CASE REPORT

Fibrous dysplasia of the thoracic spine

Zong-Syun Wu\textsuperscript{a,b}, Shiuh-Lin Hwang\textsuperscript{a,b,c,*}

\textsuperscript{a} Division of Neurosurgery, Department of Surgery, Kaohsiung Medical University Hospital, Kaohsiung, Taiwan
\textsuperscript{b} Faculty of Medicine, College of Medicine, Kaohsiung Medical University, Kaohsiung, Taiwan
\textsuperscript{c} Graduate Institute of Medicine, College of Medicine, Kaohsiung Medical University, Kaohsiung, Taiwan

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Summary
Fibrous dysplasia occurs rarely in the spine, especially in the thoracic spine. According to the literature, less than 20 cases have been reported. We report in this paper a 48-year-old female patient with fibrous dysplasia of the first thoracic spine and left side of the first rib with left thoracic first root compression. She complained of severe pain of the left arm, numbness on the ulnar side of the left forearm, and weakness of the middle to little fingers. Computed tomography and magnetic resonance imaging showed the characteristic appearance of fibrous dysplasia, which includes ground-glass opacity, an expansile nature, and lytic lesions with sclerotic rims. She underwent surgical intervention with radical tumor resection combined with stabilization and fusion with a mesh graft. The pain was relieved. The thoracic first root compression signs improved after the operation. Nineteen months after the operation, there was no recurrence of the tumor. We suggest that radical tumor resection combined with stabilization and fusion with an anterior or posterior approach using a mesh graft for thoracic fibrous dysplasia can achieve definite decompression and prevent tumor recurrence.

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1. Introduction

Fibrous dysplasia (FD) is usually a benign fibro-osseous developmental anomaly caused by the replacement of the medullary component of one or several bones with fibrous connective tissue and irregular osteoid formation (malignant transformation occurs in <1% of cases).\textsuperscript{1} This disease primarily affects adolescents and young adults and accounts for 7% of benign bone tumors. Most lesions occur in the ribs or craniofacial bones, especially the maxilla.\textsuperscript{2} Fibrous dysplasia commonly presents in a monostotic form or may present in a polyostotic form (25% of people with the polyostotic form have >50% of the skeleton involved with associated fractures and skeletal deformities).\textsuperscript{2}
Fibrous dysplasia is often subcategorized based on the number of bones involved. Monostotic lesions involve only one bone, whereas polyostotic lesions involve many bones. Many asymptomatic lesions are discovered incidentally; the remainder present with the symptoms of swelling, deformity, or pain. The polyostotic form of FD has been associated with McCune–Albright syndrome and Mazabraud’s syndrome, in which skeletal abnormalities are associated with characteristic café au lait spots and endocrinal abnormalities. The etiology remains unclear.

Fibrous dysplasia has a characteristic radiographic appearance. Most patients do not require intervention, but some patients are managed surgically with curettage, bone grafting, and (in some patients) internal fixation. We report a rare case of FD of the thoracic spine and review the literature.

2. Case Report

A 47-year-old woman presented with progressive upper back and neck pain for 10 years. The pain was located in the left subscapular area with radiation to the left arm, the ulnar side of the forearm, and the middle to little fingers. [The visual analog scale (VAS) score was approximately 5–6.] The pain was aggravated by neck hyperextension. She complained of numbness with severe paresthesia on the left ulnar side of the forearm.

Physical examinations revealed weakness of the left finger flexors, finger abductors (the muscle power was 4–5), and abnormal pin-prick sensation in the left cervical 7–8 dermatome and the thoracic 1–2 dermatome. Laboratory investigations revealed no abnormal findings.

Plain radiographs of the cervical and thoracic spine demonstrated an osteolytic lesion of the left T1 and the left first rib with a pathologic compression fracture of these bones, which suggested a metastatic tumor (Fig. 1). Computed tomography (CT) of the thoracic spine showed osteolytic lesions that involved the vertebral body with ground-glass opacity, the laminae on both sides, the spinous process of T1, and the left first rib (Fig. 2). Magnetic resonance imaging (MRI) demonstrated a multiloculated lesion with septa-like structures and marginal sclerosis extending from the T1 vertebral body to the adjacent left posterior part of the first rib. This lesion showed low signal intensity on T1-weighted and T2-weighted images with mild enhancement. We therefore highly suspected FD.

A herniated C5–6 intervertebral disc with posterior osteophyte caused mild thecal sac compression and bilateral neural foraminal encroachment (Fig. 3). The differential diagnosis of the lytic bone lesions included primary neoplasm, secondary neoplasm, or an infectious process. We suggested an en bloc resection of the tumor for decompression and pathological diagnosis.

An anterior approach with mesh graft fusion/fixation was planned to prevent postoperative instability. We performed C5–6 disectomy and replaced it with an artificial disc.

During the operation, the bone tumor was white, elastic, and did not exhibit the expected hypervascularity. Surgical treatment consisted of C5–6 disectomy with artificial cage fusion [Bryan® (Medtronic Spinal and Biologics, Memphis, Tennessee)], en bloc tumor resection, T1 corpectomy with C7–T2 mesh and plate fixation. Pathological examination of the surgical specimen revealed curvilinear trabeculae of woven bone arising in the background of fibrous tissue (Fig. 4). There was no evidence of malignancy in the specimen. We also performed bone scintigraphy, which did not show disease at any other site.
The patient’s postoperative course was uneventful. Fig. 5 shows the patient’s postoperative radiography. By 6 months postoperatively, the patient’s pain score fell from 5–6 to 1–2 (based on the VAS) and the numbness and weakness improved (the muscle power increased from 4 to 5). Nineteen months postoperatively, a follow-up MRI showed no recurrence (Fig. 6).

3. Discussion

Fibrous dysplasia represents approximately 7% of all benign tumor-like bone lesions. However, the spine is affected in only 2.5% of patients. Fibrous dysplasia of the spine very rarely exists without being present elsewhere in the body. Spinal involvement occurs mostly in the polyostotic form of FD; it is unusual for it to occur in the monostotic form, especially in the thoracic spine. A literature review of the years 1961–2013 revealed only 16 cases of monostotic FD of the thoracic spine.

Fibrous dysplasia has a characteristic radiographic appearance. Plain radiographs, CT scans, and magnetic resonance images show the presentation of an eccentric lesion with intact cortex bone and marginal sclerosis in vertebral bodies without involving the vertebral appendix and extraosseous soft tissue. The radiographic hallmark of monostotic FD involving the spine is a well-defined multiloculated expansion of the medullary cavity with a ground glass appearance and variable degrees of marginal sclerosis. The CT image and MRI are superior to plain radiographs in defining the extent of involvement of the spine. The MRI may be helpful in demonstrating the characteristics of fibrous lesions. Fibrous dysplasia usually shows low signal intensity on T1-weighted images, heterogeneous mixed signal intensity on T2-weighted images, and mild enhancement after the administration of a contrast material. A sclerotic “rind” that encapsulates the expansile bone lesion is always present in typical cases of FD. Spinal cord compression can be well demonstrated on MRI.

There is no standard treatment for FD. Treatment includes pharmacological therapy and surgical intervention. Oba et al. managed FD that involved only the 10th vertebra with tumor resection, which was followed by bone grafting with hydroxyapatite material. Arazi et al. treated FD in the sixth vertebra with the transthoracic approach and used a costal graft and interbody fusion after en bloc resection. Tezer et al. cite the work of Przybyski et al who performed a posterior fusion and instrumentation combined with an anterior strut graft and 360° spinal fusion, after resecting the tumor in FD located in the thorax, which showed kyphotic deformation and neurological progression. Chapurlat et al. cite the work of Nabarro and Giblin who treated thoracic FD with tumor resection with anterior and posterior approaches in combination with arthrodesis.

In treating FD, stabilization is as important as the excision of the lesion. A very important aim in treating spinal tumors is to perform one radical surgery and avoid repeated surgeries. For this reason, the lesion should be resected as extensively as possible. In large resection sites, stabilization
and fusion should be performed cautiously to avoid problems related to pseudoarthrosis. With the goal of 360° stabilization and fusion with an anterior or posterior approach, using a strut or mesh graft is an effective treatment in the polyostotic form of FD involving successive vertebrae. The prognosis is generally good, although poor outcomes are more frequent in younger patients and in patients with polyostotic forms of the disease. The risk of malignant transformation is low.

An alternative treatment for FD is bisphosphonate drugs. Pamidronate is the most effective bisphosphonate and is used at a dosage of 180 mg every 6 months. Prospective studies have been conducted in increasingly large numbers of patients. However, there are no reports of patients with monostotic FD of the spine who were treated with bisphosphonates. In addition to the bisphosphonates, calcium and vitamin D supplementation may benefit patients with FD.

As for our patient, we used nonsteroidal an anti-inflammatory drug (NSAID) and supplementation with calcium and vitamin D for approximately 6 months after the operation. No specific complaints were noted.

Fibrous dysplasia of the thoracic spine is rare, especially in the thoracic spine. Our patient had FD limited to the thoracic spine that mimicked a neoplastic or infectious process. Fibrous dysplasia should be included in the differential diagnosis of patients with lytic bone lesions. A combination of medical and surgical management can lead to excellent outcomes. As in the present patient, although located in the thoracic region, cystic lesions in segments of the whole spine that are not successive or adjacent to each other should be evaluated for the possibility of FD with detailed radiographical studies. We also suggest radical tumor resection combined with stabilization and fusion with an anterior or posterior approach using a mesh graft for thoracic FD to achieve definite decompression and prevent recurrence.

References


