal conus and should be transected distal to functional sacral roots. The filar type expands the filum terminale, often up to the conus and can be transected distal to the conus medullaris. The most difficult is the intermediate or combined form is a blend of the dorsal and caudal types of lipoma (Figure 6A,B). There are often functional nerve roots coursing through the lipoma and dissection occurs at the terminal dural sac where the lipoma and sac fuse.

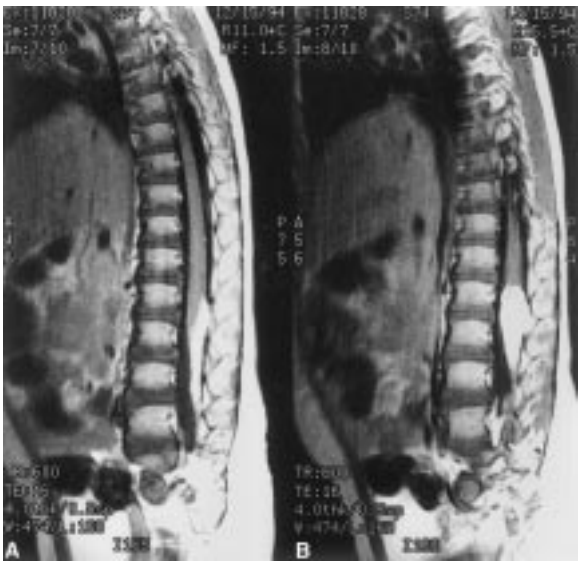

Figure 6A and B. Sagittal, unenhanced T1 weighted MRI images of transitional lipoma with nerve roots coursing through the lipoma.

Water tight reconstruction of the dura should be performed to avoid CSF leak. Use of graft such as lumbodorsal fascia, cadaveric dura or bovine pericardium is often necessary. It is important that the conus medullaris is free of the dura and not constricted by the dural closure. Lipomas of the spinal cord can be challenging procedures even in the most experienced of hands. Incomplete removal of the lipoma is appropriate and the mass should be debulked so as to prevent compression and to allow closure of the dural sac. Retethering is not uncommon. McLone reports up to a 34 percent incidence of retethering in infants who have undergone untethering. As in the original situation, it is the tethering of the spinal cord to the surrounding dura which appears to be the problem [35,52].

Of children who were normal at the time of operation, 95 percent will retain normal bladder function in long term follow up after untethering. A small number of children will experience late deterioration and require re-operation and re-release of their tethered cord [69]. Fully 50 percent of children get some improvement in motor or sensory function after surgery [66]. Improvement in bladder function occurs in less than 20 percent of patients [32]. Pain also seemed improved in most patients postoperatively.

Neurenteric cyst

The neurenteric cyst is an intramedullary or intradural extramedullary cyst which can cause compression of the spinal cord. Embryologically the cyst is derived from endodermal tissue which is displaced dorsally into the spinal canal. This endoderm fused to the developing notochord during the third week of gestation. There are often abnormalities of the vertebral body and trans vertebral communication between the intraspinal neurenteric cyst and posterior mediastinal cyst. These are extremely rare lesions and histologically consist of simple or pseudostratified columnar epithelium with or without a muscle layer. Coexisting mediastinal cysts must be considered. In most cases there is direct communication through the vertebral body to the anterior cyst [70–73].

Presentation of neurenteric cysts is usually secondary to compression of the spinal cord. A few patients may present with meningitis. Removal of the entire cyst wall is necessary to prevent reaccumulation of the cyst. The cyst wall can be quite adherent to the spinal cord and careful dissection is necessary. Often these lesions may first present as a chest mass with

extension into the spinal canal. A combined approach by the general surgeon and the neurosurgeon may be necessary. In the adult the presentation may mimic other mass lesions and include pain, weakness and dysesthesias.

Evaluation of these cysts can require a number of modalities. MRI scanning demonstrates the spinal component of these lesions as well as their extension anteriorly (Figure 7) [74]. Additional information about the vertebral body abnormalities can be obtained from CT scan. Myelography can be helpful in distinguishing these cysts from anterior sacral meningoceles or dural ectasia.

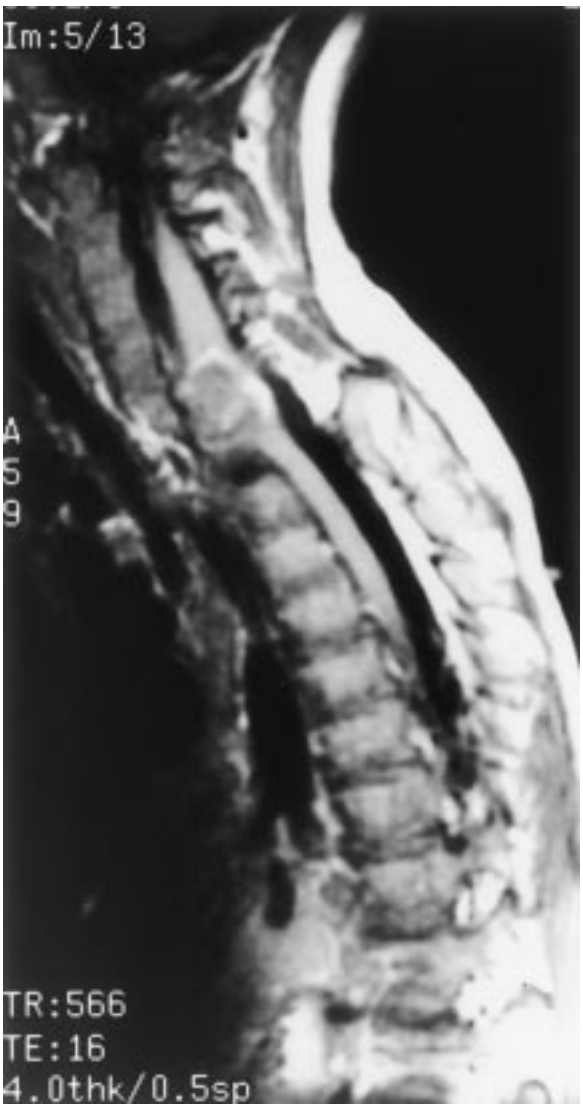

Figure 7. Sagittal, enhanced T1 weighted MRI image of child with cervical neurenteric cyst and multiple vertebral body abnormalities.

Compressive symptoms can be relieved by removal of the cyst in its entirety [75]. Consideration for a combined approach can be important if there is extension anteriorly into the mediastinum or abdomen. In such situations, a staged procedure is often necessary. In most instances, the cyst can be approached via a laminectomy. Section of the dentate ligaments can afford further access to the ventral aspects of the cyst. There is often a thickened, fibrous capsule associated with the cyst and this can require considerable effort to free from the spinal cord. Microsurgical dissection of the entire cyst capsule from the spinal cord is necessary to assure that recurrence does not occur [72,76].

The outcome from decompression is usually excellent. Recurrences have been shown to be caused by incomplete removal of the cyst capsule. Symptoms are improved in the majority of patients [72].

References

1. French B: Midline fusion defects and defects of formation. In: JR Youmans (ed), Neurological Surgery. 3rd edn., WB Saunders, Philadelphia, 1990, pp 1081–1235
2. Holmes L: Congenital malformations. N Engl J Med 295: 204–207, 1976
3. James H, Oliff M, Mulcahy J: Spinal dysraphism: a comprehensive diagnostic approach. Neurosurgery 2: 15–21, 1978
4. Lemire: Variations in development of the cauda neural tube in human embryos (Horizons XIV–XXI). Teratology 2: 361–369, 1969
5. Passarge F, Lenz W: Syndrome of caudal regression in infants of diabetic mothers. Pediatrics 37: 672–675, 1966
6. O'Rahilly R, Gardner F: The timing and sequence of events in the development of the human nervous system during the embryonic period proper. Z Anat Entwicklungsgesch 134: 1–12, 1971
7. Osaka K, Tanimura T, Hirayama A et al.: Myelomeningocele before birth. J Neurosurg 49: 711–724, 1978
8. Patten B: Embryological stages in the establishing of myeloschisis with spina bifida. Am J Anat 93: 365–395, 1953
9. Patten B: Patten's Human Embryology. Elements of Clinical Development. McGraw-Hill Book Co, New York, 1976
10. Kunitoma: The development and reduction of the tail and the caudal end of the spinal cord. Contrib Embryol 8: 161–198, 1918
11. Al-Mefty O, Kandzari S, Fox J: Neurogenic bladder and the tethered spinal cord syndrome. J Urol 122: 112–115, 1979
12. Yamada S, Zinke D, Sanders D: Pathophysiology of 'tethered cord syndrome'. J Neurosur 54: 494–503, 1981

13. Reimann A, Anson B: Vertebral level of termination of the spinal cord with report of a case of sacral cord. Anat Rec 88, 1944
14. Nazar G, Casale A, Roberts J et al.: Occult filum terminale syndrome. Pediatr Neurosurg 23: 228–235, 1995
15. Oakes W: The borderlands of the primary tethered cord syndrome. Clinical Neurosurg 43: 188–202, 1996
16. Warder D, Oakes W: Tethered cord syndrome: the low-lying and normally positioned conus. Neurosurgery 34: 597–600, 1994
17. Reigel D: Tethered spinal cord. Concepts Pediatr Neurosurg 4: 142–164, 1983
18. Kang J, Kim M, Kim D et al.: Effects of tethering on regional spinal cord blood flow and sensory-evoked potentials in growing cats. Child's Nerv Syst 3: 35–39, 1987
19. Purtzer T, Yamada S, Tani S: Metabolic and histologic studies of chronic model of tethered cord. Surg Forum 36: 512–514, 1985
20. Tani S, Yamada, Knighton R: Extensibility of the lumbar and sacral cord. Pathophysiology of the tethered spinal cord in cats. J Neurosurg 66: 116–123, 1987
21. Yamada S, Schreider, Ashwal S et al.: Pathophysiologic mechanisms in the tethered spinal cord syndrome. In: Holtzman RNNSB (ed), The Tethered Spinal Cord. Thieme Stratton, New York, 1985, pp 29–40
22. Albright A, Cartner J, Wiener F: Lumbar cutaneous hemangiomas as indicators of tethered spinal cords. Pediatrics 83: 977–980, 1989
23. Levitt MAPM, Rodriguez G, Gaylin DS, Pena A: The tethered spinal cord in patients with anorectal malformations. J Pediatr Surg 32: 462–468, 1997
24. Reigel D, McLone D: Tethered spinal cord. In: Cheek WRMA, McLone DG, Reigel DH, Walker ML (eds), Pediatric Neurosurgery – Surgery of the Developing Nervous System. WB Saunders, Philadelphia, 1994, pp 77–95
25. Warf B, Scot TR, Barnes P et al.: Tethered spinal cord in patients with anorectal and urogenital malformations. Pediatr Neurosurg 19: 25–30, 1993
26. Lapras CLPJ, Huppert J, Bret P, Mottolese C: The tethered cord syndrome (expenence of 58 cases). J Pediatr Neurosci 1: 39–50, 1985
27. Hendrick E, Hoffman H, Humphreys R: The tethered spinal cord. Clin Neurosurg 30: 457–463, 1983
28. Hoffman H, Hendrick F, Humphreys R: The tethered spinal cord: its protean manifestations, diagnosis and surgical correction. Child's Brain 2: 145–155, 1976
29. Pang D, Wilberger J: Tethered cord syndrome in adults. J Neurosurg 57: 32–47, 1982
30. Fuki J, Kakizaki T: Urodynamic evaluation of tethered cord syndrome including tight filum terminale. Urology 16: 539–552, 1980
31. Kaplan W: Management of the urinary tract in myelomeningocele. Probl Urology 2: 121–131, 1988
32. Karlin I: Incidence of spinal bifida occulta in children with and without enuresis. Am J Dis Child 49: 125–134, 1935
33. Sharrard W: Paralytic pes cavus and claw toes. In: RL M (ed), Myelomeningocele. Grune & Stratton, New York, 1977, pp 469–474
34. Mc Cullough D, Levy L, DiChiro G et al.: Toward the prediction of neurological injury from tethered spinal cord: investigation of cord motion with magnetic resonance. Pediatr Neurosurg 16: 3–7, 1990–1991
35. McLone D, Naidich T: Laser resection of fifty spinal lipomas. Neurosurgery 18: 611–615, 1986
36. Koyanagi I, Iwasaki Y, Hida K et al.: Surgical treatment supposed natural history of the tethered cord with occult spinal dysraphism. Child's Nerv Syst 13: 268–274, 1997
37. Lunardi PMP, Ferrante L: Long-term results of surgical treatment of spinal lipomas: report of 18 cases. Acata Neurochir (Wien) 104: 64–68, 1990
38. Yoneyama T, Fuki J, Ohtsuka K et al.: Urinary tract dysfunctions in tethered spinal cord syndrome: improvement after surgical untethering. J Urol 133: 999–1001, 1985
39. Zoller G, Schoner W, Ringert R: Pre and postoperative urodynamic findings in children with tethered spinal cord syndrome. Eur Urol 19: 139–141, 1991
40. Cheek W, Laurent JP: Dermal sinus tracts. Concepts Pediatr Neurosurg 6: 63–75, 1985
41. Smith G, Altman D: Occipital dermal sinus. Am J Dis Child 98: 713–719, 1959
42. Altman R: Dermoid tumor of the posterior fossa associated with congenital dermal sinus. J Pediatr 62: 565–570, 1963
43. Bailey I: Dermoid tumors of the spinal cord. J Neurosurg 33: 676–681, 1970
44. Perloff M: Congenital dermal sinus complication by meningitis. Report of a case. J Pediatr 44: 73–76, 1954
45. Petterson G, Werkmaster K: Intraspinal dermoid cysts in children. Acta Paediatr (Stockh) 52: 187–189, 1962
46. Walker A, Bucy P: Congenital dermal sinuses: a source of spinal meningeal infection and subdural abscesses. Brain 57: 401–421, 1934
47. Baldi P, Paini G, Bertolino G et al.: Diastematomyelia in adults. Surg Neurol 25: 501–504, 1986
48. Burrows F: Some aspects of occult spinal dysraphism: a study of 90 cases. Br J Radiol 41: 496–607, 1968
49. Barkovich A, Edwards M, Cogen P: MR evaluation of spinal dermal sinus tracts in children. AJNR 12: 123–129, 1991
50. Kuharik M, Edward M, Grossman C: Magnetic resonance evaluation of pediatric spinal dysraphism. Pediatr Neurosci 12: 213–218, 1985–1986
51. Bruce D, Schut L: Spinal lipomas in infancy and childhood. Child's Brain 5: 192–203, 1979
52. McLone D, Mutluer S, Naidich T: Lipomyelomeningoceles of the conus medullaris. Concepts Pediatr Neurosurg 3: 170–177, 1983
53. Atala A, Bauer S, Dyro F et al.: Bladder functional changes resulting from lipomyelomeningocele repair. J Urol 148: 592–594, 1992
54. Foster L, Kogan B, Cogen P et al.: Bladder function in patients with lipomyelomeningocele. J Urol 143: 984–986, 1990
55. Kanev P, Berger M: Lipomyelomeningocele and myelocystocele. In: JR Youmans (ed), Neurological Surgery. WB Saunders, Philadelphia, 1996, pp 861–872
56. Kanev P, Lemire R, Loeser J et al.: Management and long term follow-up review of children with lipomyelomeningocele. J Neurosurg 73: 48–52, 1990

57. Hoffman H, Taecholarn C, Hendrick E et al.: Management of lipomyelomeningoceles: experience at the hospital for sick children, Toronto. J Neurosurg 62: 1–8, 1985
58. Hoffman H, Taecholarn C, Hendrick E et al.: Lipomyelomeningoceles and their treatment. Concepts Pediatr Neurosurg 5: 107–117, 1985
59. Villarejo F, Blazquez M, Gutierrez-Diaz J: Intraspinal lipomas in children. Child's Brain 2: 361–370, 1976
60. Komiyama M, Hakuba A, Inoue Y et al.: Magnetic resonance imaging: lumbosacral lipoma. Surg Neurol 28: 259–264, 1987
61. Brunberg J, Latchaw R, Kanal F et al.: Magnetic resonance imaging of spinal dysraphism. Radiol Clin North Am 26: 181–205, 1988
62. Sato K, Shimoji, Sumie H et al.: Surgically confirmed myelographic classification of congenital intraspinal lipoma in the lumbosacral area. Child's Nerv Syst 1: 3–11, 1985
63. Chapman P, Davis K: Surgical treatment of spinal lipomas in childhood. Concepts Pediatr Neurosurg 3: 178–190, 1983
64. Schut L, Bruce D, Sutton L: The management of the child with a lipomyelomeningocele 30: 464–476, 1983
65. Quencer R, Montalvo B, Naidich T et al.: Intraoperative sonography in spinal dysraphism and syringohydromyelia. Am J Neurorad 8: 329–337, 1987
66. McLone D, Hayashida S, Caldarelli M: Surgical resection of lipomyelomeningoceles in 18 asymptomatic infants. J Pediatr Neurosci 1: 239–244, 1985
67. Chapman P, Beyerl B: The Tethered Spinal Cord with Particular Reference to Spinal Lipoma and Diastematomyelia. Blackwell Scientific Publishers, Boston, 1986
68. Chapman P: Congenital intraspinal lipomas: anatomic considerations and surgical treatment. Child's Brain 9: 37–47, 1982
69. Sakamoto H, Hakuba A, Fujitani K et al.: Surgical treatment of the rethethered spinal cord after repair of lipomyelomeningocele. J Neurosurg 74: 709–714, 1991
70. Agnoli A, Laun A, Schonmayr R: Enterogenous intraspinal cysts. J Neurosurg 61: 834–840, 1984
71. Ahmed S, Jolleys A, Dark J: Thoracic enteric cysts and diverticulae. Br J Surg 59: 963–968, 1972
72. Menezes A, Ryken T: Craniocervical intradural neurenteric cysts. Pediatr Neurosurg 22: 88–95, 1995
73. Scoville W, Manlapaz J, Otis R et al.: Intraspinal enterogenous cyst. J Neurosurg 20: 704–706, 1963
74. Brooks B, Duvall F, el Gammal T et al.: Neuroimaging features of neurenteric cysts: analysis of nine cases and review of the literature. AJNR 14: 735–746, 1993
75. Piramoon A, Abbassioun K: Mediastinal enterogenic cyst with spinal cord compression. J Pediatr Surg 9: 543–545, 1974
76. Alrabeeah A, Gillis D, Giacomantonio M et al.: Neurenteric cysts: a spectrum. J Pediatr Surg 23: 752–754, 1988

Address for offprints: Karin M. Muraszko, Department of Pediatric Neurosurgery, University of Michigan, 2128 Taubman Health Center, 1500 East Medical Center Drive, Ann Arbor, MT 48109-0339, USA; Tel.: 734-936-5062; Fax: 734-615-3772; E-mail: karinm@umich.edu