Trifid Cord; Very Rare Presentation of Split Cord Malformation

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Abstract

A child with a very rare anomaly of spinal cord is described. The patient presented at 9 months of age with trifid cord at thoracolumbar region and extensive split cord malformation (SCM) from T12-L5. She had a composite type SCM including type II at T12-L1 and SCM type I from L2, L3, to L4 levels. The condition seemed to be something like three distinct cords at T12-L1 level composed of a thin cord at left side terminating before L2 level to a tiny fibrous band, and two thick middle and right cords which joined together at conus at L5 level.

The patient had a large hairy patch on her back with asymmetric legs without any movements in her smaller foot. Cord untethering and removing the bony ridge causing SCM were performed with laminectomy of L2 to L5 and laminectomy of T12 and L1. Her neurological status was unchanged after surgery and remained stable during 6 month follow up.

Keywords: Split cord malformation; Trifid cords; Type

Introduction

Split cord malformation (SCM) is a rare close neural tube defect, mostly discovered during investigations for an abnormal skin marking especially hairy patch, foot deformity and/or weakness or scoliosis. In this malformation two hemicords can be separated with either a bony ridge or fibrous band which is classified as SCM type I or II, respectively. Some other rare types of SCM as intermediate and composite types have been reported, as well. In the intermediate type the two hemicords are separated with soft tissue bands and the intermediate hemicord tissue is bony only in a part of its length, particularly in the posterior portion. Composite-type SCM is characterized by different level SCMs with intervening normal cord in between, found in the same patient [1,2]. Here, we report a rare type of SCM in a 9-month-old girl presenting with foot abnormality, who was found to have something like three cords with SCM types I and II. To our knowledge, this is the first reported case of such triple cords with SCM of different types at three different levels.

Case Report

The patient was a 9-month old girl with a large hairy patch on her lumbar skin. Gestational period was uneventful and family history was unremarkable. She was recognized to have smaller left foot compared to the other side soon after birth.

Physical examination revealed the left lower extremity to be smaller than the right side, with no noticeable movements in the smaller foot. Other clinical examinations and developmental milestones were normal.

Lumbar spine magnetic resonance imaging (MRI) indicated split cord malformation below L1 level with two separate dural sleeves and one hemicord in each side. Left dural sleeve and hemicord were considerably smaller than the right side (Figure 1). In lumbar spine computed tomography (CT) scan, thick intracanal bony spurs were noted in L2, L3, and L4 levels. Low-lying conus at L5-S1 level was found as well (Figure 2).

Urologic evaluations revealed neurogenic type bladder with high pressure voiding, detrusor sphincter dyssynergia, and large amount of post-voiding residue in urodynamic study and VCUG, respectively.

The patient was scheduled for surgery to untether the cord at SCM level and also in distal part. Through a midline lumbar skin incision, paravertebral muscles were stripped and posterior elements of T12 to L5 were exposed. To prevent postoperative kyphosis we tried to do laminectomy as much level as possible. In levels with abnormal thick lamina or bony ridge causing SCM type I we had to perform laminectomy but in other levels without abnormal lamina osteoplastic laminotomy was done. Meticulous laminectomy of L2 to L5 and laminotomy of T12 and L1 were performed, through which both dural sleeves were exposed from L2 to L5. Removal of large thick bony spurs was carried out with high speed drill prior to dural opening, and subsequently intradural components were explored carefully. Laminectomy was performed at the levels of L2-5 that there were thick lamina and huge bony ridge.

Figure 1: Sagittal MRI shows the thick bony ridge at L2-4 levels with cord tethering.

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We found trifid cords at T12-L1 level; one hemicord was separated from right cord proximal to L1 and traversed to the left side, where was terminated to a thin fibrous band proximal to L2 level before dural duplication and was considered as an SCM type II. A nerve root was found along lateral side of this hemicord as well (Figure 3). Right hemicord was thick in the whole length and became duplicated for the second time at L1 level, alongside dural sleeves duplication. This time, the left side neural tissue was similar to a thin single root and was derived from the left border just beneath the previous left hemicord of SCM type II, arching the distance toward L2-3 level bony spur. This neural tissue became thicker passing from left dural sleeve and converted to a thick hemicord distal to L4 bony level (Figure 2). Afterwards, both hemicords joined each other and made a single cord which was terminated in a lipomatous and thick filum terminale. The bony spur, medial dura adjacent to bony spur in both sides, and all arachnoid adhesions were removed and the thick filum terminale was cut too. Patch of dural graft from lumbar fascia was used in order to make a large single dural sac. Postoperative period was uneventful. There were no changes in neurological status during 6 month follow up. She was prescribed lumbar orthosis for 3 months. Neurogenic bladder was managed through urologic consultation.

Discussion

The Current reported case is a very rare type of SCM with three cords, separated from each other at T12-L1 level at two different places, and one of them resembling SCM II was terminated to a thin fibrous band before L2 level after emerging one root from it. The other two cords, manifesting as SCM I, joined each other distally at L5 level and terminated to cauda equina and thick filum.

Split cord malformations are rare congenital malformations of the spinal cord with two known subtypes [3]. A number of theories have been proposed to explain the pathogenesis of this anomaly during embryonic period. One of the most accepted theories, proposed by Pang et al. and called as the unified theory, explains the origin of abnormality in formation of an accessory neuroenteric fistula between the yolk sac and amnion in the third week of gestation. The abnormal communication between ectoderm and endoderm results in cleaving the notocord and neural plate. Afterwards, concentration of mesenchymal cells around the accessory fistula forms a tract within the neural tube and dependent of its final composition, each of the subtypes will develop [2-5]. If endomesenchymal tract forms a bony septum, the result will be type I and if the mesenchymal tract degenerates to a fibrous band dividing the hemicords in a single dural sac, type II SCM will be formed [5,6]. Usually there is a single anomalous accessory neuroentric canal but rarely multiple canals may be present in early embryonic stage which results in two or more septa splitting the spinal cord into two hemicords in more than one level, making “tandem” or “composite” split cord malformations [6].

Another theory explains the etiology of anomaly as hyperproliferation of basal cells of the neural tube and then the walls fail to be merged as the result [7].

Some other researchers have suggested notochord cleavage as the etiology of SCM. Connection of endoderm and ectoderm as the result of notochord cleavage leads to double neural tube and hemicord formation [7]. Embryonic studies have revealed spinal cord widening at lumbar region and presence of ectodermal groove within two neural tubes in human embryos with diastematomyelia [1].

Whatever the etiology and embryonic nature, the composite type is extremely uncommon. Pang, et al. has reported two cases of composite type SCMs among 39 patients. Although one of the cases was believed to have tripartite cords separated by two parallel bony spurs at first glance, careful reconstruction of images demonstrated oblique bony spurs rostrocaudally settled along the length of involved cord segments and found to be two hemicords only [2]. Ersahin, et al. has reported four cases of composite type SCM among 74 patients. Two of four unusual SCM cases were found to have composite type supporting the role of multiple accessory neuroentric canals in the development of composite type SCMs [8,9]. Vaishya and Kumorjain have reported 3 cases of composite type SCM with different combinations of SCM typing and level of involvement [10]. Besides, Akay, et al. reported a rare composite type SCM with two different types at 3 levels [6]. Morya, et al. presented a unique variant of SCM with co-existing segmental spinal dysgenesis in which left hemicord with a small remnant of sub arachnoid space passed through an intervertebral cleft in a vertebral anomaly [11].

In our patient, we found a type II SCM at T12-L1 level with a large hemicord in the right side and a very thin left hemicord. Although
Hemicords commonly differ in size, in our patient the smaller hemicord was terminated in a thin fibrous band in caudal L1 level while the other hemicord was continued to L5. This pattern of asymmetric termination of hemicords is not usual however. We propose that this patient was a kind of composite SCM with type II at proximal (ie. T12-L1), and then a SCM I at L2-4 level with a huge bony spur. We think that both two cords at left side, including the short cord finished to a fibrotic band and the thin left cord joining distally to right cord at L5 level, should be one cord embryonically. Then a process like an infarct or degeneration should happened in this single left cord and the remained neural tissue could differentiate to these two cords.

We have encountered SCM type I in L2, L3, and L4 levels, splitting the right hemicord into two further separate parts with two dural sleeves which could be explained by the potential presence of an anomalous paramedian accessory neurtric canal, dividing the right neural plate in the first stages of embryonic life.

Unusual thickening of distal left hemicord may be due to aberrant neural fibers from right side, making a loop in distal part of left hemicord, before moving cephalad or caudal in the right cord. On the other hand, it could be correlated with lumbar enlargement of spinal cord as routine increase in cord diameter in cervical and lumbar area because of neuronal bodies of brachial and lumbosacral plexuses neural fibers.

In conclusion composite type of SCM is very rare. Here we report a complex type of composite SCM and described the surgical intervention To prevent more neurological deficits in such complex cases, meticulous laminectomy at the levels of SCM type I and careful intradural dissection for cord untethering are necessary.

References