Case Report

Monostotic fibrous dysplasia of the thoracic spine: A case report

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Abstract.
BACKGROUND AND OBJECTIVE: Fibrous dysplasia (FD) is a benign bone lesion manifested by local pain, swelling and deformity change. We report a case of monostotic fibrous dysplasia of the first thoracic vertebrae that treated by radical removal and reconstruction.

CASE REPORT: A 29-year-old man with monostotic fibrous dysplasia of the first thoracic vertebrae was admitted to our department because of persistent, dull back pain for 3 months. Radical removal of the first thoracic vertebrae and reconstruction were performed in a combined posterior-anterior approach. This patient experienced complete pain relief without any complication.

CONCLUSION: This report presents a rare case of monostotic fibrous dysplasia of the first thoracic vertebrae, with symptoms of chronic back pain that was successfully treated with radical excision and reconstruction, providing a good option to the patient.

Keywords: Monostotic, fibrous dysplasia, spine, surgical treatment

1. Introduction

Fibrous dysplasia (FD) is an uncommon bone formation disorder characterized by replacement of medullary bone component with immature fibro-osseous tissue [1]. It accounts for approximately 7% of benign bone tumors and 2.5% of all bone lesions [2]. The exact etiology remains unknown. It is well recognized that mutation of the Gs alpha gene on chromosome 20 leads to an increase in the generation of cyclic adenosine monophosphate in mutated cells, which influences the function of osteoblastic cells resulting in osteogenic process abnormalities [3–5]. According to the numbers of bone involved, it could be divided into monostotic fibrous dysplasia (MFD) and polyostotic fibrous dysplasia (PFD). Lesions of FD are most commonly found in the ribs, followed by the femur, tibia, and craniofacial bones. Although MFD represents 70% of this lesion, involvement of axial bone is rare and it is frequently seen in PFD. McCune Albright syndrome is a severe form of PFD that couples with cutaneous changes (café-au-lait spot) and endocrine abnormalities.

We present a case of MFD of the first thoracic vertebrae that was managed with complete excision and reconstruction in a combined posterior and anterior approach.

2. Case report

A 29-year-old male patient was admitted to our department because of persistent, dull back pain for 3 months. The pain localized to the first thoracic vertebrae area without radiation. The VAS was 5 on the day of admission. The pain was aggravated by motion, especially backward movement and wasn’t relieved by rest. There was no history of trauma recently. His past medical history and family history were unremarkable.
Fibrous dysplasia (FD) is a benign bone lesion that was first described by Lichtenstein in 1938 [1]. It is characterized by the replacement of bone marrow with poorly organized spicules of immature bone. There is no gender predomiance. It may affect a single bone (MFD) or multiple bones (PFD). Vertebral involvement in MFD is exceedingly rare. The majority of cases involving spine were discovered incidentally on radiography since most of the patients were asymptomatic, whereas symptomatic cases may present as local pain, swelling and deformity change.

Radiological data plays an important role in diagnosing this disease. The most characteristic features are multiloculated expansion with sclerotic margin and “ground-glass” appearance change on X-ray. CT and MRI are superior to plain film in revealing information about the extent of spinal involvement, which is critical in the management strategy selection. Histological examination is the gold standard for the final diagnosis. CT-guided percutaneous biopsy is recommended as a safe and effective technique in the evaluation of lesion with reported accuracy rate about 90% [6,7]. Wu FL reported that nature of lesion, diameter of biopsy trocar and puncture site are three most important factors in determining the accuracy of biopsy [8]. However, his study showed that the rate of correct preoperative pathological diagnosis by computed tomography-guided biopsy was low for patients with suspected spinal FD. In our case, what we got from our sampling was yellowish fluid. Percutaneous biopsy was performed by medical doctor instead of radiologist in our hospital. The doctor’s experience may influence the result of correct rate. Other possible reasons for our failure in sampling are the puncture site and the diameter of trocar. To obtain a correct result, a representative specimen must be obtained with all components, especially for sclerotic lesions. We used an 8G biopsy needle that was not suitable to get more amount of samples. However, specimens were obtained during the operation and the final histological result revealed FD changes. So, We recommend that open biopsy be performed when spinal FD is suspected.

To date the treatment strategy still remains controversial because of the rarity of this disorder and lack of systematic analysis. To the best of our knowledge, the location, size of the lesion and patients’ symptom are the most crucial factors in determining treating method. Conservative management includes simple observation, analgesic medication and periodic follow-up examination. However, once conservative management fails, more aggressive management should be taken into consideration. It is well recognized, that FD is caused by activating an autosomal mutation of the Gs alpha subunit of protein G resulting in an increased c-AMP concentration and thus in retarding of osteoblast differentiation with abnormal bone formation. There is also an increase in interleukin-6 levels expressing bone resorption, which is the rationale for administering bisphosphonates in the management of these patients. Some studies demonstrated bisphosphonates as a promising drug, had successfully relieved FD associated pain. Study by Chapurlat showed that pamidronate therapy using 180-mg infusion every 6 months was able to reduce bone pain.
and bone turnover in most patients with FD, while about half of the patients also display radiological improvement [9]. Jayaraman M’s case report showed that oral alendronate significantly reduced bone pain in a PFD patient and the patient’s plain radiography showed reduction in lesion size [10]. Percutaneous vertebroplasty (PVP) and balloon kyphoplasty (PKP) have been widely and successfully used in pain relief and early immobilization in compression fracture patients [11–13]. Michinari reported that hydroxyapatite was sufficient to replace bone defect owing to its chemical similarity to bone [14]. Some investigators reported that simple curettage of lesion and bone graft can provide a good outcome [15,16], but studies also
showed the possibility of recurrence of FD. Osteosarcomatous, fibrosarcomatous and chondrosarcomatous are three types of malignant transformation of fibrous dysplasia [17]. Rodenberg J reported a case of sarcomatous transformation of fibrous dysplasia, which affected the spine, costae and hemipelvis [18]. Since the less favorable results after incomplete resection, some clinicians advocated the radical removal of the affected regions to prevent recurrence or malignant transformation [19–21]. Rosenblum B reported a fibrous dysplasia case affecting the first thoracic vertebral body. The patient underwent complete resection of the mass and anterior fusion with an autogenous bone graft that gave us good evidences of this method [2]. However, it is well recognized that FD is a benign bone formation disorder without destructive capacity and the malignant transformation is rare. So the possibility of malignant transformation is not an indication for massive surgery. Actually, as pain is the common occurrence in FD, a step-wise approach to its management, starting with NSAID is recommended in symptomatic patients. Meanwhile, we strongly encourage periodic follow-up of our patient after treatment.

In our report, immobilization and analgesic medication didn’t help in relieving patient’s pain that bothered his daily life. The patient was at high risk of pathological fracture because of extensive destruction of the first thoracic vertebrae according to radiological findings. Vertebroplasty was not recommended because of minor-fracture of posterior cortex and possibility of cement leakage. Sclerotic rims will also limit diffusion of cement. We performed a total resection of the first thoracic vertebrae using a combined posterior and anterior approach. The posterior approach was performed first to achieve rapid decompression and immediate stabilization. After that we removed the vertebral body of the first thoracic vertebrae and reconstructed it with titanium mesh and autogenous iliac crest graft. Anterior plate was used to maintain the stability of titanium mesh. This patient recovered soon and VAS was 0–1 when he was discharged. Currently, the patient was back to full activities. 1-year follow up imaging studies were satisfactory (Fig. D1, 2).

4. Conclusion

This report presents a rare case of monostotic fibrous dysplasia involving the first thoracic vertebrae, with symptoms of chronic back pain that was successfully treated with radical excision and reconstruction, providing a good option to the patient.

Conflict of interest

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References


