

# Fibrous Dysplasia of the Spine – A Case Involving Three Levels of Thoracic Spine

Kenneth M Joyce<sup>1</sup>, Sven O'hEireamhoin<sup>1</sup>, Michael Tan Chien Sheng<sup>2</sup>, Aiden Devitt<sup>1</sup>

## What to Learn from this Article?

*Unusual Presentation of Fibrous Dysplasia of Spine with its Management.*

### Abstract

**Introduction:** Involvement of the spinal column in either monostotic or polyostotic form is rare, with fewer than thirty-five cases discussed in the literature. Most of the cases of polyostotic fibrous dysplasia of spine have involvement of the appendicular skeleton.

**Case Report:** We report a case of a 74-year-old Irish man with a two month history of back pain. Investigations revealed a diagnosis of fibrous dysplasia involving three levels of the thoracic spine in isolation. The patient underwent T2-T9 stabilization and bone grafting.

**Conclusion:** A case of fibrous dysplasia involving three levels of the thoracic spine in isolation has never previously been reported. The extreme rarity of this type of presentation can pose a diagnostic dilemma, and in cases with spinal involvement, a consensus of management has not yet been achieved.

**Keywords:** Fibrous - dysplasia – thoracic - spine.

### Introduction

Fibrous dysplasia is a non-neoplastic fibroosseous lesion of the bone, accounting for 7% of benign bone tumours and 2.5% of all bone lesions. It is characterized by the metaplastic replacement of the medullary component of one bone (monostotic) or less commonly of several bones (polyostotic) with fibrous tissue and irregular osteoid formation. Portions of the bone are replaced by fibrous connective tissue and poorly formed trabecular bone. The process originates in the medullary cavity. It is caused by a post-zygotic mutation in the guanine nucleotide stimulatory protein (GNAS1) gene—. It is more of a skeletal dysplasia than a true

neoplasm.

Fibrous dysplasia most commonly presents in the teens or twenties. It may occur in any bone but is most common in the proximal femur, tibia, ribs, and skull. It affects slightly more males than females. Most patients with fibrous dysplasia are asymptomatic. However fibrous dysplasia may be painful or cause swelling. It can cause repeated pathologic fractures or severe bone deformity, such as the "shepherd's crook" varus deformity of the proximal femur .

When present in the elderly with multiple vertebral lesions, a biopsy may be indicated because metastatic disease or

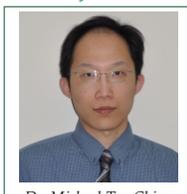
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Dr. Kenneth M Joyce



Dr. Sven O'hEireamhoin



Dr. Michael Tan Chien Sheng



Dr. Aiden Devitt

### Reviewer's Photo Gallery



Dr. Krunal Desai



Dr. Ralf Gahr

<sup>1</sup>Department of Trauma & Orthopaedic Surgery, Galway University Hospital, Galway, Ireland.

<sup>2</sup>Division of Anatomic Pathology, Department of Histopathology, Galway University Hospital, Galway, Ireland.

### Address of Correspondence

Dr. Kenneth Joyce, Department of Trauma & Orthopaedic Surgery, Galway University Hospital, Galway, Ireland. Phone: 353879654586. E-mail: kennethjoyce1@gmail.com

multiple myeloma may simulate a benign non-aggressive process. Therefore, active diagnosis and radiological familiarity of spinal fibrous dysplasia are thought to be essential for preventing unnecessary examinations or procedures.

### Case Report

We report the case of a seventy four year old man who presented to the Emergency Department complaining of back pain of two months duration. This was a dull mid-thoracic back pain with radiation to the chest and upper abdomen. It was initially intermittent associated with exertion, but now was increasing in frequency and severity to the extent that it was interfering with his daily living activities. There was no history of trauma. His family history was unremarkable.

His only significant past medical history was that of hypercholesterolaemia for which he was prescribed a statin by his general practitioner. He underwent surgery for varicose veins in his right leg 7 years previously. His regular medications also included aspirin. He had no known drug allergies and smoked 10 cigarettes per day. He was a retired farmer who lived with his wife and was in good physical condition from an anaesthetic point of view (ASA grade I).

On examination of his back the overlying skin was unremarkable although a mild thoracic kyphosis was present. Physical examination revealed conspicuous tenderness and percussion pain in the thoracic region. The straight leg raising test was negative on both sides. His neurological system was intact on examination. Systemic examination was also unremarkable.

Laboratory studies including total blood count, sedimentation rate, C-reactive protein, electrolyte values, and alkaline phosphate levels were within the normal ranges. Chest radiography showed widening of the right paratracheal stripe, of concern for lymphadenopathy. There were also calcified left hilar lymph nodes. Plain radiographs of the thoracic spine showed mid thoracic kyphosis with loss of distinction of the posterior cortices of T7 and T8.

An MRI of the cervical and thoracic spine was carried out

[Fig 1,2]. This showed an infiltrative process involving the vertebral bodies of T6 and T7, associated with expansion and some destruction. There was extensive demineralisation and destruction of the vertebral body of T6. The process extended inferiorly into the vertebral body of T7, and appeared to cross the disc space. The process also involved the superior and inferior articular facets on the left side at T5. Computerized tomography images [Fig 3] further characterised the extent of the dysplastic process. Reconstructed images revealed the infiltrative process in further views [Fig 4]. This process extended into the pedicles bilaterally and was associated with some compromise of the foramina at T6 and T7. There was no definite nerve root compression identified and no other focal bony abnormalities were seen. Significant multilevel degenerative disc disease was seen in the cervical spine, associated with reversal of normal cervical lordosis resulting in significant multilevel spinal canal stenosis, and some evidence of signal changes in the cord, although focal cord compression was not seen. The findings of an aggressive destructive process were highly suspicious for a malignant process, although the presence of a sclerotic edge suggested an indolent aspect to the process.

The following day this patient underwent T2-T9 stabilization and bone grafting. Pedicle screws were inserted proximally into T3 and T4 pedicles bilaterally [Fig 5], and distally into three levels at T7/T8/T9 [Fig 6]. Spinous processes of T2-T9 were excised. Autologous bone graft was harvested from left iliac crest and used as an onlay graft. The patient's post-operative course was uneventful, managed with analgesia and physiotherapy to assist with mobilisation. He was discharged home day four post-operatively.

Multiple biopsies of bony tissue were sent for histological examination. Histology revealed multiple pieces of woven bone with prominent periosteal fibroblastic reaction and areas showing new bone formation with medullary space fibrosis. In addition, there were focal areas showing irregularly oriented osteoid without osteoblastic rimming but surrounded by fibroblastic proliferation [Fig 7,8]. The overall features were most suggestive of reparative changes

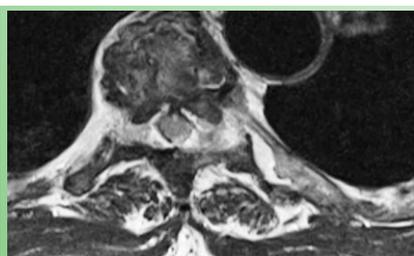


Figure 1: MRI Thoracic Spine at Level of T6.



Figure 2: MRI Thoracic Spine.



Figure 3: CT of Thoracic Spine.



Figure 4: Reconstructed CT of Thoracic Spine.



Figure 5: Intra-operative Screening of Pedicle Screws in T3 And T4.

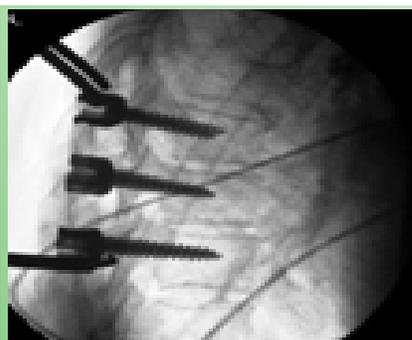


Figure 6: Intra-operative Screening of Pedicle Screws in T7, T8, T9.

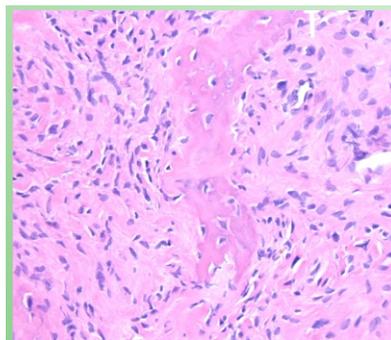


Figure 7: Intra-operative Screening of Pedicle Screws in T7, T8, T9.

rather than a neoplastic process with some features which could be in keeping with fibrous dysplasia in the appropriate clinical settings. The patient had an unremarkable post-operative follow-up. He continues to be asymptomatic with no evidence of recurrence on serial imaging [Fig 9,10].

### Discussion

Fibrous dysplasia was first described in 1891. It is a multisystem disorder that may involve the CNS, endocrine organs, skin and the skeletal system. The skeletal involvement can be monostotic (70– 80%) or polyostotic (20–30%)–. In the monostotic form, the long tubular bones are often involved. On the other hand, the axial bone is more often involved in the polyostotic form. Although fibrous dysplasia accounts for approximately 7% of benign bone lesions, the prevalence of spinal involvement is thought to be very low, particularly in the monostotic form[1]. The presence of multiple lesions of fibrous dysplasia in the thoracolumbar spine in isolation is rare. In a recent study by Leet et al involving a large cohort of patients, none of the cases had isolated spinal localization[11]. McCune-Albright syndrome was not present in this case as there were no associated appendicular lesions, cutaneous manifestations or

endocrinopathies. This syndrome would typically be associated with polyostotic fibrous dysplasia[10].

When symptoms occur, they can include pain, irregular bone growth and increased susceptibility to bone fractures (rare). The patient we described was symptomatic due to pain caused by the bone expansion from T5-T7. In spinal fibrous dysplasia, the vertebral body is affected most frequently, owing to the relatively large amount of cancellous bone[12]. Because of the close relationship to the vertebral body, involvement of the pedicles has been described in nearly all cases in which the vertebral bodies are affected [13].

On plain radiographs, fibrous dysplasia appears as a lytic lesion in the metaphysis or diaphysis with a "ground glass" appearance. There is expansion of the bone and possible bowing. The cortical bone is thinned with a scalloped, undulating pattern due to endosteal erosion[14]. Periosteal reaction usually is absent unless there is a pathologic fracture. MRI is useful for evaluating the soft-tissue components and the entire extent of a lesion. The MRI characteristics of fibrous dysplasia are variable, typically showing signal intensity that is intermediate to low on T1-weighted images and intermediate to high on T2-weighted[15]. The features of no cortical bone violation, no obvious soft-tissue involvement, low signal intensity on T1W sequence and fine well demarcated border between the lesion and normal bone are

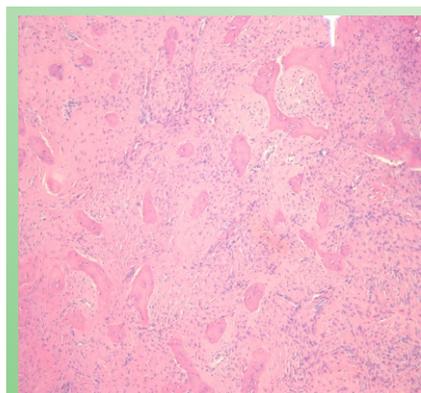


Figure 8: Histology Image of Fibrous Dysplasia.



Figure 9: Lateral Post-operative X-ray.

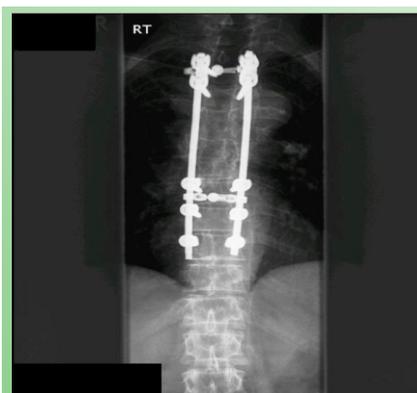


Figure 10: Anteroposterior Post-operative X-ray.

characteristics of a benign lesion[15].

Establishing the diagnosis of fibrous dysplasia in adult can be difficult due to broad differential diagnoses which include metastases, plasma cell myeloma, bone tumours and chronic infectious spondylitis. Furthermore, the lesion may be complicated by fractures and reparative changes. In cases of fibrous dysplasia with lytic lesions, the differential diagnosis should include haemangioma, giant cell tumour and aneurysmal bone cyst. In cases of fibrous dysplasia with blastic lesions, it should include Paget's disease and osteoblastoma[16]. Osteolytic metastasis rarely presents as expansile vertebral lesions because it is invariably associated with vertebral collapse[17]. Most cases of fibrous dysplasia in adult should be established following clinicopathological correlation.

The treatment of fibrous dysplasia depends upon the presence of symptoms. Asymptomatic patients may be observed every six months with serial radiographs as well as analgesic medication[18]. Children with large lesions or lesions in the proximal femur or other weight-bearing bones are observed more frequently[19]. Surgery is indicated for confirmatory biopsy, correction of deformity, prevention of pathologic fracture, and/or eradication of symptomatic lesions[18]. The indication for surgery depends on location and size of the lesion, symptoms and deformity. In cases with spinal involvement, a consensus of management has not been achieved. Surgery is performed when the patient has persistent pain, neurologic impairment, vertebral collapse, instability, and/or cord compression. Bone grafting may be indicated for selected adult patients with monostotic disease. Allograft is preferred to autograft to eliminate donor site morbidity[20]. There may also be an indication for the use of allograft with internal fixation for selected cases where the graft material provides temporary augmentation for the internal fixation[18]. The natural history and ideal treatment for this condition remain poorly understood. Surgical approach is advocated to be tailored to the involvement of the dysplastic tissues.

The deformity of fibrous dysplasia may progress with skeletal growth[3]. Fibrous dysplasia usually is static after growth ceases but may be reactivated with pregnancy[21]. Malignant transformation, which is very rare, must be suspected when there is endosteal or cortical erosion, especially if there is cortical destruction with associated soft-tissue mass. Malignant transformation occurs with a frequency of 0.5% in monostotic form but it may rise to 4% in McCune-Albright syndrome[22].

## Conclusion

A case of fibrous dysplasia involving three levels of the thoracic spine in isolation has never previously been reported. The extreme rarity of this type of presentation can pose a diagnostic dilemma, and in cases with spinal involvement, a consensus of management has not yet been achieved.

## References

### Clinical Message

The disengagement of the humeral head component of the prosthesis is uncommon and has not been previously reported. Systematic and correct rehabilitation exercises are highly important for optimal recovery of shoulder function. Postoperative functional exercise should be carried out in an ordered manner. Also the motivation of the patient is especially important for the treatment success.

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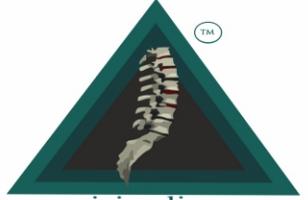
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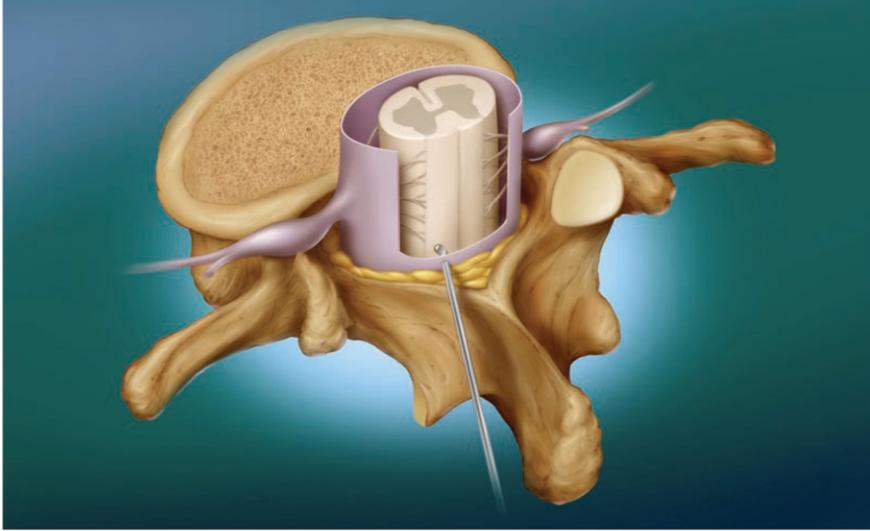


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