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CASE REPORT

Duplication of spine with hemi-lipomyelomeningocele

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Abstract
Background Duplication of the spine is very rare, and this malformation is generally considered as a severe form of type I split cord malformations. To the best of our knowledge, this is the first reported case of spine duplication associated with lipomyelomeningocele.
Case We report an exceptional case of 14-year-old, asymptomatic and neurologically intact girl with duplication of the spine and marked separation of bony elements at thoraco-lumbar region. One of the split thecal sacs includes a tethered spinal cord whereas other thecal sac has no visible neural content, and there is a neighbor lipomyelomeningocele located in the midline.
Conclusion A surgical operation was planned to release the tethered cord and instrumentation and fusion for scoliosis; however, the operation was declined by the patient.

Keywords Scoliosis · Spinal dysraphism · Multislice computed tomography · Magnetic resonance imaging

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Introduction

Duplication of spine with marked separation of bony elements is a rare malformation, and there is limited number of cases published as case reports in the literature [1–5, 8–11, 14]. According to classification by Pang et al. at 1992 [12, 13], this malformation is considered as a severe form of Type 1 split cord malformations (SCM). However, Pang et al. have not reported any patients with such extensive duplication of the bony elements. Therefore, some authors define this entity as unclassified today. We report a neurologically intact adolescent case of thoraco-lumbar duplication of spine with a hemi-lipomyelomeningocele. To the best of our knowledge, this is the first reported case of spine duplication associated with lipomyelomeningocele.

Case report

A 14-year-old Kosovo female patient was referred to our outpatient clinic of institution due to spinal deformity. She had a full-term normal vaginal delivery history. Her parents were healthy without genetic disorders. Her weight and height were in normal range. She had spinal deformity diagnosis in her first year of life. Unfortunately, the spinal deformity was progressed since then. She had a surgical resection of a focal lump at the lumbar region at the age of 6 months. Since the medical reports for the patient could not be retrieved, details of the operation and nature of the excised tissue were unknown. The patient's father depicted that she did not have any cutaneous defect and any complaint except the aforementioned lump at that time. The patient, without consanguineous marriage of her parents, had three siblings and none of them had a similar health

problem; however, her uncle also had scoliosis background.

On physical examination, her weight was 145 cm and height was 45 kg. Her intelligence and speech were completely normal. She had no facial dysmorphism. Detailed examination of the musculoskeletal system was normal except the spinal deformity. Hypertrichosis at lower lumbar posterior region especially located in the midline was noted on physical examination. Examination of the spine in coronal plane revealed left-sided thoraco-lumbar scoliosis, left-sided rib hump, shoulder asymmetry, and marked truncal shift. Severe kyphosis at thoraco-lumbar junction and increased lumbar lordosis were seen on the examination of the spine in sagittal plane. Her spinal deformity was also rigid in bending and axial traction. Separate two spinal processes line were palpated below the thoraco-lumbar junction. Her upper and lower extremities were neurologically intact, including reflexes and skin sensation.

Plain radiographs and CT examination performed with a 64-slice multidetector CT scanner (Aquilion 64, Toshiba Medical Systems Corporation; Tokyo, Japan) revealed marked duplication and separation of the spine after T8 vertebra (Fig. 1). Both spine components consist of incomplete dysmorphic vertebral elements, especially on the left side. The right-sided vertebral elements were showing continuity with lumbosacral vertebras whereas the left-sided spine component, consisting of more defective eight vertebral elements and showing marked rotation, was terminating near the level of L4–L5 vertebra. Other findings on CT were rotoscoliosis, T5 hemivertebra, T8 butterfly vertebra, partial fusion of vertebral bodies on L4–L5 level, and multiple posterior vertebral fusion abnormalities on lumbosacral levels. On the anteroposterior radiographs using the method described by Cobb, right thoraco-lumbar scoliosis was measured 87° between T3 and L3 level. There was a 37° compensatory left lumbar curve below the main curve.

MRI examination was performed by a 1.5 T MRI scanner (Signa Excite HD, GE Healthcare, Milwaukee,

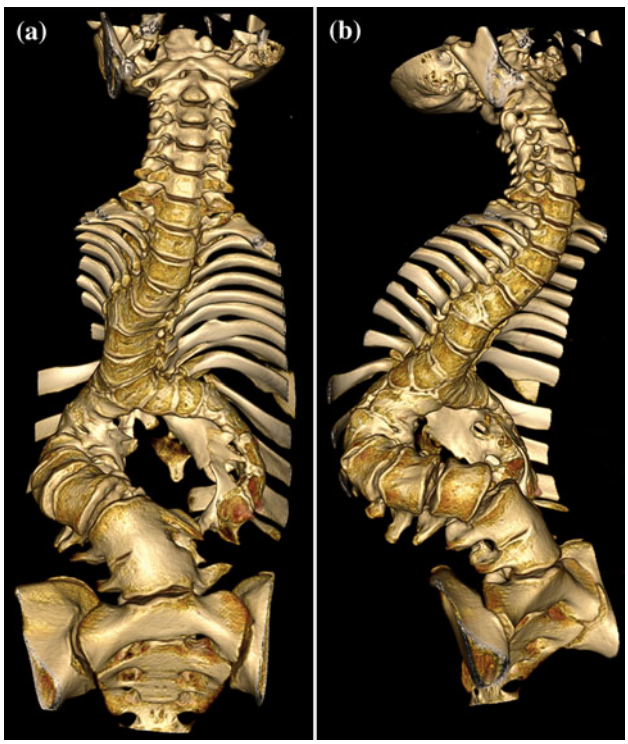

Fig. 1 Coronal (**a**) and right oblique (**b**) 3D volume rendering CT images show marked duplication and separation of the thoraco-lumbar spine and rotoscoliosis

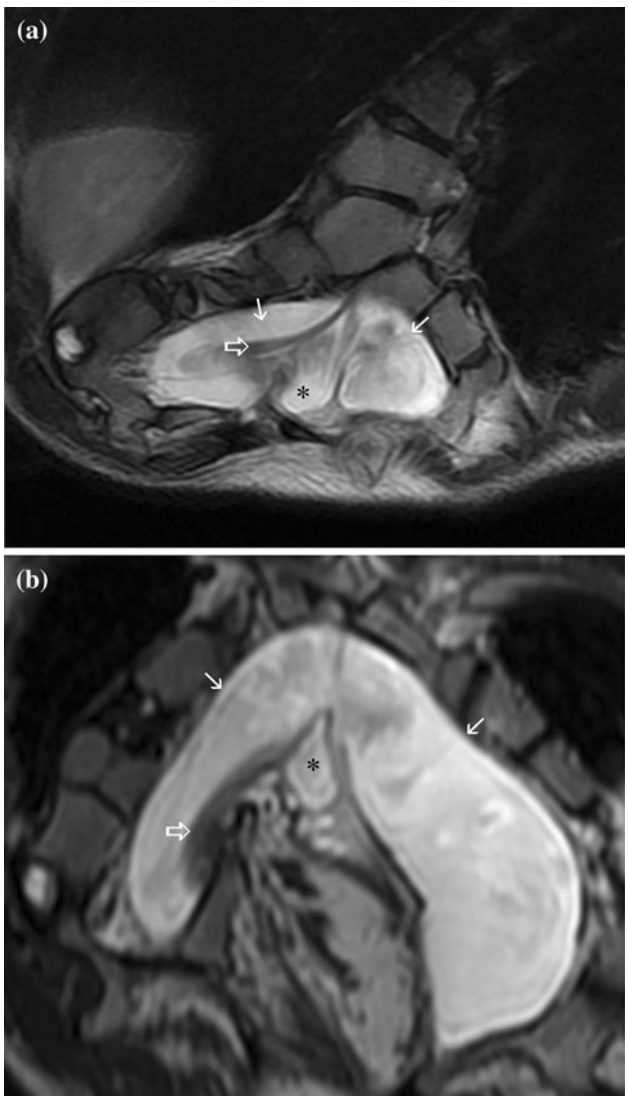

Fig. 2 T2-weighted axial (**a**) and coronal (**b**) MR images show splitting of the thecal sac into two discrete sacs (*arrows*) where each one of them follows the duplicated spine components. The right-sided thecal sac includes spinal cord (*open arrow*). Note a third cystic structure containing neural elements and surrounded by fat tissue located in the midline (*asterisk*)

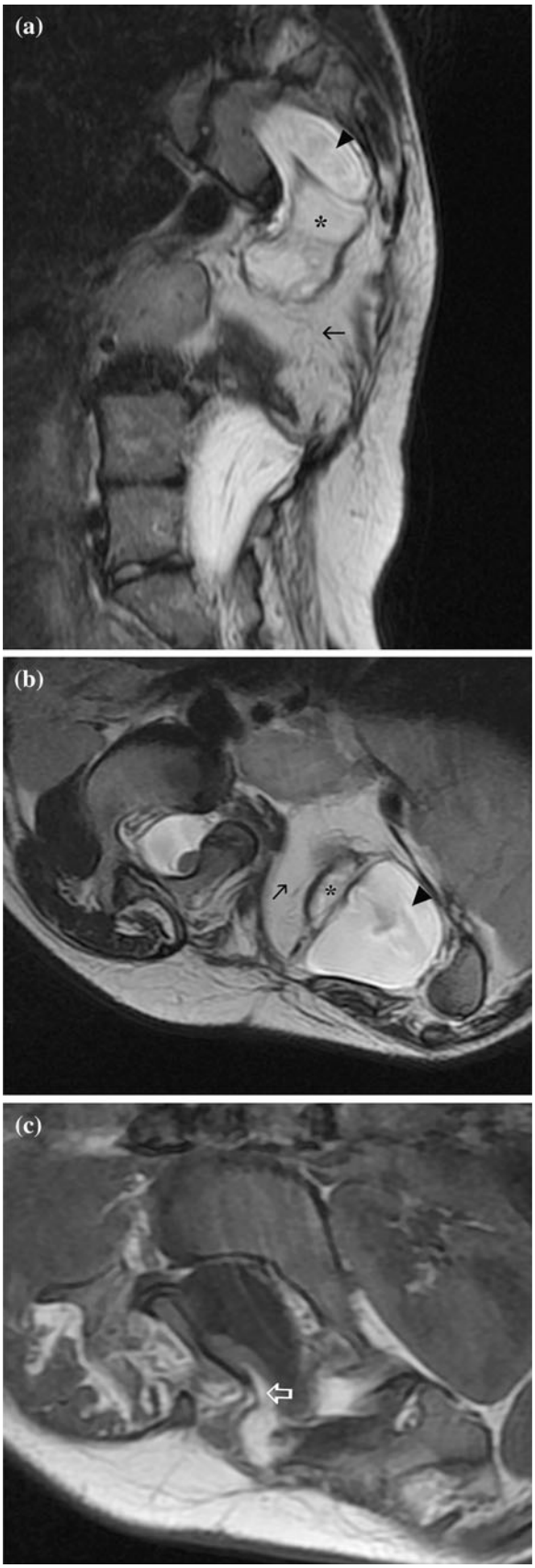

Fig. 3 T2-weighted sagittal (**a**) and axial (**b**) MR images show the lipomyelomeningocele sac (*asterisk*) and surrounding fat (*arrow*) adjacent to the left-sided thecal sac (*arrowhead*). T1-weighted axial image (**c**) shows the fat tissue (*open arrow*) enters into the right sided thecal sac and adheres to the tethered spinal cord

Wisconsin, USA). Below the T8 level, the thecal sac splits into two discrete thecal sacs; each one of them followed the duplicated spine components (Fig. 2). A third cystic structure containing neural elements and surrounded by fat tissue located in the midline, is adjacent to the left-sided thecal sac. This lipomatous tissue whose intensity was slightly different compared to subcutaneous fat, continues with left pararenal fat at anterior side, enters into right-sided thecal sac and adheres to the spinal cord at L4 level in the inferior side (Fig. 3). Due to the neural content that arises from the spinal cord at the duplication level that can be followed until the cyst-fat tissue interval, this cystic-lipomatous lesion assumed as a lipomyelomeningocele. The left-sided thecal sac which had no visible neural content conjugated again with the right-sided thecal sac at the L4–L5 level. A tethered spinal cord structure originated from spinal cord at the duplication level and terminated at L4–L5 level can be seen in the right-sided thecal sac. The nerves could be traced into bilateral neural foramina along the right-sided spine.

Except the spine duplication, no other duplication of abdominal structures was detected. A surgical operation was planned to release the tethered cord and instrumentation and fusion for scoliosis; however, the operation was declined by the patient.

Discussion

Pang et al. proposed a theory of embryogenesis of double spinal cord malformations based on the surgical findings of 39 patients with SCM and two postmortem cases [12, 13]. According to this "unified theory", the whole spectrum of split cord syndromes originates from one basic ontogenetic error occurring around the time when the primitive neuroenteric canal closes. This basic error is the formation of an "accessory neuroenteric canal" through the midline embryonic disc that enables continued contact between ectoderm and endoderm within the canal. This abnormal fistula which is subsequently invested with mesenchyme to form an endomesenchymal tract causes regional splitting of the notochord and the overlying neural plate. Final appearance of matured SCM depends on the ability of the embryo to heal around the endomesenchymal tract, the variable extent to which the endomesenchymal tract persists, and the ultimate developmental fates of the dislocated

midline mesenchyme and endoderm. Pang et al. detailed a new classification for SCM into types I and II according to whether the hemi-cords are within double sacs separated by a rigid osseocartilaginous spur or within a single dural sac with a fibrous midline septum, respectively.

Duplication of the spine is very rare, and this malformation is generally considered as a severe form of type I SCM on the limited case reports in the literature. Ahmed et al. [1] reported a case of 6-year-old, asymptomatic and neurologically normal girl except a lump on her back, which is the most similar case to our case. She had similarly extensive duplication of the spine from T9 to L5 level, each containing a thecal sac and hemi-cord. The subcutaneous mass was a large lipoma that attached to both thecal sacs. Cebesoy et al. [5] also reported a 44-year-old asymptomatic man that had incidentally found with lumbar spine duplication. Goldberg et al. [8] reported a case of an asymptomatic 13-year-old girl with scoliosis and whose spine shows partial duplication at lumbosacral region. A distal large lipoma with intradural extension through a sacral vertebral cleft, a neuroenteric cyst in the distal thoracal region, and a single kidney were the other findings of this case. Incesu et al. [10] reported another adolescent case of a 15-year-old girl complaining of back pain, and diagnosed asymmetric lumbar spine duplication with spinal cord tethering secondary to a filum lipoma in the sacrum. The other cases of duplication of spine were symptomatic with neurovascular, genitourinary, or gastrointestinal abnormalities [4, 9, 11]; some of these cases could be regarded as a form of caudal duplication [2, 6, 14] and in some cases, there is a cleft in the spine rather than two formed spinal columns with two hemi-cords [3]. To the best of our knowledge, any of the reported cases of duplication of spine describes a lipomyelomeningocele as in our case.

The limitation in our case is the inability to reach the old medical reports including the operation procedure of the patient. Patient's father clearly depicted that there was no any cutaneous defect, and the operation is limited to resection a subcutaneous lump. Depending on patient's anamnesis and current imaging findings, we guess that the operation was limited probably to the subcutaneous lipomatous tissue.

The surgical treatment for SCM is to release the tethered hemi-cords by removal of bony spurs, dural sleeves, fibrous septae, or any fibro-neurovascular bands (myelomeningoceles manque) that might transfix the split cord and anterior/posterior spinal fusion with instrumentation for kyphoscoliosis [5, 12]. It is generally argued that all patients with SCM should be surgically treated due to the concerns regarding the clinical consequences of the tethering of the cord, which is common in all of these syndromes [7, 13]. However, none of the asymptomatic cases of duplication of the spinal cord reported in the literature undergo surgical management [1, 5, 8, 10]. We decided for surgery including intraspinal procedures and fusion with instrumentation; however, she denied it due to neurologic risks of surgery.

Conflict of interest None of the authors has any potential conflict of interest.

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