

Chapter 12

Split Cord Malformations



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12.1 Introduction

Split cord malformations (SCM) represent a subgroup of congenital abnormalities related to spinal dysraphism, in which the spinal cord is split into two hemicords along a portion of its length. They are relatively rare accounting for 3.8–5% of all spinal dysraphisms and are often associated with other forms of spinal dysraphism, mainly tethered spinal cord syndrome [1, 2]. Diagnosis is usually established during early childhood and only scattered cases are encountered during adulthood [3]. Due to the risk of neurologic impairment of patients suffering from split cord malformations, early diagnosis and proper management are mandatory.

12.2 Embryology

Gastrulation is the embryonic developmental process of gestation, through which all three germ layers are produced—ectoderm, mesoderm, endoderm. Ectodermal germ cells form the neural tube and neural crests from which the central and peripheral nervous systems are formed respectively in a process called neurulation.

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Mesoderm is responsible for the formation of the notochord, which gives rise to the nucleus pulposus, and the somites, thus ultimately forming the spinal column. Failure at different points during these developmental stages, results in various congenital abnormalities of the nervous system. Split cord malformation pathogenesis is thought to be initiated by the presence of an ecto-endodermal adhesion, which will eventually form two hemineural plates and two heminotochords, during neurulation [4].

12.3 Classification

Over the years, split cord malformation nomenclature underwent changes. Terms such as diastematomyelia, diplomyelia and dimyelia have been used in the past. Diastematomyelia describes a single cord, which is split caudally by a septum with two distinct dural sacs. Diplomyelia refers to an accessory spinal cord dorsal or ventral to the original one, encased in a single dural sac. Dimyelia describes the presence of two separated spinal cords with two distinct dural sacs. Pang et al. in 1992 proposed a classification system that described two distinct types of split cord malformations, split cord malformation type I and type II [5]. Split cord malformation type I consists of two distinct hemicords, each one encased in its own dural sac, which are separated from an anteroposterior bony or fibrocartilaginous spur. This type of malformation is usually located in the lumbar and lower thoracic spine. Split cord malformation type II refers to the presence of two hemicords contained into a single dural sac and separated intradural by fibrous bands [6]. This type of malformation can also be found in the cervical region. In 2005 Mahapatra and Gupta proposed a new classification system for type I SCM, based on the location of the bony septum that produces the split. The proposed classification includes four categories: Ia in which there is a bone spur in the center with an equally duplicated cord above and below the septum, Ib the bone spur is located at the superior pole with no space above it and a large duplicated cord lower down; Ic, the bone spur is found at the lower pole with a large duplicated cord above; and Id, a bone spur straddling the bifurcation with no space above or below the spur [7].

12.4 Relation to Other Abnormalities

Split cord malformations are often associated with several congenital abnormalities. These abnormalities arise from anomalies related to all three germ layers. Most of them represent complex craniospinal congenital malformations produced by ectodermal-endodermal abnormal adhesions. Common findings include myelomeningoceles, intramedullary lipomas, dermal sinuses, neurenteric cysts, hemivertebrae, Klippel-Feil syndrome and Chiari malformation [8]. Extra-craniospinal

abnormalities have also been reported, such as intestinal duplications and diverticula.

12.5 Clinical Presentation

The clinical features of split cord malformation include a wide range of manifestations. Some patients can be asymptomatic, but in most cases various symptoms can be seen. Both SCM types are tethering lesions. The majority of children may be asymptomatic at birth and neurological deterioration usually begins within the first 2 to 3 years of life [7]. Symptoms involve several neurologic deficits, deformities of the spinal column or the extremities and various cutaneous markers. Patients usually complain of lower extremities pain and persistent back pain. Neurologic deficits most commonly involve the lower limbs. Motor and sensory deficits such as weakness, atrophy of the lower limbs, gait disturbances, radicular pain, hypoesthesia or paresthesia represent common manifestations. Bladder and bowel disturbances can also be seen in about 20–40% of patients and should raise clinical suspicion [2, 7]. Skeletal deformity can also be present, usually in the form of scoliosis or kyphoscoliosis and are more common in SCM type I. Thus, all patients with congenital and progressive scoliosis should be investigated with MRI. Congenital talipes equinovarus is commonly encountered in these patients and should be carefully evaluated when present. Cutaneous markers can be seen in the form of capillary hemangiomas, subcutaneous lipomas, hyperpigmented patches, though hypertrichosis is the most common skin anomaly encountered [9]. SCM type I has more severe symptomatology, whereas in type II presenting symptoms might be subtle or can be incidentally detected [8].

12.6 Imaging

Diagnosis of split cord malformations is obtained through careful clinical evaluation and proper imaging techniques. X-rays of the spine have been traditionally used in the initial investigation of patients. Their usefulness relies in depicting several skeletal anomalies associated with split cord malformations, such as kyphoscoliosis, vertebral or rib anomalies, but lack sensitivity in diagnosing split cord malformations. Computed tomography can provide more detailed information related to skeletal pathology of the spine and can also demonstrate the presence of a bony or fibrocartilaginous septum, which is indicative of split cord malformation [Fig. 12.1]. The gold standard imaging modality is magnetic resonance imaging (MRI). Due to its superiority in demonstrating the neural elements, it can establish the diagnosis of split cord malformation by depicting the presence of hemicords and the dural sac or sacs containing them, depending on the type of malformation

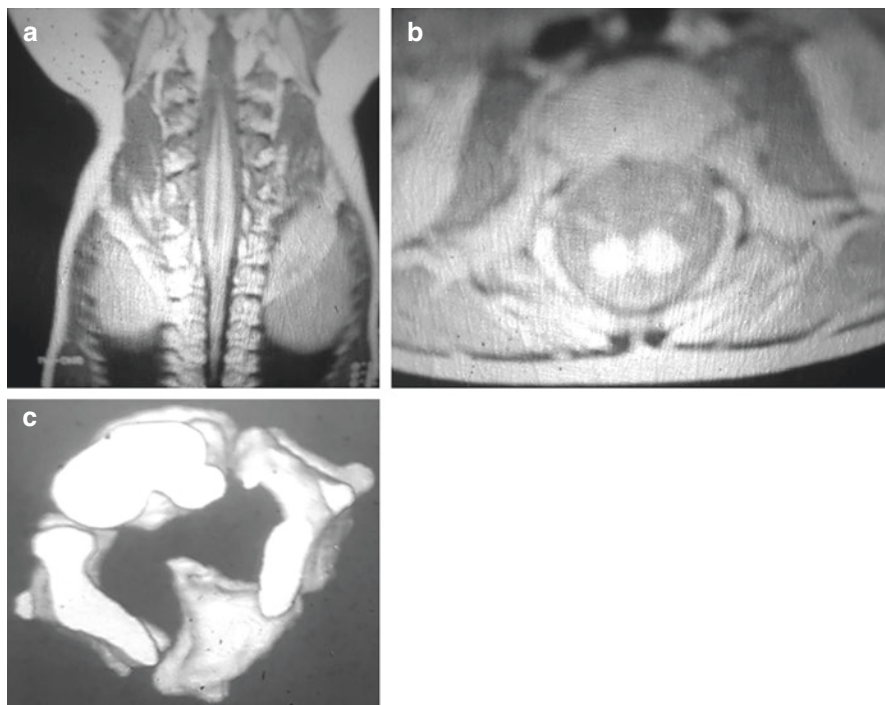


Fig. 12.1 Type I split cord malformation. (a) Coronal and axial (b) MRI revealing the two hemisacs. (c) 3D Computed Tomography (CT) reconstruction scan revealing the bony spur. The patient was operated via a laminotomy. The bony spur was dissected extradurally between the two dural sacs and removed piecemeal

encountered, as well as by demonstrating other anomalies related to split cord malformation, such as tethered spinal cord [10].

12.7 Management

The treatment of split cord malformation is mainly surgical. Patients with split cord malformations who remain untreated show increased rates of neurologic deterioration and have a low chance of complete recovery postoperatively. There is an association between increased age and the risk and severity of neurological deficits, due to tethering of the spinal cord. Thus, surgery is indicated in all patients with symptomatic split cord malformations and is also suggested in most asymptomatic patients at the time of diagnosis. The goal of surgery is spinal cord detethering and includes resection of the bony or fibrocartilaginous spur, removal of any other tethering attachments of the spinal cord, such as thick filum terminale, as well as management of any other coexisting craniospinal abnormalities. Type Ia is easier to treat with

surgery, whereas type Id is the most difficult to address surgically. In approximately 10% of type I SCMs cases, the bony spur may be diagonal, separating the canal into a large and a small compartment. Late symptomatic retethering is relative uncommon in adults but common in children. Reoperation should be proposed and provides years of relief [8, 11]. Severe scoliosis in type I SCM is among the most complex conditions in all types of spine deformities because the presence of the bony spur increase the risk of neurological deterioration during deformity correction surgery [12]. These patients are usually treated by two stage surgery. First, bony spur removal can be performed followed by scoliosis correction after 3–6 months. Recently, single-stage bony septum resection followed by spinal deformity correction, as well as, single-stage spine-shortening posterior vertebral column resection without prophylactic resection of bony spur have been performed with good results [12, 13].

12.8 Conclusion

Split cord malformations are rare congenital anomalies traditionally classified in two types. These malformations are often associated with other congenital abnormalities, that cause cord tethering and neurologic impairment. Careful clinical examination, early definitive diagnosis and timely surgical treatment are of paramount importance.

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