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SPLIT CORD MALFORMATION IN ADULTS: SYMPTOMS, SURGICAL TREATMENT AND RESULTS

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ABSTRACT

Objective: This is a rare report of patients with clinically silent split cord malformation (SCM) unrecognized until adulthood. The symptoms of patients with SCM rarely manifests in adulthood. When it does, it is characterized by an acute neurologic change. There are insufficient studies in the literature on the natural history of adult patients with SCM. In this article, we retrospectively present the long-term postoperative follow-up of adult patients with SCM.

Materials and Methods: Patients operated with the diagnosis of SCM between 2000 and 2021 were evaluated. It was analyzed retrospectively. Patients without at least 6 months of follow-up and patients with incomplete epicrisis and radiological images were not included in the study.

Results: Ten patients were included in the study. Patients were followed up for 6-72 months (mean 37.5 months). All patients were female. The most common symptoms were pain in the legs, back pain, lower extremity weakness, and bladder dysfunction. It was found that the symptoms started with excessive physical activation for the first time. All patients were successfully treated surgically. All those undergoing surgery experienced symptomatic relief even at the initial follow-up.

Conclusion: SCM may be asymptomatic in childhood and symptomatic in adulthood. Neurological deficits may be attributed to traction injury derived from an osseous septum due to excessive physical activation. An excellent outcome may be obtained from the resection of the septum and untethering of the filum.

Keywords: Adult diastematomyelia, diastematomyelia, diplomyelia, syringomiyelia, split cord malformation

INTRODUCTION

Split cord malformation (SCM) is an extremely rare form of congenital spinal dysraphism[1]. There is a bony or fibrous septum extending anteriorly to posteriorly leading to the formation of two split cords, each surrounded by a dural layer or a single dural sac. It is often accompanied by spina bifida. It is usually diagnosed in childhood. Patients often have skin lesions. Neurologic deficits and spinal deformities may be seen in the lower extremities, bladder and intestines. Very rarely, aseptomatic dystraphism may be diagnosed incidentally[2].

Symptoms usually appear in patients in young childhood. Therefore, very few patients remain undiagnosed until adulthood[3]. There is also a very small group of patients whose complaints appear in adulthood[3-8]. Generally, there may be a history of trauma that may cause the onset of complaints. Pain is the most common complaint and mostly involves the perianal region. Loss of strength in the legs and urologic problems are also common[6,7].

Since this disease is very rare, there are no large case series in the literature. There are small series of articles or case reports. It is therefore difficult to find scientific data on how best to treat patients or improve their quality of life. To further elucidate this rare condition, 10 cases of SCM in adults are presented, and clinical presentation, diagnostic evaluation, management and outcome are discussed.

MATERIALS AND METHODS

We performed surgical intervention on 10 adult patients with SCM between 2000 and 2021. We retrospectively analyzed the medical reports of these cases. This study was approved by Erciyes University Institutional Ethics Committee (decision no: 2022/682, date: 12.10.2022). Only patients older than 18 years with new symptoms and SCM were included in the study. All patients underwent preoperative computed tomography (CT) and magnetic resonance imaging (MRI) scans. Postoperative MRI scans were obtained in all patients. All patients underwent laminectomy or hemilaminectomy for septum removal and dura repair. After the dura was stripped

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from the bony spur, the spur was removed. The medial side of the dura of the hemichords was opened and cut. Connective formations such as fibrous band and dentate ligament were sectioned. Associated intraspinal pathologies such as dermoid and epidermoid cysts were also removed as appropriate. The filum was cut distally in all patients. A waterproof dura closure was performed (Figures 1-4). Resection of the septum was always performed before dissection of the filum to prevent ischemic damage from spinal cord retraction.

Data were analyzed in terms of demographic characteristics, admission characteristics, radiological findings and follow-up outcomes. Pain, motor deficit, sensory deficit and urinary symptoms were evaluated at follow-up.

Statistical Analysis

No statistics were made due to the low number of cases.

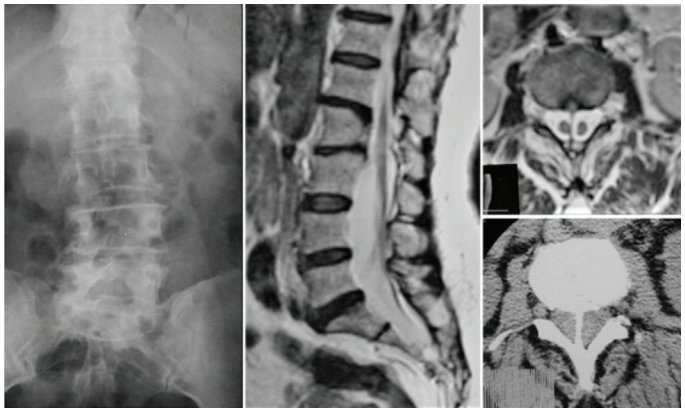

Figure 1. A 64-year-old female applied with complains of pain, numbness, weakness, neurogenic claudication and incontinence in legs after running. Plain X-ray revealed a bony spur at L2-L3 levels. CT showed a bony spur extending from the lamina of L2 through the midline of the spinal canal to the posterior margin of the L2-L3 disc space. MRI showed a bisection of the spinal cord and conus medullaris were tethered to L4 level
CT: Computed tomography, MRI: Magnetic resonance imaging

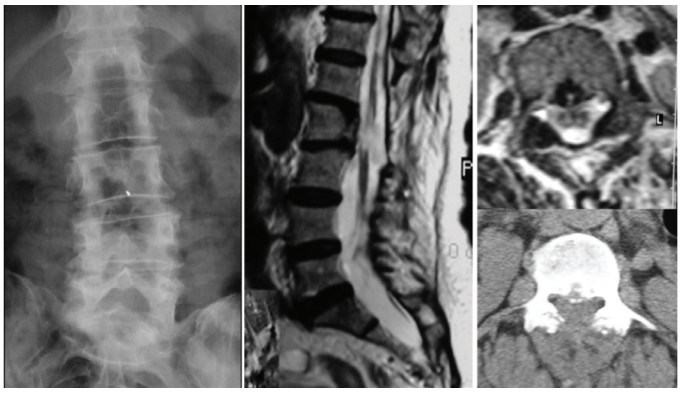

Figure 2. At 12 months follow-up, she returned to her job, she reported complete resolution of her back and legs symptoms but incontinence continues. MR and CT shows bone spur removal and no tethering of spinal cord
CT: Computed tomography, MRI: Magnetic resonance imaging

RESULTS

There were 10 female patients and no male patients. The mean age of the patients was 42.1 years (range 19 to 64, median value 43). The mean follow-up period was 37.5 months (range 6-72 months, median value 28). Pain was the predominant presenting feature in most cases. Eight patients presented with low back pain and all patients presented with leg pain. Sphincter dysfunction was present in 5 patients. Paraparesis was present in 2 patients and motor weakness in the lower extremities in 5 patients. Six of the patients had sudden onset of low back and radicular pain, neurogenic claudication and paraparesis after traumatic events such as sports activities or heavy lifting. SCM was associated with syringomyelia in 3 patients, dermoid cyst in 2 patients, neuroenteric cyst in 1 patient, congenital vertebral fusion in 2 patients and scoliosis in 3 patients.

In the postoperative period, 6 of 8 patients with low back pain and all 10 patients with leg pain were improved. All 4 patients with motor deficits were improved. However, sphincter deficits

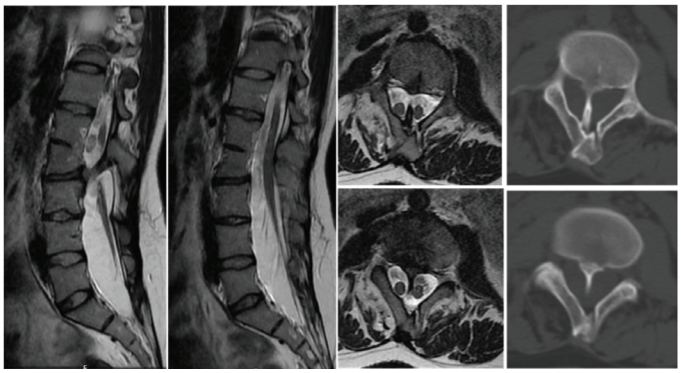

Figure 3. A 29-year-old female applied with complains of low back pain, sometimes right leg pain for 6 months. It was increases when leaning forward. Bilateral foot thumbs were dorsal fleksion 3/5. CT showed a bony spur extending from the lamina of L1 through the midline of the spinal canal to the posterior margin of the L1-L2 disc space. MRI showed a bisection of the spinal cord and conus medullaris were tethered to L4-5 levels
CT: Computed tomography, MRI: Magnetic resonance imaging

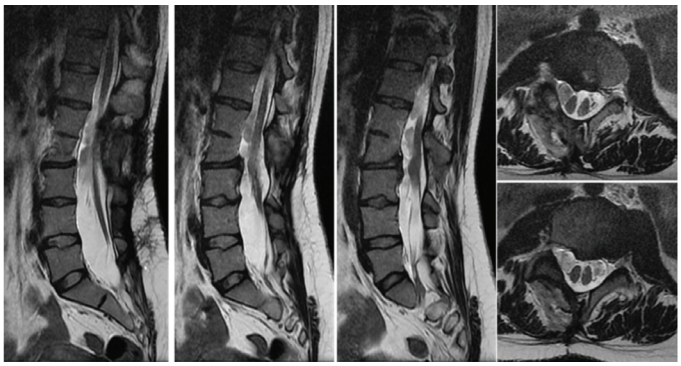

Figure 4. At 8 months follow-up, she had no symptoms. MR shows bone spur removal and no tethering of spinal cord
MR: Magnetic resonance

were improved in only 2 of 5 patients, and improvement could not be documented in the other 3 patients. One patient was continued to have frequent urination postop (Table 1). No postoperative complications were observed.

DISCUSSION

SCM is a rare form of spinal dysraphism characterized by a single cord cranially and caudally, a duplicated cord centrally, and two distinct dural sacs separated by a septum. It may be a bony, cartilaginous or fibrous septum.

Pang et al.[1] categorized SCMs into two different groups: Those in which one or two dural tubes are present, and those in which the medial septum is rigid or not rigid. In type I SCM, the separation of the osseo-cartilaginous septum results in two dural tubes. Type II SCM refers two hemichords separated with a non-rigid septum, both located within a single dural tube. Huang et al.[8] reported that 78.8% of patients had type I SCM and 21.2% had type II SCM. In our study, 90% of the patients were type I SCM. This is in contrast to the review by Karim Ahmed et al.[2] in which type I and II SCM represented 54.5% and 45.5% respectively.

Patient Demographics and Associated Conditions

In adults, SCMs are most commonly found in the lumbar spine (51.9%), followed by the lumbosacral spine (16.9%) and thoracic spine (13.2%)[3]. In our study, 90% of the patients were in the lumbar spine.

In a review of the literature consisting of 146 cases of adult SCM[3], it was reported that 25.3% of the patients were male with a mean age of 26.8 years at first presentation. The most common associated condition was tethered cord syndrome (59.8%), followed by hypertrichosis (44%) and epidermoid cyst (14.1%)[3]. In our study, all patients were female and the mean age at first presentation was 42.1 years. The most common associated conditions were tethered cord syndrome (42%) and syringomyelia (42%), scoliosis (28%), epidermoid cyst (14%) and neuroenteric cyst (14%).

Traditionally, SCMs were believed to be pediatric disorders defined by the onset of neurological deficits in early childhood. The presence of scoliosis, lumbar skin patches, progressive foot deformities, calf and foot atrophy, and bowel, bladder and gait disorders in the pediatric population draws attention to the possibility of SCMs. These are caused by traction on the conus with elongation of the spinal column in the presence of a taut cord[5-7,9-13]. Although this condition originates in embryogenesis, there is a poorly characterized subset of SCM patients who subsequently become symptomatic in adulthood[12]. Few cases have been documented in the adult population[1,5,7,8,10,12,14-22].

The etiology of diastematomyelia symptoms in adult patients is not clearly understood. Progression of neurologic dysfunction, back pain, spinal cord and cauda equina dysfunction may be seen in both childhood and adult patients. In addition to these symptoms, most of these patients have midline skin anomalies such as skin hair growth hemangiomas, lipomas and sinus tracts associated with spinal dystrophism[5,7,8,10,12].

Especially in the presence of anorectal anomaly (67%), meningocele manqué (54%) and diastematomyelia (38%), there is a high rate of syringomyelia with occult spinal dysraphism with a tethered spinal cord from a tight filum terminale[23]. Many theories have been presented to explain the origin of terminal syringomyelia. Terminal syringomyelia was found in 2 of our patients and cervicothoracic syringomyelia was found in one. The epicenter of terminal syringomyelia is almost always rostral and close to an occult spinal disraphic defect, so its pathogenesis is more likely to be related to the spinal disraphic lesion[23]. However, we did not find a relationship between cervicothoracic syringomyelia and lumbar diastematomyelia.

Clinical Features

The most common symptom in patients is low back and leg pain (68.5%), radicular pain and paresthesia (51.8%), and lower extremity weakness (50.9%)[3]. In our study, the most common symptoms were leg pain (100%) and low back pain (80%).

Neurologic symptoms are thought to result from the movement of the spinal cord within the spinal canal and subsequent local injury to neural elements due to traction by the dentate ligament[11], bony prominence[24] or tethered cord. In patients with SCM in adulthood, there is usually a trauma that forces flexion and extension of the spine before the onset of symptoms. During this trauma, the septum locally damages the spinal cord. Trauma has been documented to be a triggering factor in this disease[8,24]. Our cases also support this concept. In our study, the onset of SCM symptoms in 6 of 10 patients was associated with acute events such as aggressive sports and heavy lifting. There was no neurocutaneous stigma.

Imaging Features

X-ray evaluation is the first step in the radiologic evaluation of patients with SCM. With these images, pathologies that can be found in adult SCM patients such as scoliosis, spina bifida, tapering of the vertebral corpus, and partial anomalies in the vertebrae are seen. CT may be useful in identifying the midline osseous/osteocartilaginous septum and bony abnormalities (e.g. butterfly vertebra, hemivertebra, Klippel's Feil, spina bifida). CT scan is the preferred tool to elucidate the bony anatomy of the deformity. MRI is useful for identifying SCM and visualizing neural elements. MRI can also reveal tethered cord malformations associated with SCM, intramedullary lipoma, hydromyelia, Chiari malformations, and meningocele/myelomeningocele[1,14,22,24,25]. MRI is preferred to visualize the cord and conus region and to detect other intraspinal abnormalities associated with spinal dysraphism[4,8,10].

Treatment

Karim Ahmed et al.[2], reported that neurologic function in adult SCM remained unchanged in 90% and worsened in 10% in

Table 1. Summary of the patients

No	Age	Sex	SCM levels	Conus levels	Associated pathology	Initial symptoms	Inkontinans	NC	Skin sign	Low back pain	Leg pain	Paraparesis	Lower extr weakness	Symptom provacation	Follow-up
1	28	F	T11-12 (type II)	L2-3	Syringomyelia (C7-T1), scoliosis	SA					+/-		+/-	Walking	12
2	51	F	L2-3	L3-4	Syringomyelia (T12-L1)				+	+/-	+/-			Walking	72
3	55	F	L4	L5-S1	Dermoid cyst (L3-4)		+/+		+	+/+	+/-				14
4	29	F	L2-3	L4	Enteric cyst (L2), congenital vertebral fusion, scoliosis	HL				+/-	+/-	+/-	+/-	Bending	26
5	64	F	L2	L3	Syringomyelia (C7-T3), Klippel Feil syndrome, congenital vertebral fusion	SA	+/+	+		+/-	+/-	+/-	+/-	Bending	84
6	53	F	L1-2	L5			±/-			+/+	+/-			Bending	72
7	52	F	L4-5	S2		SA	+/±			+/-	+/-		+/-	Working	6
8	19	F	L2-3	L4	Scoliosis					+/-	+/-			Walking	30
9	35	F	L2	L5-S1	Dermoid cyst (L5-S1)	SA	+/±		+	+/-	+/-		+/-	Bending Working	33
10	35	F	L4	L5		HL					+/-				26

NC: Neurogenic cloudication, HL: Heavylifting, SA: Sport activities

the conservative treatment group. In the surgical group, pain improved in 91.1%, remained unchanged in 7.1% and worsened in 1.8%. Russell et al.[7] also reported that surgical intervention resulted in symptomatic improvement in 95.8% of patients who underwent surgical treatment. Therefore, symptomatic adult SCM should be treated surgically. Postoperative complications occurred in 4.3% of all surgical cases and reoperation was required in 2.1% of cases[3].

Prophylactic spur removal and ligament release are recommended in children with SCM. For complete bone spur removal, wide dural opening and duraplasty are often necessary[4,8,12,26]. The role of surgery in adult patients is controversial. In symptomatic patients, the bony septa must be removed to prevent neurologic damage. Routine prophylactic removal is not recommended in asymptomatic patients[7,12]. As seen in our patients, there may also be a return of neurologic findings after surgical removal of the bone spur. Hazneci et al.[27] reported that surgical resection should be performed with neuromonitoring in patients with SCM associated with spinal teratoma and that the results were better in surgeries performed at an early age.

Akay et al.[13], reported that tethering structures requiring surgical intervention are more common in type I SCM. There is an influential view that these malformations are caused by fibrous septa or bony protrusions that interfere with the attachment of both hemicords. Therefore, these attachment defects are thought to be associated with neurologic symptoms. In such cases, surgery aims to remove the septum that prevents the hemichords from connecting to each other and to free the hemichords. In addition, if the patient has a thickened filum terminale or inferior location of the conus, a filum sectioning and releasing is required. The location of the conus should be determined preoperatively[26]. In our cases, connective formations such as fibrous bands, dentate ligaments were excised, and the filum terminale was sectioned and released. In addition, associated intraspinal pathology such as dermoid and epidermoid cysts.

In our study, all symptomatic adult SCM cases were operated. Those operated on underwent hemilaminectomy or laminectomy, septum resection and dural repair and dissolution of the filum. Resection of the septum was always performed before dissolution of the filum, thus preventing ischemic damage from cord retraction against the bony septum by a suddenly dissolving cord. In our study, operative treatment resulted in recovery in all 4 patients (100%) with preoperative neurologic deficit.

Study Limitations

The fact that the number of cases in this study was 10 is a limiting factor, but it should be kept in mind that a very rare disease was analysed. Another limiting factor is that the study was performed retrospectively. The follow-up period of the patients may be longer.

CONCLUSION

SCM is an extremely rare spinal dystrophism characterized by caudal separation of a spinal cord into two or more cores. The disease is diagnosed by skin lesions or neurologic deficits in early childhood and was considered a childhood disease. There is a poorly characterized subset of SCM patients who subsequently become symptomatic in adulthood. When these patients are operated on, successful outcomes are achieved for pain and strength loss, while urinary incontinence does not appear to benefit satisfactorily. It can be said that long-term follow-up after surgery in these patients is also meaningful in terms of revealing the results.

Ethics

Ethics Committee Approval: This study was approved by Erciyes University Institutional Ethics Committee (decision no: 2022/682, date: 12.10.2022).
Informed Consent: Retrospective study.
Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: M.M., Concept: H.U., A.Ş., R.K.K., Design: A.K., H.U., Ş.O., R.K.K., Data Collection or Processing: M.M., R.K.K., Analysis or Interpretation: A.K., H.U., Ş.O., Literature Search: M.M., A.K., A.Ş., Writing: M.M., Ş.O., R.K.K.
Conflict of Interest: The authors have no conflicts of interest to declare.
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