Formosan Journal of Surgery (2014) 47, 122–126

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.e-fjs.com

CASE REPORT

Fibrous dysplasia of the thoracic spine

Zong-Syun Wu [a,b], Shiuh-Lin Hwang [a,b,c,*]

[a] *Division of Neurosurgery, Department of Surgery, Kaohsiung Medical University Hospital, Kaohsiung, Taiwan*
[b] *Faculty of Medicine, College of Medicine, Kaohsiung Medical University, Kaohsiung, Taiwan*
[c] *Graduate Institute of Medicine, College of Medicine, Kaohsiung Medical University, Kaohsiung, Taiwan*

Received 3 September 2013; received in revised form 27 November 2013; accepted 15 January 2014
Available online 15 May 2014

KEYWORDS
Fibrous dysplasia;
fusion;
no recurrence;
radical resection;
thoracic spine

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.

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).[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.[1] 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).[2]

Conflicts of interest: All authors declare no conflicts of interest.

* Corresponding author. Division of Neurosurgery, Department of Surgery, Kaohsiung Medical University Hospital, Kaohsiung Medical University, 100 Tzyou First Road, Kaohsiung 80708, Taiwan.
E-mail address: nsdoctor@yam.com (S.-L. Hwang).

http://dx.doi.org/10.1016/j.fjs.2014.01.001

Fibrous dysplasia is often subcategorized based on the number of bones involved. Monostotic lesions involve only one bone, whereas polyostotic lesions involve many bones.[3] Many asymptomatic lesions are discovered incidentally; the remainder present with the symptoms of swelling, deformity, or pain.[4] 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.[1] 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.[3] 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—), 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 diskectomy 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 diskectomy 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.

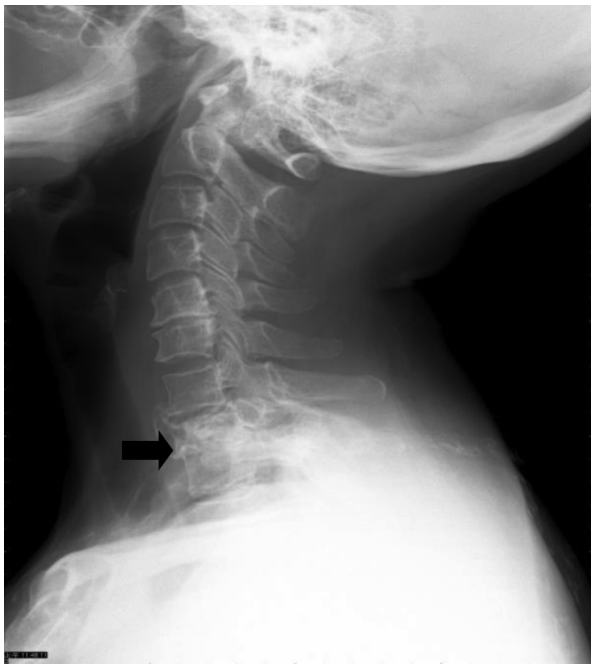

Figure 1 C-spine X-ray image (lateral view) shows an osteolytic lesion of T1 with a pathological compression fracture (black arrow).

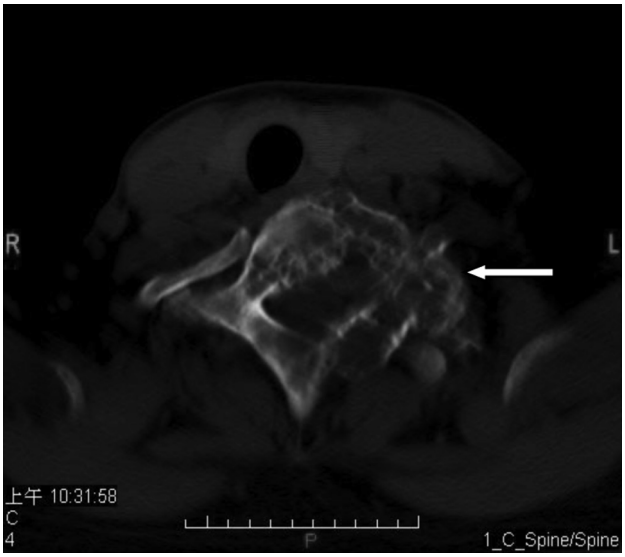

Figure 2 Axial view computed tomography image of the cervical spine in the bone window setting shows an expansile osteolyic lesion (primarily at T1) that involves the left side of the vertebral body, pedicle, lamina, and transverse process. It extends to the left side of the adjacent first rib.