

## Monostotic Fibrous Dysplasia of the Spine: Report of a Case Involving the Cervical Transverse Process and Review of the Literature

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**Abstract:** Fibrous dysplasia (FD) is a benign bone lesion; it is a disorder of bone development in which cancellous bone is replaced by fibrous tissue. Although fibrous dysplasia can involve any bone in the body, vertebral involvement is quite rare. We report a case of monostotic fibrous dysplasia of the sixth cervical vertebrae transverse process that treated by gross resection. A 32-year-old man complained of persistent, dull cervical pain for 2 months. Pre-operative CT scan demonstrated a neoplasm located at the cervical transverse process and expanded into surrounding soft tissues. After gross resection, complete pain relief without any complication was experienced.

**Key-words:** Fibrous dysplasia; cervical spine; surgical treatment.

### INTRODUCTION

Fibrous dysplasia (FD) is a benign bone lesion; it is a disorder of bone development in which cancellous bone is replaced with fibrous tissue, resulting in expansion of the bone, manifested by local pain, swelling and deformity change [1]. According to the numbers of bone involved, it could be divided into monostotic fibrous dysplasia (MFD) and polyostotic fibrous dysplasia (PFD). Although MFD represents 70% of this lesion, involvement of axial bone is rare, only 2% to 4.7% of the MFD lesions may involve the mobile spine (above the sacrum) [2]. The McCune-Albright syndrome is a severe form of PFD with endocrinopathy, which presents with the classic triad of FD, café-au-lait skin pigmentation, and endocrine dysfunction (commonly precocious puberty in the pediatric population) [3, 4].

Its treatment strategies are still controversial. The treatment strategies reported thus far vary widely and include biopsy and/or observation, surgical resection, and vertebral augmentation procedures (vertebroplasty [VP]/balloon kyphoplasty [KP])[5]. Good outcomes have been reported in almost all cases of monostotic fibrous dysplasia of the spine in the literature regardless of the therapeutic intervention used, and no factors favored a particular approach to treatment [6-8]. Thus, it seems prudent to avoid overly aggressive treatment.

We report a case of monostotic fibrous dysplasia of the sixth cervical vertebrae transverse process that treated by gross resection with satisfied clinical outcome.

### CASE REPORT

#### History

A 32-year-old man with history of 2 months persistent, dull cervical pain admitted in our hospital. He was diagnosed as cervical muscle strain at local clinic and treated with analgesics. However, the pain persisted, and he was referred to our outpatient clinic. The pain localized to the right cervical area without radiation. The VAS was 5 on the day of admission. The

pain was aggravated by motion, especially right lateral bending movement and was not relieved by rest. There was no history of trauma recently. Laboratory examinations consisted of complete blood count, electrolytes, liver, renal function tests, and urinalysis. They were all normal, except 10% evaluation of C-reaction protein level. His past medical history and family history were unremarkable. During hospital admittance immobilization and analgesic medication didn't help in relieving patient's pain which bothered his daily life.

#### Examinations

At neurologic examination neck movements were painful and restricted. Physical examination revealed normal motor and sensory function below neck of both side. The manual muscle strength test demonstrated 5/5 strength on the left and 5/5 on the right. Increase of deep tendon reflex was not noted. Babinski sign was negative bilaterally. Muscle tonus of all extremities was not increased. There was no remarkable X-ray findings except some soft tissues thickening in front of vertebral bodies (Fig.1). Computed tomography (CT) revealed an expansile lesion involving the right transverse process of the sixth cervical vertebra. A noncontrasted CT scan showed the

classic “ground glass” appearance of fibrous dysplasia, reflecting random woven bone formation; Meanwhile, The lesion was lytic with spiculate contours, with the cortical bone partially interrupted and associated with soft tissues irritation (Fig.2). Magnetic resonance imaging (MRI) confirmed the presence of a lesion in the right transverse process of C6, without involvement of the spinal cord. The lesion had a heterogeneous signal on T1-weighted images and intermediate signal on T2-weighted images, but was predominantly hypointense. Intravenous infusion of gadolinium increased the intensity of the signal of the lesion harmoniously (Fig.3).

### Surgical treatment

The differential diagnosis included aneurysmal bone cyst, osteoblastoma, giant cell tumor, eosinophilic granuloma, and chronic infection. As the neoplasm located nearby vital structures and considerable negative rate of percutaneous needle biopsy, we believed that intra-operative frozen biopsy was more suitable. As the neck pain persisted, we performed a cervical anterior approach surgery. During operation, a round shape cortical osseous tissue was exposed under cervical deep muscles. The osseous shell of the neoplasm was filled by a lot of meat like soft tissues. The frozen biopsy examination revealed fibrous tissue

within medullary bone with sparse, small irregularly shaped bony trabeculae embedded in the fibrous stroma, typical of FD. As good outcomes have been reported in almost all cases of monostotic fibrous dysplasia of the spine in the literature, regardless of the therapeutic intervention used, and no factors favored a particular approach to treatment. Thus, a gross resection was performed to avoid overly aggressive treatment.

### Pathological findings and Postoperative course

Histopathological examination confirmed the lesion to be fibrous dysplasia. The patient recovered completely without complications. After an uncomplicated hospital course, he was discharged home 1 weeks later. After follow-up for 3 months, there was no recurrence of neck pain, neurologic examination was normal, the VAS was 0. Now this patient is completely symptom free.

The design and performance of this study conformed to ethical standards of Helsinki Declaration and our national legislation. It was approved by Medical Ethical Committee of our institution. The patient was enquired whether or not willing to take part in a scientific research and informed consent forms were signed by himself.



Fig-1: Pre-operative X-ray pictures

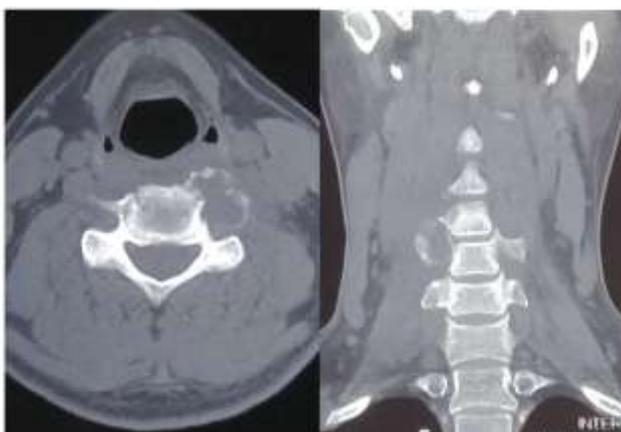


Fig-2: Pre-operative CT scans



**Fig-3: Pre-operative MR images**

## DISCUSSION

Fibrous dysplasia (FD) is a benign bone lesion that was first described by Lichtenstein in 1938 [1]. Fibrous dysplasia is caused by the replacement of normal cancellous bone in the medullary canal with immature fibro osseous tissue as a result of the mutation of GNAS1 gene. Although fibrous dysplasia presents clinically in adolescents and adults, it may be seen at any age. There is no gender predominance. It accounts for 7% [2]. Of all benign bone tumors. It may affect a single bone (monostotic) or multiple bones (polyostotic). Monostotic form is more frequent (75–80%). Polyostotic form may present with McCune–Albright (polyostotic fibrous dysplasia-caf-au-lait spots-endocrine dysfunction) and with Mazabraud syndrome (polyostotic fibrous dysplasia soft tissue mycosomes) [3, 4]. 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 pain, swelling and deformity formation. Fibrous dysplasia may cause pathological fractures. Malignant transformation occurs with a frequency of 0.5% in monostotic form but it may rise to 4% in McCune–Albright syndrome [9, 10].

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 [11]. However, the lesion was almost undetectable in X-ray films by the transverse process location. CT is superior

to plain film in revealing information about the extent of involvement, which is critical in the management strategy selection. Classically three types of computed tomography (CT) images are described: ground glass (56%), homogeneous dense (sclerotic) (23%), and radiolucent (cystic) (21%). These findings are characteristic of fibrous dysplasia [12]; however, magnetic resonance imaging (MRI) findings of FD are variable and may not be as characteristic as CT findings. There is intermediate or low signal intensity in T1-weighted MRI sequences, and intermediate or high signal intensity in T2-weighted MRI sequences. Contrast enhancement is generally heterogeneous with variable intensity [13, 14].

Histological examination is the gold standard for the diagnosis. The histologic features of monostotic fibrous dysplasia of the spine consisting of narrow, curved, and misshapen, woven bony trabeculae, supported in a background of fibroblastic tissue of variable cellularity. This woven bone never becomes transformed to lamellar bone, suggesting that the process of bone formation is arrested at an early stage [15]. The study of Wu FL showed that the rate of correct pre-operative pathological diagnosis by computed tomography guided biopsy was low for patients with suspected spinal FD [16]. Thus, we recommend open biopsy when spinal FD is suspected.

The reported treatment of monostotic fibrous dysplasia in the spine varies widely, from biopsy and observation to radical surgical resection. The location,

size of the lesion and patients' symptoms are the most crucial factors in determining treating method. Some patients received incisional biopsy experienced complete resolution of the symptoms and remained asymptomatic. Non-operative management after biopsy confirmation of the diagnosis may be considered a viable option in the absence of neurologic findings or structural compromise [17]. Percutaneous vertebroplasty (PVP) and balloon kyphoplasty (PKP) have been successfully used in pain relief and early immobilization in FD patients [18]. Furthermore, some clinicians advocated the radical removal of the affected regions to prevent recurrence or malignant transformation [19], however, it is well recognized that FD is a benign bone formation disorder without destructive capacity and the malignant transformation is rare, with only one such report in the literature [20]. So the possibility of malignant transformation is not an indication for massive surgery. On the other hand, good outcomes have been reported in almost all cases of monostotic fibrous dysplasia of the spine in the literature regardless of the therapeutic intervention used. All patients had a favorable outcome, and no factors favored a particular approach to treatment. Thus, it is a good choice to avoid overly aggressive treatment. In our report, immobilization and analgesic medication didn't help in relieving patient's pain that bothered his daily life. After gross resection, this patient experienced complete pain relief without any complication. Meanwhile, periodic follow-up of our patient after treatment was encouraged.

Recently, the pain associated with fibrous dysplasia lesions has been successfully treated with bisphosphonates. In a phase-II trial including patients with both polyostotic and monostotic fibrous dysplasia, there was a substantial decrease in bone pain with intravenous pamidronate after thirty-nine months of follow-up [21]. On the basis of such findings, bisphosphonates should be considered for patients with pain related to FD.

## CONCLUSIONS

Monostotic fibrous dysplasia of the spine, though very rare, can affect either sex with equal frequency and present at any age. This report illustrates a rare case of monostotic fibrous dysplasia involving the sixth vertebrae transverse process of cervical spine, with symptoms of chronic cervical pain that was successfully treated with intra-operative biopsy, gross excision and close follow-up, providing a good option to the clinical practice.

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