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CASE REPORT

Cervicothoracic spine duplication: a 10-year follow up of a neurological intact boy

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Abstract

Purpose Spine duplication is a very rare condition with the literature being composed of only case reports. All previously reported cases were thoracolumbar spine duplications. Here, we report cervicothoracic spine duplication in a neurological intact male. According to our knowledge, it is the first case in the literature of cervicothoracic spine duplication.

Clinical presentation A 3-year-old patient presented to a primary physician with a complaint of short stature. He was referred to our department with suspected spinal deformity. Computerized tomography imaging revealed anterior bony structure duplication and posterior dysmorphic elements at the C5–T9 levels. Magnetic resonance imaging revealed a syrinx cavity which splits cord at the duplication level and the relation of the syrinx with posterior mediastinum through anterior bone defect. He was followed up for 10 years.

Conclusion In the literature, spine duplication has been classified as a severe form of split cord malformation because of the concurrence of bone duplication with split spinal cord malformation (SCM). This case presents a distinct form of SCM which shows non-duplicated dural tube as unclassified and cervicothoracic duplication level without neurological deficitis. Treatment of SCM was based on removal of splitting fibrous/osseous process. Neurologic intact spine duplication could be followed up without surgical intervention.

Keywords Cervicothoracic spine duplication · Split cord malformation · Syrinx

Purpose

Spinal column duplication is a rare abnormality of the vertebral column. According to a literature review, the oldest report dates back to 1980 [1]. After case report by McKay and Nason, many published papers reported neurological symptomatic or asymptomatic thoracolumbar and lumbar spine duplications [2–7].

In the literature, spine duplication (SD) has been classified as a severe type of split cord malformation because of the accompaniment of bony duplication with split spinal cord malformation. Most of the previously reported cases of SD involved two hemicords in separated dural tubes with bone duplication, which were classified as type 1 split cord malformation (SCM) [2–7].

Here we present the first case of cervicothoracic spine duplication in the English literature. The patient was diagnosed with the disease when he was 3 years old, and was closely followed up for 10 years. He has a single enlarged dural tube with a syrinx cavity at duplication level. Historically, treatment of SD based on removal of fibrous/osseous intramedullar splitting process. As previously reported cases without neurologic deficits, cervicothoracic duplication also could be followed up conservatively.

Clinical presentation

After informed consent had been obtained from patient's family, and institutional review board approval was obtained from local institution (44-314 January 19, 2016), the patient's hospital chart was reviewed and presented in this paper.

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We report the case of a 13-year-old male patient who was first diagnosed with cervicothoracic spine duplication when he was 3 years old. He had a short body stature compared with his peers and was taken to a primary health care center by his family. He was referred to our spine center with a suspicion of spine abnormality.

He had a history of full-term normal vaginal delivery. His parents were healthy without any genetic disorders or consanguineous marriage. The patient's sister had no spine abnormality and was healthy. The patient had a short stature compared with his peers. He had a height of 127 cm and weighed 37 kg. He was stage 3 according to Tanner scale. He had no facial dysmorphism and his intelligence was normal. Physical examination of the musculoskeletal system revealed a normal presentation, except for spinal deformity. Coronal plane spine examination did not show shoulder asymmetry, thoracic hump, or pelvic obliquity. Sagittal plane examination showed thoracic hypokyphosis (C5–T12 Kyphossi angle: 22°) with nearly normal lumbar lordosis (Lumbar Lordosis Angle: 51°) (Fig. 1). The processus spinousus were hard to palpate in the cervicothoracic region, but could be palpated easily in the lumbar region. Neuromotor evaluation revealed that light touch, pin prick, proprioception, and vibrational sensations were intact. Reflexes in all extremities were symmetric without hyperreflexia or clonus. He was found negative for Romberg, Hoffman, and Babinski signs and the rectal sphincter tone was normal. Bilateral upper extremity and lower extremity muscles were grade 5 according to manual muscle testing, without weakness and muscle atrophy. Pigmentation or hypertrichosis was not seen on physical examination. The patient underwent detailed evaluation of his gastrointestinal and genitourinary systems for abnormalities and the results were normal for both systems. His pulmonary function test revealed mild obstructive pulmonary disease [Forced air volume expelled in 1 s FEV1 (L/s): 0.62; total forced air volume FVC (L/s): 0.85; FEV1/FVC: 79.8%] without clinical significance. Total SRS 22R score was 4.15 without management domain and 4.4 in pain domain. He is attending the standard education schedule and daily sports activities with success like his peers.

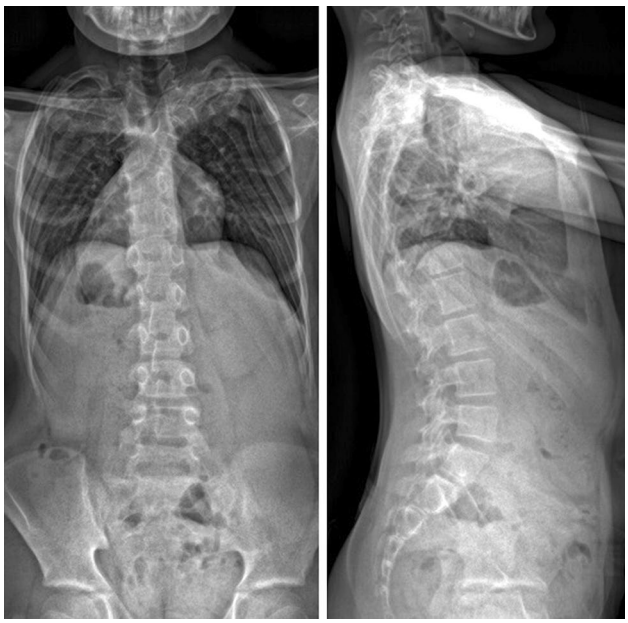

Fig. 1 EOS imaging standing anteroposterior image showed well balanced shoulder and pelvis without limb length inequality.EOS standing lateral image showed nearly normal sagittal alignment of spine except the hypokyphosis at duplication level

The patient underwent EOS, computerized tomography (CT), and magnetic resonance imaging (MRI) of the whole spine including the craniocervical junction. Sagittal spinal variables measured with a digital software [Surgimap Spine (Nemaris Inc., New York, NY, USA)] lateral standing EOS [C2–S1 height: 33.23 cm C2–T12 height: 18.69 cm lumbar lordosis L1–S1: 51° pelvic incidence: 59° sacral slope (SS): 36° pelvic tilt (PT): 23° sagittal vertical axis (SVA): − 15.8 mm coronal balance (Cor Bal.): + 9 mm]. He was grade 2 according to Risser classification. CT examination revealed marked duplication of the spine between C5 and T9 vertebra. Both spine components consist of incomplete dysmorphic vertebral elements, each of the duplicated spinal column was steered laterally and showed fusion abnormalities (Fig. 2). There was a bone defect with a dimension of 2 cm at the duplication level. Through this bone defect, the anterior epidural area was close to the posterior mediastinum. The spinal cord was a single structure in enlarged single duramater with syrinx cavity. Bone defect contained a cystic formation filled with cerebrospinal fluid (CSF), which was continuous with the intramedullary syrinx cavity (Figs. 3, 4a, b). The spinal cord ended at T12. There was no abnormality of the craniocervical junction such as Chiari malformations.

Discussion

Split spinal cord malformations are congenital spinal anomalies in which spinal cord is split longitudinally by a rigid or fibrous septum. Pang classified SCMs, according to number of dural tube and splitting process within spinal cord. A type 1 SCM consists of two hemicords, each contained its own dural tube and seperated by a dura-sheated rigid median septum. A type 2 SCM consists of two hemicords housed in a single dural tube seperated by a non-rigid, fibrous median septum [8].

According to Dachling Pang, SCMs form in the third week of gestation because of lack of obliteration of the primitive neurenteric canal and the communication between the amnion and yolk sac, and instead, a secondary neurenteric

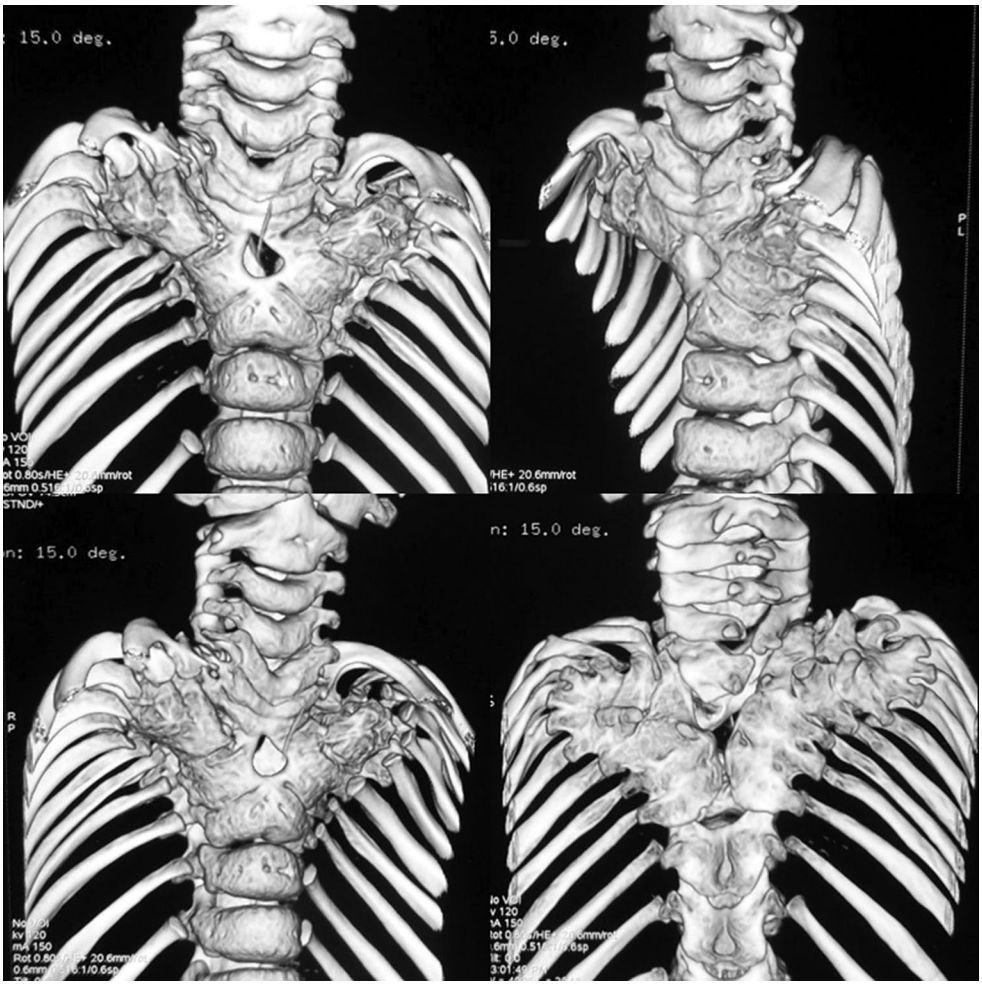

Fig. 2 Volume rendering images of Computerized Tomography examination, anterior and posterior projections reveals spinal column duplication between C5 and T9 levels. Both of the duplicated spine column was steered to laterally and showed fusion abnormalities. There is an anteriorly located bone defect at duplication level

canal forms that causes the split. The split can be limited to the cord and dura, or can extend to the vertebral bodies [8].

Duplication of spine with marked separation of bony elements is a rare malformation, and there are limited number of cases published as case reports in the literature. According to classification by Pang et al. [9] SD is considered as a severe form of Type 1 split cord malformations. However, Pang et al. have not reported any patients with such extensive duplication of the bony elements. Therefore, some authors define SD as unclassified today.

According to our best knowledge, it is the longest follow up time in the literature. Previously reported cases were incidentally identified cases in screening studies or at different outpatient services without follow up periods.

Yigit et al. reported an interesting SD case with hemilipomyelomeningocele. In the presented case, the thecal sac was split to two separate thecal sacs below T8 level; the right thecal sac which included spinal cord and the left sided thecal sac which had no visible neural content, conjugated again at L4–L5 level [6]. Also in our case, spinal cord was a single structure in enlarged single duramater similar to Yigit's case without split of spinal cord. It' s controversial to classify both cases as type 1 SCM, maybe it is logic to classify them as unclassified.

Spine duplication may have different clinical presentations that range from mild to severe and symptoms may differ. Cebesoy reported an adult patient diagnosed during an evaluation for other systemic pathologies. The condition was

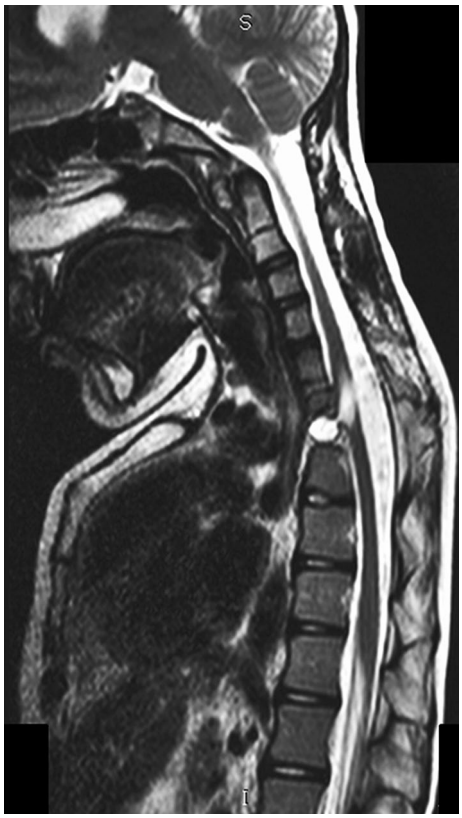

Fig. 3 Sagittal T2 weighted Magnetic Resonance Image shows duplicated spine and a bony defect. Medulla spinalis is unicord structure in enlarged single durameter. Anterior epidural area was in communication with posterior mediastinum through a bone defect at the center of dublication. This defect contained a cystic formation filled with CSF which showed continuity with intramedullar syrinx cavity

not diagnosed until adulthood [2]. Incesu et al. presented an asymptomatic adolescent girl who had been diagnosed with spine duplication during an evaluation for back pain [3]. Goldberg presented a lumbar spine duplication case suspected as adolescent idiopathic scoliosis during a school screening program [4]. The aforementioned cases were all thoracolumbar-lumbar spine duplications without any neurologic deficits. The remaining cases of spine duplication were symptomatic with other system anomalies [9–11] and some of the cases could be regarded as a form of caudal duplication [12–14]. The last mentioned cases were severely affected by multisystem dysfunction and neurologic deterioration. Our patient presented with short body stature and further evaluation revealed cervicothoracic spine duplication. He was followed up for 10 years without any neurologic deterioration and functional limitation.

Here, our case did not show additional system abnormality except mild obstructive pulmonary disease without clinical significance. Pulmonary function test (PFT) give the most useful information about conditions affecting lungs but patient cooperation to the test is very difficult especially in young population. At this point two-dimensional measurements (pelvic inlet width, T1–T12 height, T1–S1 height and coronal chest width) could give information but their value on predicting lung capacity is controversial [15]. We could not measure T1–T12 height because of the C5–T9 duplication levels, fortunately he could performed PFT exactly.

Pang et al. concluded that SCMs were tethering lesions of the spinal cord, likely to cause neurologic deficit, and should be treated. The suggested surgical treatment included the release of the tethered hemicords and removal of fibroosseous sleeves and spurs which might have transfixed the split cord [16]. Large series on SCM proposed that all patients with SCM should have prophylactic surgery, even if they are asymptomatic [17]. However, none of the asymptomatic cases reported in the literature underwent surgical intervention [2–7]. The patient presented here did not show any neurological compromise during the 10-year follow-up. Since he is neurological normal, we will continue to observe this patient. From first diagnosis at 3 years up to now the patient has not shown coronal decompansation in terms of shoulder asymmetry and pelvic tilt. He is attending daily sportive activities without difficulty.

Conclusion

To the best of our knowledge, this patient is the first case of cervicothoracic spine duplication with a 10-year follow-up; unlike other reports, he presented with an enlarged dura matter and a single spinal cord without any split anomaly. This case presents a distinct form of SCM which shows non-duplicated dural tube and cervicothoracic duplication level without neurological deficitis. Treatment of SD based on removal of fibrous/osseous intramedullar splitting process. As previously reported cases without neurologic deficits,

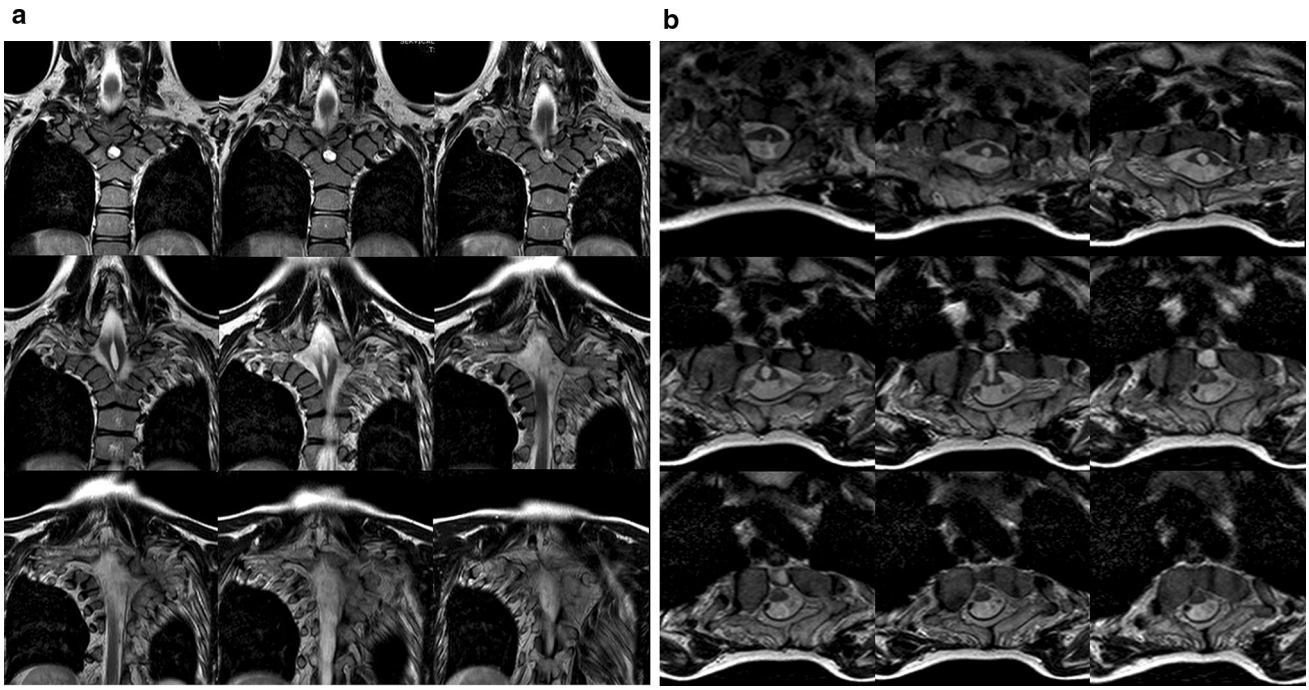

Fig. 4 **a,b** Coronal (**a**) and axial (**b**) consecutive T2 weighted MR images demonstrate medulla spinalis as unicord structure in enlarged single durameter and cyst formation in a bone defect at duplication level showed communication with intramedullar syrinx cavity

cervicothoracic duplication without cord abnormality also could be followed up conservatively.

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Compliance with ethical standards

Conflict of interest None of the authors has any potential conflict of interest for this paper.

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