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CHINESE SECTION

Unusual association of intraspinal extramedullary teratoma with congenital scoliosis in an elderly adult: case report and literature review

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Abstract
Introduction Intraspinal teratomas associated with congenital scoliosis are extremely rare, especially in an elderly adult.
Materials and methods We report the seventh case of intraspinal extramedullary teratoma coexisting with congenital scoliosis in a patient older than 50 years, possibly the oldest patient documented in literature. A 56-year-old male suffered from low back pain that increased with calf numbness and foot weakness. Conventional radiography showed a congenital scoliosis due to incomplete segmentation of the L2 and L4 vertebras, and magnetic resonance images revealed a heterogeneous intraspinal extramedullary mass located at L4-S1.
Results The tumor was totally removed, and was confirmed as a mature teratoma on biopsy. The patient remains asymptomatic at 34-month follow-up.
Conclusions Rare intraspinal teratoma should be included in the differential diagnosis of intraspinal mass, especially in patient with congenital scoliosis. Patient with mature teratoma may survive with out any symptoms in the long term. Progressing neurological deficit is a main indication for surgery. Excellent clinical outcomes could be achieved by surgical resection and dural sac decompression.

Keywords Teratoma · Congenital scoliosis · Surgery

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Introduction

Spinal teratomas associated with congenital scoliosis are very uncommon [1], which have been previously reported with the majority occurring in the pediatric population [2–7]. Intraspinal teratoma coexisting with congenital scoliosis in the adult is considered to be an extremely rare entity. We present an unusual case of lumbar teratoma in a 56-year-old patient with congenital scoliosis, possibly the oldest patient recorded in the literature.

Case report

A 56-year-old man was admitted to our hospital with a 1-year history of low back pain and radiating pain in both posterior calves. In the last 2 months, his symptoms progressed gradually to calf numbness and foot weakness. He had neither history of trauma nor any other spinal procedures. Neurological examination revealed weakness of the both extensor hallucis longus. Straight leg raise was positive at 40° bilaterally. A sensory deficit involved left anterior tibial and dorsalis pedis. No abnormal hairy patch or dimple was found in the skin over the lumbosacral region. The X-ray of the lumbar spine demonstrated a left curve of 42° (Fig. 1a), and a kyphosis of 60° (Fig. 1b). Incomplete segmentation of the L2 and L4 vertebras was noted on plain radiographs. Magnetic resonance (MR) images indicated a heterogeneous intraspinal extramedullary mass located at L4-S1 levels. The intraspinal lesion showed inhomogeneous high and low signal intensity on T1WI and T2WI (Fig. 2a, b). MR images of the whole spine displayed no cord tether, syrinx or other dysraphic abnormality.

As the patient denied the surgical correction of scoliosis, the surgeons proceeded to remove the tumor without

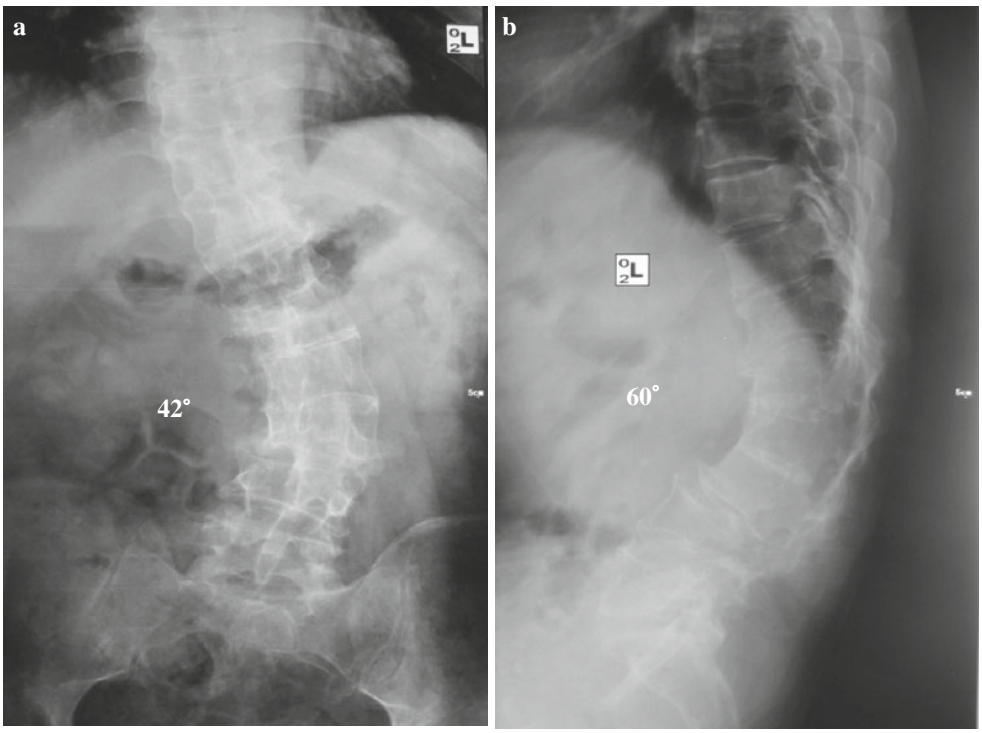

Fig. 1 A 56-year-old male with a 42° left lumbar scoliosis and a 60° kyphosis due to unilateral unsegmented bar

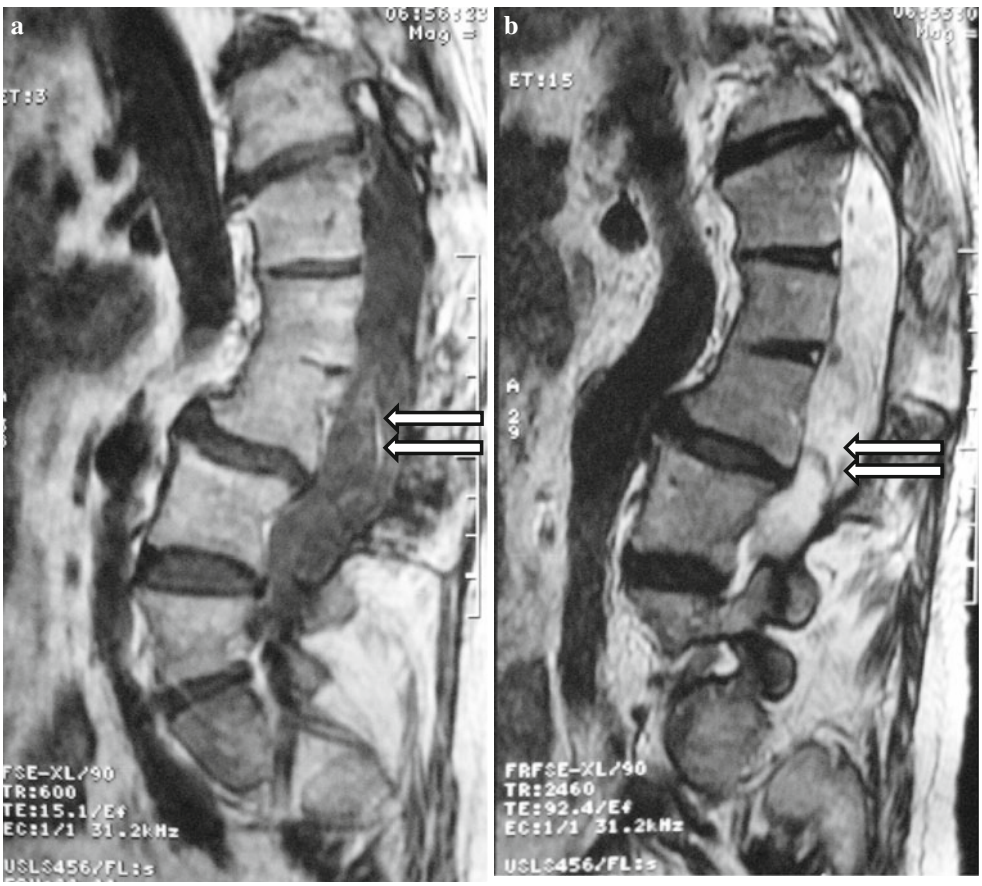

Fig. 2 MR images revealing a T1-hyperintense (**a**) and T2-hypointense (**b**) intraspinal mass at L4-S1 levels

Fig. 3 Postoperative photographs demonstrating the tumor with hair shaft, cartilage, fibrous tissue and adipose tissue

treating spinal deformity. During surgery, a L4-L5 total laminectomy and S1 partial laminectomy were performed. After the dura was opened, the cystic mass could be visualized without any firm attachment posteriorly to the dura. Consequently the tumor was totally removed, which contained brown hair and cartilaginous tissue (Fig. 3). Histological examination demonstrated a mature tridermal teratoma with fully differentiated components including stratified squamous epithelium, neural tissue, enteric glandular cells and cartilage (Fig. 4). The patient had complete resolution of their neurological symptoms within 3 months postoperatively. There was no obvious progression of the kyphoscoliosis at 34-month follow-up.

Discussion

The incidence of intraspinal teratomas is low, and these tumors account for approximately 0.5–2.2 % of all intraspinal tumors [1, 8]. Intraspinal teratomas have been commonly reported in relation to abnormal neural tube including diastematomyelia, myelomeningocele and tethered cord syndrome [9, 10]. The occurrence of intraspinal teratomas associated with congenital scoliosis is rare, and is more common among infants and adolescents than among adults [8]. Only six studies on spinal teratomas coexisting with congenital scoliosis have been previously reported [2–7] (Table 1). To our knowledge, we present the seventh case of a mature intraspinal teratoma associated with congenital scoliosis in a 56-year-old male who may be the oldest patient documented in the literature.

The etiology of spinal teratoma has not been fully understood. Several hypotheses have been proposed,

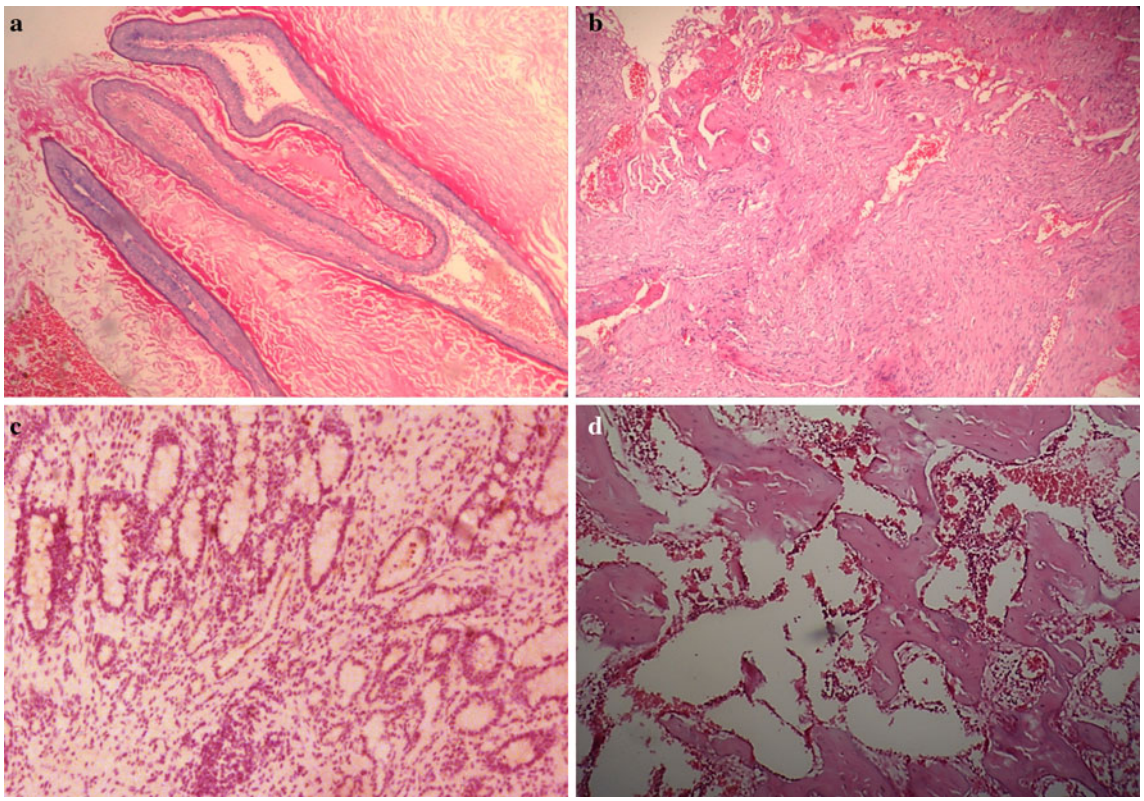

Fig. 4 Photomicrographs of the tumor specimen showing tissues derived from all three germ layers: **a** stratified squamous epithelium, **b** neural tissue, **c** enteric glandular cells, **d** cartilage, hematoxylin and eosin, original magnification (×100)

Table 1 Cases of intraspinal teratomas associated with congenital scoliosis

	Case author	Year	Age	Gender	Location of teratoma	Spinal anomalies	Treatment of teratoma	Outcome
1	Cameron [2]	1957	NB	M	Extramedullary, lumbar	Congenital kyphoscoliosis	None	Died
2	McMaster [3]	1984	4 y	F	Intradural, thoracolumbar	Scoliosis, left lower limb hypoplasia, thoracolumbar cutaneous lipoma	Surgery (extent not specified)	Improved at 8 y
3	Koen et al. [4]	1998	14 m	M	Intradural, thoracic	T5–6 Hemivertebra, absent 6th rib, scoliosis	Gross total resection	NS
4	Hader et al. [5]	1999	16 y	F	Intramedullary, thoracic	T7–9 Hemivertebra, scoliosis	Subtotal resection	Symptom free at 2 m
5	Jarmundowicz et al. [6]	2004	13 y	F	Intraspinal, lumbar	Split cord malformation, scoliosis	Subtotal resection	NS
6	Suri et al. [7]	2006	8 y	M	Intraspinal, thoracic	Hole in T4 vertebral body, butterfly vertebra, scoliosis	Gross total resection	Symptom free at 1 y
7	Present case	2008	56 y	M	Intraspinal, lumbar	L3–4 Incomplete segmentation, scoliosis	Gross total resection	Symptom free at 3 m

NB newborn, *NS* not specified, *m* months, *y* years

including the dysembryogenic theory and misplaced germ cell theory [11]. Our case is considered to support misplaced germ cell theories because, it is an instance of adult teratoma without dysraphism. Additionally, several authors reported that intraspinal teratoma can occur in adults after trauma or surgical interventions [12, 13]. However, the patient in the present study had neither history of trauma nor any other spinal procedures.

Preoperative diagnosis of spinal teratoma is not easy. The role of plain radiography is limited to detect the vertebral abnormalities including hemi vertebrae, butterfly vertebrae and block vertebrae [14]. In the current case, X-ray images revealed incomplete segmentation of the L2 and L4 vertebras leading to lumbar kyphoscoliosis. Furthermore, MR scans may provide more information about intraspinal lesions. Heterogeneous signal intensity on MR images indicated a solid and cystic composition of the tumor, which would be helpful for diagnosis of teratomas in the early stage [11, 13]. Correspondingly, teratomas in histopathology are composed of the heterogeneous remnants of all three germ cell layers, and are classified as mature, immature or malignant type [10]. Mature teratomas, represented by our case, contained fully differentiated tissue elements such as cartilage, squamous epithelial cells, glands, mucosal tissue and neural elements.

As posterior correction and fusion were currently widely used for the treatment of congenital scoliosis [15], two-stage surgical treatment was recommended in our preoperative procedures including tumor resection and scoliosis correction. However, the patient in this case denied the scoliosis correction, because of socio-economic restriction. Fortunately, his neurological symptoms had been completely resolved within 3 months after tumor removal. Nonetheless, we strongly recommend that surgical procedures for intraspinal teratoma coexisting with congenital scoliosis be performed in two stages, and the intraspinal anomalies be treated surgically before management of scoliosis.

Conclusion

Intraspinal extramedullary teratoma associated with congenital scoliosis is extremely uncommon, especially in patients over 50 years. Heterogeneous signal intensity on MR images indicated a solid or cystic composition of the tumor, which would be helpful for diagnosis of teratoma in early stage. The good outcome could be achieved by surgical resection, because the clinical symptoms may be mainly attributed to dural sac compression by intraspinal teratoma.

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Conflict of interest None.

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