Split cord malformation associated with scoliosis in adults

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ABSTRACT

Split cord malformations (SCMs) are rare congenital abnormalities arising from a primary neurulation defect during embryological development of the spinal cord. SCMs can lead to progressively worsening scoliosis and gait difficulties if left untreated. We report on two patients who atypically presented with SCM in adulthood and review previous reports.

KEYWORDS Adult scoliosis; spinal cord dysraphism; split cord malformation; tethered cord syndrome

Split cord malformations (SCMs) are rare congenital abnormalities arising from a primary neurulation defect during embryological development of the spinal cord. SCMs can lead to progressively worsening scoliosis, torticollis, motor deficits, and gait difficulties if left untreated.^{1,2} Traditionally, SCMs have been classified into two categories based upon the presence or absence of an individual dural sac around each hemicord and the presence or absence of a rigid septum separating the two hemicords. Although SCMs are rare in incidence within the general population, it is increasingly unlikely for clinical symptoms to present in adulthood.³ The following two cases illustrate how adult patients with an SCM may present.

CASE 1

A 58-year-old English man with severe scoliosis since childhood presented with worsening back pain, left leg pain, and difficulty with ambulation. The patient's pain had continued despite medication trials and epidural steroid injections. In the months after his first clinic evaluation, his pain worsened, causing him to have a severe antalgic gait, which forced him to rely upon a wheelchair. Examination revealed occult spina bifida, sensory deficits in his lower extremities, and near full strength in both legs.

The standing radiograph revealed multilevel thoracolumbar scoliosis. A rightward curvature with apex at T9–T10 was seen, as well as a leftward curvature with apex at L3–L4. Magnetic resonance imaging (MRI) revealed an SCM

beginning at the apex of the dextroconvex curve at the T9–T10 levels and extending to the S2 level.

Computed tomography (CT) of the thoracic and lumbar spine revealed severe dextroconvex and dextroconvex scoliosis centered at T9 with compensatory levoconvex curvature at the cervicothoracic junction and L4. More specifically, partial ankylosis at T12–L1, moderate bilateral facet hypertrophy with disc bulge at L2–L3, severe right neural foraminal narrowing at L4–L5 with L4 pars defect, and a clearly discernible cutaneous defect seen in the midline at the level of the L2–L3 disc were observed. CT myelogram confirmed an SCM with the left hemicord larger than the right with an associated plaque-like ossification along the posterior dural wall. Above this ossification, the spinal cord was unitary. Below, there were two hemicords, left larger than right, in a single thecal sac extending to S2.

Our patient underwent T9–T11 laminectomies with resection of the intradural calcified lesion and release of adhesions tethering the spinal cord. The spinal contents were contained within one dural sac. Microsurgical techniques were used to resect the calcified plaque from extensive arachnoid adhesions from the dura to the spinal cord (*Figure 1*). The scar tissue remaining was explored and revealed an SCM in the distal part of the exposed area.

At the 6-week follow-up examination, the patient had a normal neurologic examination with no complaints other than a lingering tugging sensation in the lumbar region, which had improved significantly since the immediate post-surgical period. At his 6-month visit, the patient could walk

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Figure 1. Case 1. Removal of calcified plaque.

about 100 yards before needing a walker or resting with his cane. At his 21-month visit, the patient could ambulate without assistive devices.

CASE 2

A 28-year-old Hispanic woman was found to have asymptomatic scoliosis at 12 years of age. She presented with progressively worsening upper back pain beginning at 15 years of age. At 20 years, she gave birth to her last child and had since experienced worsening of her back pain, particularly exacerbated by bending at the waist. At the age of 25, her pain became associated with numbness in her upper extremities, right greater than left, when turning her head. Examination revealed decreased range of motion of her upper extremities and a right thoracic kyphosis.

Standing radiographs revealed rightward curvature of the thoracic spine from the superior endplate of T6 to the inferior endplate of T12, apex at T8, and no compensatory leftward curve. CT showed a fused C5–C6, left T3 and T7 hemivertebra, right T5 hemivertebra, spina bifida occulta at C5 and C6, varying degrees of fusion of C5–T6, and a small osseous septum that extended sagittally through the central canal at T2. MRI revealed an SCM at the level of T1–T4 in the form of a thoracic cord split extending from C7 to T4 (*Figure 2a*). Above C7 and below T4, the spinal cord remained unitary.

The patient underwent C5–C7 laminectomies with resection of the intradural calcified lesion from the arachnoid adhesions. The spinal contents were contained within one dural sac. The calcified lesion was demonstrated by showing instrumentation of the void above and below the bony septum (*Figure 2b*).

At 6-week follow-up, the patient had a normal neurologic exam and some right posterior thoracic pain, along with pain in her right wrist and the proximal aspects of her right digits. The back pain she experienced prior to surgery was gone. At the 6-month follow-up visit, she still had lingering pain in the middle of her upper back, but the right upper extremity pain had resolved. At 14-month follow-up, she felt happy with her progress and was standing more erect.

DISCUSSION

The most widely agreed-upon theory regarding the pathogenesis of SCM was proposed by Pang et al, who

Figure 2. Case 2. (a) Axial T2 MRI demonstrating two hemicords in a single thecal sac. (b) Calcified lesion demonstrated by showing instrumentation of the void above and below the bony septum.

described an accessory neurenteric canal that begins as an ecto-endodermal adhesion when the two primary germ layers are in contact in the midline, forming a fistula. This leads to the formation of two hemineural plates and, with subsequent neurulation, two heminotochords.⁴ Normally, the integration of the two paramedian notochords along the midline leads to the embryological development of the spinal cord.² SCMs can manifest if this process is abnormal in the setting of a longitudinal division in the form of a fistula.^{4–6} Therefore, the formation of the fistula itself is the initial pathway that is mutual to all of the SCMs. Incidence of an SCM was found to be bimodal, where most patients presented in infancy or their teens and later presentations happened in latter parts of the fourth decade of life.^{7–17}

There are two general types of SCMs. In type I SCM, the hemicords are enveloped within individual dural sacs and separated by a rigid (bony or cartilaginous) median spur. In type II SCM, the hemicords are encased within a single dural sac and the median structure is composed of nonrigid fibrous tissues.^{2,18} Our cases do not fit nicely into this classification system because both patients had hemicords separated by a bony spicule and were contained within a single dural sac. A peculiar detail of our cases was the orientation between the anatomic location of the bony spicule and the level of SCM hemicord split. Although the unified theory by Pang et al has been effective at classifying the majority of SCMs, it did not fully categorize our patients.

A new classification system proposed by Mahapatra and Gupta further divides type I SCM into four categories: Ia, bony spur in the center with equally duplicated cord above and below the spur; type Ib, bony spur at the superior pole with no space above and a large duplicated cord below; Ic, bony spur at the lower pole with a large duplicated cord above; and Id, bony spur straddling the bifurcation with no space above or below the spur.¹ Given this classification system, our patients can be classified as type Ib and type Ia, respectively. Due to the rarity of adult type I SCM, recent case reports by Viswanathan et al describe older patients with SCM who presented with similar complaints of axial back and radicular pain, which were relieved with surgery.¹⁹ Although the authors did not subclassify the type I SCM based upon Mahapatra and Gupta's new classification

system, the primary endpoint of pain relief and improved quality of life was demonstrated, as it was in our patients. We therefore emphasize that the most common sign associated with SCM is skin stigmata, usually in the form of hypertrichosis, and clinicians should remain vigilant in seeking out these occult physical exam findings.^{1,9} Plain imaging, based upon a study from Mahapatra and Gupta, revealed scoliosis or kyphoscoliosis in 41% of patients, a finding that our patients also shared.¹ According to Jindal and Mahapatra, MRI is highly accurate in confirming this diagnosis.²⁰

The long-term prognosis for patients with adult presentations of SCM is not clearly defined; however, our patients continued to experience great relief of their preoperative symptoms at the 1-year mark, a finding that has been shared and commented upon by Proctor and Scott, as well as Mahapatra's review of 300 cases.^{21,22}

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