Case Report

Type I Split Spinal Cord Malformation: Literature review, Case presentation and Surgical Technique

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Abstract

Background: Split Spinal cord Malformation (SSCM) is a form of spinal dysraphism in which any or all of the spinal cord, cauda equina, and filum terminale are divided by a bony or fibrous spur. There are two described types of SSCM, namely Types I and II. In Type I SSCM, there are separate hemi-cords contained in two dural sacs, separated by a bony spur; in Type II SSCM, there is a single dural sac containing two hemi-cords separated by a fibrous band.

Purpose: We present here a case presentation of an adult patient with Type I SSCM with tethered spinal cord. Also presented is a literature review on the embryogenesis, anatomy and imaging findings of SSCM.

Conclusions: In untreated cases of diastematomyelia, neurological deficits are progressive in nature in both the adult and pediatric populations. Thus, many authors advocate early surgical intervention. Further studies are needed to evaluate the incidence of re-tethering after surgical treatment of SCCM.

ABBREVIATIONS


INTRODUCTION

Split Spinal cord Malformation (SSCM) can be defined as a form of spinal dysraphism in which any or all of the spinal cord, cauda equina, and filum terminale are divided into two lateral parts by a dorsal-ventral bony or fibrous spur [1]. There are two described types of SSCM, namely Types I and II. Presented here is a literature review on the embryogenesis, anatomy and imaging findings of SSCM as well as a case presentation of an adult patient with Type I SSCM with tethered spinal cord and our surgical approach.

Embryogenesis

The most widely accepted theory about the embryological mechanisms that produce a Split Spinal cord malformation (SSCM) is the one proposed by Bremer and updated by Pang [2] which is based on the existence and persistence of an accessory neuroenteric canal. According to this theory, in the first weeks of gestation there is a primitive neuroenteric canal (PNC), which temporarily connects the yolk sac, which is of endodermic origin, with the amnion, which is ectodermic in origin [1-4]. It is thought that following the closure of the PNC, a new neuroectodermic communication (the accessory neuroenteric canal-ANC) develops; according to Bremmer, the ANC explains the range of anatomic malformations that may occur, including split cord malformations [1].

The persistence of the anterior end may cause intestinal duplication, formation of a fibrous band that interferes with intestinal rotation and development of a neuruectodermic cyst, while persistence of the posterior end results in the formation of dermal sinuses, dermoid or epidermoid cysts or cutaneous abnormalities such as angiomas, umbilical lesions and hypertrichosis [5,6].

The persistence of the intermediate part of the ANC causes the notochord and the neural placode to divide [4]. Division of the notochord interferes with the formation of vertebral bodies, causing hemivertebrae, bilid or butterfly vertebrae, fusion of adjacent vertebral bodies, hypertrophic or hyperplastic vertebrae; while division of the neural placode into two hemiplaciodes leads to the formation of two hemicords, causing a split cord malformation.

If the menenchyme surrounding the hemicords (meninx primitiva), the ANC forms the median
part of the dural sac with its external aspect and a bony spur with its internal aspect; this is an SSCM type I. The lateral part of the dural sac forms, as normal, from the regional mesenchyme. However, if the mesenchyme of the ANC has not incorporated precursor meningeal cells, then it is not involved in the formation of the dural sac. The dural sac derives from the mesenchyme, which surrounds the neural tube; the ANC either disappears or is transformed into an intradural fibrous spur situated between the two hemicords, thus constituting a SSCM type II [2-4]. It has been postulated that the persistence of the ANC could occasionally interfere with the neurulation process, driving the formation of a meningocele or a myelomeningocele associated with SSCM. In other cases the tract could interfere with the neurulation process only unilaterally, thus causing a hemi-meningocele or a hemi-myelomeningocele [4].

**ANATOMY AND IMAGING FINDINGS**

Type I SSCM account for 40–50% of all SSCM; they consist of two hemicords separated by an extra-dural bony or partially bony spur, covered by a double dural sac, contained within a double spinal canal [2,3,7]. This spur can be strictly anterior-posterior and divide the canal and the cord into two symmetrical halves, or can be slanting in the axial plane, dividing the spinal canal asymmetrically with one normal and one abnormal, hypoplastic hemicord. The most common location of the spur is in the lumbar spine, with spurs occurring less frequently in the cervical and lowers thoracic regions; they are exceptionally rare in the superior thoracic area and in the sacrum [4]. The hemicords are separated at the level of the spur but usually join together below it. However, they may continue separately, ending in two independent fila terminalia. In these cases the presence of a composite SSCM with more than one spur should be suspected [8-10]. The hemicords frequently do not join immediately above the spur, persisting through several vertebral segments.

Type II SSCM accounts for 50–60% of all cases and comprises of one dural sac, one spinal canal, and two equal, symmetrical hemicords between which there may be an anterior-posterior, fibrous intradural band not extending over more than one or two vertebral segments in height [2,3,7].

**Clinical features**

Although the majority of SSCM cases are diagnosed in infancy, such malformations can occur at any age, from the prenatal period right up to adulthood. They are more frequent in women (with a female to male ratio of 1.6:1) and have a highly variable clinical presentation [1,11-14]. SSCM may be diagnosed prenatally by ultrasound during the evaluation of a normal or pathologic pregnancy. Its diagnosis in newborns and infants is prenatal only unilaterally, thus causing a hemi-meningocele or a hemi-myelomeningocele [4].

**CASE PRESENTATION**

A 58 year-old gentleman presented to clinic with a 2-year history of progressively worsening left lower extremity weakness, urinary retention, constipation, abnormal gait and erectile dysfunction. He also reported bilateral foot numbness attributed to diabetic neuropathy, but did not require an assistive device for ambulation. His medical history is significant for diabetes mellitus, pilonidal cyst, hypospadias and urethral stricture.

On physical examination, he was noted to have full range of motion with no pain with forward flexion or extension of his spine, and no radicular pain. The patient had 5/5 strength in his bilateral upper extremities and his right lower extremity; on his left lower extremity he had 4/5 strength on hip flexion and 4+/5 strength distally. He had bilateral foot numbness and a tuft of hair on his lower lumbo-sacral spine with no obvious sinus tract or skin abnormalities. He walked with a broad based gait, had a negative romberg sign and 1+ reflexes diffusely.

MRI of the lumbar spine showed spina bifida occulta at L3-5, a low lying conus ending at L3-4, tethered cord, diastematomyelia with a bony septum at L3-4 with syringomyelia above the level of the split, and levo-scoliosis. A 10cm renal cyst was found on Ultrasound and CT of the abdomen and pelvis; this was aspirated and found to be benign. The patient was referred for baseline urodynamic testing and bladder ultrasound pre-operatively. Figure 1.

**Surgical technique**

Using Fluoroscopic guidance, we made a linear incision centered on L3, and performed sub-periosteal dissection, exposing the laminae of L2, L3 and L4. At this point, we performed a posterior decompression using a matchstick cutting burr and drilled down troughs bilaterally on the lamina of L3 down to the inferior lamina of L4 and the inferior part of the spinous process of L4 and the superior spinous process and superior lamina of L2. We then drilled the spinous processes and laminae from a lateral to medial fashion over the ligamentum flavum and dura.

The bony septum of the split cord malformation was continuous with the L3 lamina (Figure 2a). We took great care not to pull on the lamina or disrupt the spinal cord where it was tethered and split. We therefore drilled down the lamina in a lateral to medial fashion until we were left with a small island of bone in the middle of the thecal sac (Figure 2b). This island of bone met in the midline and went intradurally, splitting the spinal cord into two halves. We then brought in the microscope for visualization, illumination and magnification.

We opened the dura immediately inferior to the bony septum, and then around the septum in an elliptical fashion; this was then

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Figure 1 Saggital and axial T2- weighted MRI of the Lumbar spine showing syringomyelia, low-lying conus at L3-4, tethered cord at L4-5, diastematomyelia with a bony septum at L3-4 with syringomyelia above the level of the split.

Figure 2

continued in the midline, superior to the bony septum (Figure 2c). Next, we released the arachnoid adhesions and tacked up the dura with 4-0 neurolon sutures. We then identified the plane between the bony septum and both sides of the spinal cord, minimizing any manipulation of the spinal cord. Using a 3-mm diamond burr, we drilled down the bony septum until it was in smooth continuity with the posterior longitudinal ligament and the anterior aspect of the dura (Figure 2d). The nerve roots that were closely associated with the filum terminale were carefully separated. The filum was then identified and was cauterized in two locations caudally. It was then transected in two locations, with good un-tethering of the spinal cord. Results of intra-operative Motor Evoked Potentials (MEP) and Somato-Sensory Evoked Potentials (SSEP) were re-assuring, showing no changes from the patient’s pre-operative baseline.

We then performed a complex closure of the dorsal aspect of the dura using 4-0 Neurolon suture in running fashion. This was then covered with a piece of DuraGen and a layer of Tisseel was placed over it. Next, we closed the muscle and fasical layer with interrupted 0-Vicryl sutures and nylon sutures for the skin.

Follow-up

The patient reported urinary retention and urgency in the immediate post-operative period but was back to his pre-operative state by the 6-week follow up visit; he reported complete resolution of his urinary difficulties by the 4-month post-operative visit.

In the immediate post-operative period, the patient had some gait imbalance and required a front wheel walker for ambulation. He also reported urinary urgency and retention. He had 4/5 hip flexion, 4/5 knee extension, 4/5 knee flexion, 3+/5 EHL, 3+/5 dorsiflexion, 4/5 plantar flexion in the Left lower extremity; and 3+/5 dorsiflexion and 4/5 plantar flexion in the Right lower extremity. He was discharged to a rehabilitation facility and had
graduated to walking approximately 100 feet with the use of a front wheel walker upon discharge home.

By the 6-week post-operative visit, his motor strength was approximately 4+/5 in hip flexion, 4+/5 knee extension and knee flexion, 4/5 hip flexion, 4/5 dorsiflexion and 4+/5 plantar flexion in the left lower extremity; he also had 3+/5 dorsiflexion and 4+/5 plantar flexion strength in the right lower extremity. He was still using a front wheel walker for ambulation and his urinary retention and urgency had improved to his pre-operative state.

By the 4-month post-operative visit, he had some residual weakness with 3/5 dorsiflexor and EHL strength in the left lower extremity. He still used a front wheel walker for long distances and a cane for short distances at home. His urinary retention and urgency had completely resolved.

An MRI of the Lumbar spine obtained 6 months post-operatively showed that the spinal cord had migrated up to the L3 level with significant improvement of his syringomyelia Figure 3.

Outcomes

In a case series by Schijman [1] the author retrospectively reviewed 22 patients, 17 with SSCM type I and 5 with SSCM type II, who’s ages at diagnosis ranged between newborn and 20 years. These patients were divided into.

Three groups:

Group A: Patients with neuro-orthopedic abnormalities (9 cases). In this group, the most frequent signs were muscular atrophies and joint deformities in the lower limbs (4 cases) and scoliosis (5 cases).

Group B: Patients with cutaneous stigmata (6 cases). In this group the results of the neurological examination were normal in 4 patients, while 2 had an asymmetrical motor deficit in the lower limbs.

Group C: In this group there were 7 cases: 3 with hemi-myelo-meningocele, 3 with myelo-meningocele and 1 with meningocele. All these were surgically repaired soon after the patients’ birth. All 7 patients presented with a non-progressive neurological deficit attributable to the pre-existing dysraphism [1].

Of the seventeen patients operated on, three of the four patients that were neurologically intact pre-operatively remained intact after surgery, while the fourth patient had transient post-operative urinary retention that resolved after 20 days. The remaining thirteen patients had pre-operative neurological deficits but remained at their neurological baseline post-operatively.

Based on the progressive nature of symptoms in previously asymptomatic patients, and favorable surgical outcome obtained in his series, Shijman [1] advocates surgical intervention in all cases of Type I SSCM and in most cases of Type II SSCM, except in severely compromised patients with myelo-meningocele.

In a case series by a Mahapatra [11], the largest published case series on SCCM so far, the authors reviewed 300 cases of SCCM treated over a sixteen-year period. They reported improvement in neurological deficits in 50% of the patients, arrest of progression of neurological deficits in 44% of cases and neurological deterioration in 6% of cases in the immediate post-operative period. However, these patients had improved to their pre-operative baseline by post-operative day 10. The authors concluded that since the risk of developing neurological deficits increases with age, all patients with SCM should be surgically treated prophylactically, even if they are asymptomatic.

In a case series of 53 patients (30 with Type I SSCM and 23 with Type II SSCM) by Borkar and Mahapatra, the authors used a new sub-classification system proposed by Mahapatra and Gupta to further sub-classify patients with Type I SCCM and found it to have significant prognostic value.

- Type 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 lower down
- Type Ic. Bony spur at the lower pole with a large duplicated cord above
- Type Id. Bony spur straddling the bifurcation with no space above or below the spur [21].

Seven patients who were asymptomatic pre-operatively remained asymptomatic post-operatively.
However, eight of the asymptomatic patients had neurological deterioration post-operatively; five had deterioration in their strength and three had urinary retention. Five of these patients had type IId split, two had type IC split, and one had type Ib split. Among the eight patients who deteriorated post-op, four had improved to preoperative status by the time of discharge; the remaining four, all with a Type IId split, had long-term deficits. These results significantly substantiate recommendations by other researchers for early surgical intervention in all cases of SSCM [22].

CONCLUSION

Based on the progressive nature of neurological deficits in untreated cases of SSCM and the dramatic arrest in progression with considerable improvement in neurological deficits, many authors strongly advocate early surgical intervention once a diagnosis of diastematomyelia is made. Studies with longer follow-up periods may provide more knowledge on the long-term effects of surgical intervention and the incidence of re-tethering after surgical management of SSCM.

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REFERENCES