

A Rare Case of Monostotic Spinal Fibrous Dysplasia Mimicking Solitary Metastatic Lesion of Thyroid Carcinoma

Mohd Ariff SHARIFUDIN¹, Zamzuri ZAKARIA², Mohamed Saufi AWANG³, Mohamed Azril MOHAMED AMIN¹, Azian ABD AZIZ⁴

Submitted: 25 Jan 2015

Accepted: 10 May 2015

¹ *Musculoskeletal Oncology Unit, Department of Orthopaedics, Traumatology and Rehabilitation, Kulliyah (Faculty) of Medicine, International Islamic University Malaysia, 25200 Kuantan, Pahang, Malaysia*

² *Spine Unit, Department of Orthopaedics, Traumatology and Rehabilitation, Kulliyah (Faculty) of Medicine, International Islamic University Malaysia, 25200 Kuantan, Pahang, Malaysia*

³ *Neurosurgery Unit, Department of Surgery, Kulliyah (Faculty) of Medicine, International Islamic University Malaysia, 25200 Kuantan, Pahang, Malaysia*

⁴ *Department of Radiology, Kulliyah (Faculty) of Medicine, International Islamic University Malaysia, 25200 Kuantan, Pahang, Malaysia*

Abstract

Monostotic fibrous dysplasia of the vertebra is a rare entity. A case of a 53-year-old lady who presented with an 8 months history of pain in the thoracic spine region with paraparesis is discussed. She had a history of papillary thyroid carcinoma and had undergone total thyroidectomy one year prior to her current problem. Magnetic resonance imaging revealed isolated osteolytic lesion over the posterior element of the T12 vertebra with narrowing of the spinal canal causing compression of the cord. The diagnosis of fibrous dysplasia was made histologically. Fibrous dysplasia rarely occurs in axial bones compared with peripheral bones. This case illustrates that osteolytic lesion of the vertebrae should be evaluated with detailed radiological and histopathological examination before an empirical diagnosis of spinal metastasis is made in an adult with a background history of primary malignancy well-known to spread to the bone.

Keywords: *fibrous dysplasia, monostotic, thoracic vertebra, thyroid carcinoma, spinal cord compression*

Introduction

Fibrous dysplasia (FD) is a developmental intrinsic defect of osteoblastic differentiation and enchondral bone maturation resulting immature pattern of ossification. It is characterised by replacement of normal medullary bone by disorganized cellular fibrous connective tissue proliferation stippled with irregularly shaped trabeculae of woven bone leading to structural weakening and pathological fractures (1). It is sporadic in occurrence with equal gender distribution. FD may affect any bone with solitary (monostotic) form being the most common, observed in 70-80% of reported cases (2). Spinal involvement accounts for only approximately 2.5% of cases; mostly occurs in the polyostotic form (1, 3,4). Although it may present at any age, patients are typically in their first two decades of

life at the time of initial presentation (1,2).

This report presents a case of back pain and paraparesis in a middle-age lady with a history of surgically treated papillary thyroid carcinoma. The rarity of monostotic spinal FD in adults prompted the authors to report this case and highlight the challenges that were encountered in confirming the spinal tumorous lesion.

Case Report

A 53-year-old Malay lady presented with an 8-month history of back pain in the lower thoracic region with paraparesis requiring full assistance for her daily activities. The pain was sudden in onset, aggravated by movement and gradually worsened over a period. It was not relieved by oral

analgesics. She developed urinary incontinence a few months after the onset of paraparesis requiring the use of a continuous bladder drainage catheter. She had a history of papillary thyroid carcinoma and underwent total thyroidectomy one year prior to the current presentation. Initial staging with positron emission tomography scan showed no evidence of distant metastasis.

Physical examination at current presentation revealed no obvious deformity of the cervical and thoracolumbar spine but spinal tenderness was elicited at T12 vertebral level. Motor power was Medical Research Council (MRC) Grade 2 with hypotonia and hyporeflexia and sensory loss L2 downward. There were no cutaneous stigmata or clinical signs of endocrinological dysfunction, respectively. Plain radiographs showed an isolated osteolytic lesion involving the T12 vertebra with minimal narrowing of the spinal canal at the respective level (Figure 1).

Magnetic resonance imaging (MRI) of the thoracic spine showed a diffuse lesion with minimal enhancement on gadolinium involving the T12 vertebra body including the pedicles and spinous process. The spinal canal was narrowed causing compression to the cord. The paravertebral soft tissue was normal (Figure 2). Erythrocyte sedimentation rate was raised as a consequence of urinary tract infection. Other blood investigations were within the normal range. Computed tomography (CT) guided percutaneous biopsies were performed twice and both reported as inconclusive with no malignant cells seen. Although the biopsy report was inconclusive, our provisional diagnosis was still on spinal metastasis in view of history of papillary thyroid carcinoma. The aggressive destructive process observed in this case was highly suspicious for a malignant process. Restaging showed no other bony lesions.

The patient underwent posterior decompression with fusion and stabilization of T10-L2 vertebrae with pedicle screw system following deterioration in the symptoms. The indications for decompression were mainly for neural structure release and tissue diagnosis following inconclusive results from the previous procedures. Intra-operatively, soft greyish lesion involving the posterior element of T12 vertebra was observed. Part of the lesion was adhered to the dura, with oedematous and pale surrounding paraspinal muscles. Specimens taken from posterior elements and muscles were sent for histopathology analysis. Histopathology examination revealed fibrous dysplasia (Figure 3).

Postoperatively, she had made good recovery.

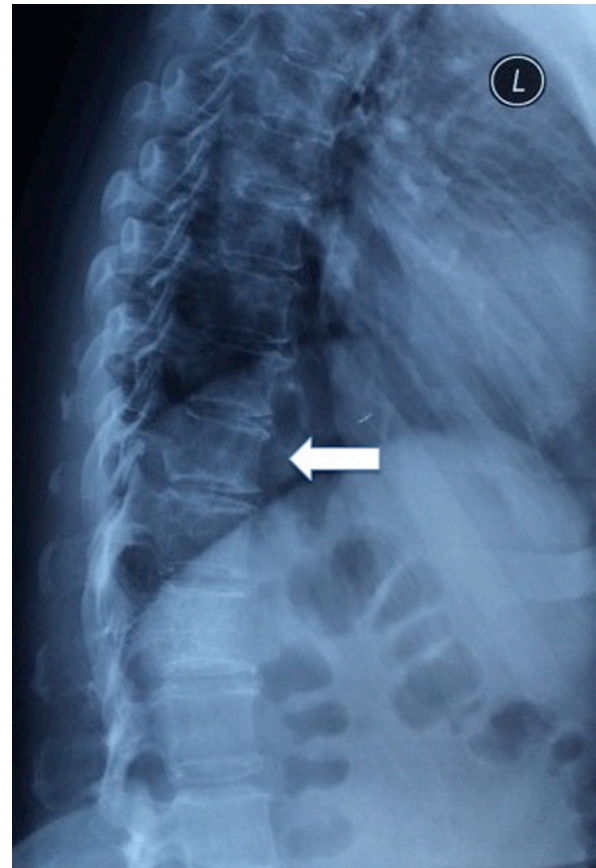


Figure 1: Lateral view plain radiograph of the thoracolumbar spine revealed osteolytic lesion of T12 (white arrow) with minimal loss of vertebral body height and minimal narrowing of canal.

She was discharged two weeks after surgery and her condition improved gradually. She was also started on intravenous infusion of pamidronate 60 mg monthly. At two years, she was able to ambulate without aid. Currently, she is capable of performing daily activities independently and shows no symptoms of recurrence.

Discussion

Monostotic form of FD presenting as solitary spinal lesion in adult is extremely rare. It occurs mostly in the polyostotic form, which is usually associated with specific syndromes and endocrine disorders (2,3). These important associations were not observed in our patient. Furthermore, the progression of bony lesion in FD tends to regress as the bone reached maturity. FD with vertebral involvement has been described in each segment of the vertebrae, with highest prevalence



Figure 2: MRI thoracolumbar spine (a) Sagittal T2, (b) Sagittal T2 STIR, (c) Sagittal T1, (d) Post IV Gadolinium and (e) Axial T1 of T12 vertebrae. T12 vertebra including its pedicles and spinous process is hypointense on T1 and hyperintense on T2 and T2 STIR (white arrow). Only minimal enhancement is observed post IV gadolinium. The spinal canal is narrowed at this level causing compression to the cord. No paravertebral soft tissue mass is visualised. The remaining vertebral body and intervertebral disc spaces (not shown) are normal.

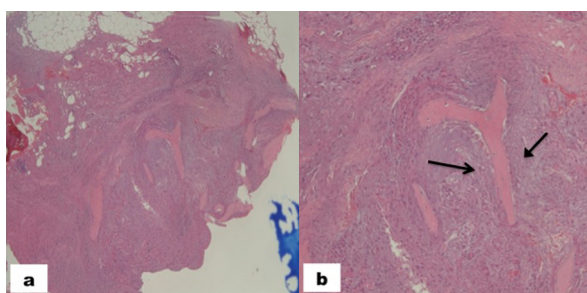


Figure 3: Histopathology examination (a) hematoxylin and eosin x40 and (b) hematoxylin and eosin, x100 showed fragments of chondroid and lamellated bones, fibrous tissue and muscle bundle. Normal and fibrotic marrow were also observed. In some of the most fibrotic area, few narrow, curved and misshapen woven bone trabeculae without osteoblastic rimming was noted (black arrows). There was no malignant epithelial cells infiltration observed.

around the lumbar region. Most reported cases of lumbar spine FD involved lower lumbar vertebrae (1,3). Reported thoracic spine lesions were mainly found in the vertebrae body and posterior elements with no predilection for any thoracic level (3,4).

Patients with monostotic FD usually have painless and asymptomatic lesion, which most

of the time manifest as incidental findings on radiographic examination (1,4). Patient can be symptomatic because of nerve root or cord compression following insufficiency fracture or directly by the tumour itself. Pain is usually proportional to the degree of vertebral body involvement. However, monostotic lesion generally becomes inactive and rarely expands following puberty, with the exception of pregnancy-induced activity within the dysplastic lesion (1,2).

On plain radiographs, monostotic FD of the spine appears similar to other extraspinal lesions, characterized by expanding ground-glass matrix of cancellous bone with variable degree of marginal sclerosis and narrow transition zone (3). Some authors advocate the use of radionuclide bone scintigraphy to demonstrate other bony involvements but false-negative results may occur as the lesions mature (1,2). CT and contrast MRI are indicated to evaluate cortical destruction and soft tissue extension. The features on MRI are variable, typically hypointense on T1-weighted images and hyper- or hypointense on T2-weighted images (3-5).

The gross appearance of FD is soft-firm greyish-white tissue within the medullary cavity, surrounded by shell of cortical bone. Histologically, it appears well circumscribed and formed by the trabeculae of the immature bone. Uniformly cellular fibrous tissue containing proliferation of spindle cells can be found without any features of malignancy and lack of osteoblastic rimming (15). It is also important to recognize

features of possible sarcomatous transformation of FD. Sarcomatous transformation of both monostotic and polyostotic forms are rare, with reported incidence of 0.4% and 2.5%, respectively. The most common malignant histologic subtypes reported were osteosarcoma, fibrosarcoma and chondrosarcoma. Hence, it is highly recommended to correlate the imaging and histologic findings. Features of intralesional calcification, cortical destruction, periosteal reaction and extensive soft tissue extension on imaging are suggestive of malignancy (5). Histologic evaluation will show more atypical cells and mitotic activities, with abundant malignant osteoid deposition in irregular trabeculae (1, 2,5).

In this patient, the diagnosis of spinal metastasis was higher in the list because of the history of papillary thyroid carcinoma. Bone is the third most common target of metastatic cells after the lungs and liver. Metastases from the lungs, breast, prostate, renal and thyroid carcinomas, represent almost 80% of all skeletal metastases. Irrespective of the primary tumour involved, the vertebral column is the most common site for skeletal metastases, particularly the anterior and middle columns. Thoracic spine is the most common, representing almost 70% of cases, followed by lumbar and cervical segments (6,7).

In terms of clinical symptoms, patients with spinal metastases may have a spectrum of presentations varying from back pain (with or without pathological fracture) to spinal instability and cord compression (6,7). In some cases, the metastatic lesions are found only during routine staging procedures. Obviously, there are not much different in terms of clinical presentations between symptomatic FD with spinal metastasis, making the diagnosis often difficult. Diagnosing FD is even more difficult in adults because cases in older patients, particularly that involving the spine, are rare (3,4). The present patient manifested a mechanical type of pain, based on its relationship to her activity and resistance to analgesics. But the effected vertebra did not have the typical ground-glass appearance of a FD lesion on plain radiograph, or features of an aggressive tumorous spinal lesion (Figure 1). Furthermore, findings on MRI did not show significant insufficiency fracture that may cause severe mechanical pain and neurologic compromise. The narrowing of the spinal canal may be caused by direct compression of the tumour (Figure 2).

Treatment of FD depends on the presence and severity of symptoms. Asymptomatic patients with stable lesions require only clinical observation and serial plain radiographs every

6 months (1,2). There is no consensus in the management of cases with spinal involvement. Some authors necessitated complete tumour resection followed by reconstruction of the spine in cases involving the vertebral body and posterior elements. While this approach still remains controversial, as the traditional understanding is that FD is a benign lesion without progressive destructive capabilities, surgery is still indicated for confirmatory biopsy, fracture prophylaxis, deformity correction and symptomatic therapy (1,2,4,5). Bisphosphonates, mainly pamidronate, have been used in cases where surgery is difficult or contraindicated, as well as in polyostotic form of FD. Another form of treatment for FD that is still under research is the use receptor activator of nuclear factor kappa-B ligand (RANK-L) inhibitor denosumab which is reported to have a more significant effect on bone turnover than following bisphosphonate therapy (1,8). Our patient responded well to the treatment given. Following surgery, there was some residual back pain and neurological deficit, which improved further with bisphosphonate therapy.

Another benign bony tumour-like lesion more commonly seen in the paediatric age group was reported in an adult with extensive involvement of the spinal column and thyroid gland. The extensiveness, age of presentation and its rarity made the diagnosis difficult. It was initially thought to be a secondary metastasis until it was confirmed on histopathology examination (9). We did not find any correlation between FD and thyroid carcinoma in the literature. A review of the literature showed a case reported in 1977 in which the authors suggested that a coexisting dystrophic fibrosis with goitre is suggestive of a metastatic lesion from a thyroid carcinoma (10). However, associations between polyostotic FD and endocrine diseases have been established (2, 3,5).

Conclusion

Monostotic form of FD involving the thoracic spine is a rare entity. The extreme rarity of this presentation may pose a diagnostic dilemma, particularly in adults with risk of metastatic disease. Benign bony lesions should be included in the differential diagnoses of adult patients with osteolytic lesions in the vertebral body and posterior elements. These benign lesions, although uncommon, may have a risk of sarcomatous transformation, with radiation therapy as one of the risk factor.

Acknowledgement

The authors wish to thank the staff of the Department of Orthopaedic, Traumatology and Rehabilitation, and Neurosurgical Unit of Department of Surgery, International Islamic University Malaysia for their contribution and support in completing the report.

Funds

None.

Conflict of Interest

None.

Authors' Contributions

Conception and design: MAS, MSA
Analysis and interpretation of the data: ZZ, AAA
Drafting of the article: Critical revision of the article for important intellectual content: MAS
Final approval of the article: MAS, ZZ, MAMA
Collection and assembly of data: MAMA

Correspondence

Associate Professor Dr Zamzuri Zakaria
MD (UKM), MSurg (Ortho)(UKM)
Department of Orthopaedics Traumatology and Rehabilitation
Kulliyah (Faculty) of Medicine
International Islamic University Malaysia
Jalan Sultan Ahmad Shah
25200 Kuantan
Pahang, Malaysia
Tel: +609-570 6131
Fax: +609-514 4451
Email: zamdr@yahoo.co.uk

References

1. DiCaprio MR, Enneking WF. Fibrous dysplasia. Pathophysiology, evaluation, and treatment. *J Bone Joint Surg Am.* 2005;**87(8)**:1848–1864. doi: 10.2106/JBJS.D.02942.
2. Ippolito E, Bray EW, Corsi A, De Maio F, Exner UG, Robey PG, et al. Natural history and treatment of fibrous dysplasia of bone: a multicenter clinicopathologic study promoted by the European Pediatric Orthopaedic Society. *J Pediatr Orthop.* 2003;**12(3)**:155–177. doi: 10.1097/01.bpb.0000064021.41829.94.
3. Gogia N, Marwaha V, Atri S, Gulati M, Gupta R. Fibrous dysplasia localized to spine: a diagnostic dilemma. *Skeletal Radiol.* 2007;**36(Suppl 1)**:S19–S23. doi: 10.1007/s00256-006-0102-8.
4. Oba M, Nakagami W, Maeda M, Kobayashi K. Symptomatic monostotic fibrous dysplasia of the thoracic spine. *Spine (Phila Pa 1976).* 1998;**23(6)**:741–743. doi: 10.1097/00007632-199803150-00019.
5. Fitzpatrick KA, Taljanovic MS, Speer DP, Graham AR, Jacobson JA, Barnes GR, et al. Imaging findings of fibrous dysplasia with histopathologic and intraoperative correlation. *Am J Roentgenol.* 2004;**182(6)**:1389–1398. doi: 10.2214/ajr.182.6.1821389.
6. Hage WD, Aboulaflia AJ, Aboulaflia DM. Incidence, location and diagnostic evaluation of metastatic bone disease. *Orthop Clin N Am.* 2000;**31(4)**:515–528. doi: 10.1016/S0030-5898(05)70171-1.
7. Singh K, Samartzis D, Vaccaro AR, Andersson GBJ, An HS, Heller JG. Current concepts in the management of metastatic spinal disease. The role of minimally-invasive approaches. *J Bone Joint Surg Br.* 2006;**88(4)**:434–442. doi: 10.1302/0301-620X.88B4.17282.
8. Ganda K, Seibel MJ. Rapid biochemical response to denosumab in fibrous dysplasia of bone: report of two cases. *Osteoporosis Int.* 2014;**25(2)**:777–782. doi: 10.1007/s00198-013-2585-1.
9. Mohd Ariff S, Joehaimey J, Ahmad Sabri O, Zulmi W. Langerhans cell histiocytosis with extensive spinal and thyroid gland involvement presenting with quadriparesis: an unusual case in an adult patient. *Malays Orthop J.* 2011;**5(3)**:28–31. doi: 10.5704/MOJ.1111.004.
10. Bielicki F, Dolinski J, Noczynski L, Wolek M. Case of dystrophic fibrosis with coexisting goitre suggesting metastases of thyroid carcinoma. *Endokrynol Pol.* 1977;**28(6)**:551–555.