



## SARCOMATOUS TRANSFORMATION IN FIBROUS DYSPLASIA- A RARE CASE.

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### ABSTRACT

Malignant transformation in a Fibrous Dysplasia is a rare entity. Osteosarcoma is the most common sarcoma that arises from Fibrous Dysplasia followed by fibrosarcoma and chondrosarcoma. A 70 years old gentleman presented with right knee pain for one month. He is a known case of Fibrous Dysplasia diagnosed 10 years ago. Magnetic Resonance Imaging revealed lytic lesion in distal third of right femur suggestive of Giant cell tumour. Wide resection of the distal femur was performed. Pathological examination revealed pale white, solid and fleshy nodule in the diaphysis measuring 4x3.1x2.8cm. On microscopy, a cellular spindle cell lesion was noted with cells exhibiting moderate nuclear pleomorphism, nucleomegaly, vesicular to hyperchromatic nuclei and small nucleoli. Necrosis, malignant osteoid, malignant chondroid, bizarre tumour cells or giant cells were not evident. A diagnosis of secondary Fibrosarcoma, Grade 2 arising in a Fibrous Dysplasia was rendered. We are reporting this case for its rarity in two aspects- fibrosarcoma of the bone and malignant transformation of Fibrous dysplasia. This case highlights strict scrutiny of symptoms and signs like pain, swelling or lytic bone lesions in a patient with Fibrous Dysplasia as they could be indication for a malignant transformation.

### KEYWORDS

Fibrous dysplasia, fibrosarcoma, malignant transformation

### ARTICLE HISTORY

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### INTRODUCTION

Fibrous dysplasia(FD) is a benign developmental disorder of the bone where fibrous tissue replaces normal bone and bone marrow, as a result of which the bone is weakened and prone to fractures, deformities and functional impairment. There are two forms of fibrous dysplasia-monostotic involving one bone and polyostotic involving multiple bones. The bones most commonly involved are craniofacial bones, ribs and long bones<sup>1</sup>. Malignant transformation in FD is very rare accounting for 0.5% of monostotic FD and 4% of polyostotic FD<sup>2</sup>.

### Case History

A 70 years old gentleman presented with pain and swelling in the right knee and difficulty in walking more than 50 metres for the past one month. He is a known case of FD diagnosed 10 years ago following a fracture for which he underwent curettage, bone cementation and internal fixation with plate and screws in the distal right femur. Routine investigations were normal. Plain radiography showed an expansile lesion in distal end of right femur. Magnetic Resonance Imaging revealed a large expansile osteolytic lesion with fluid levels involving the distal third of right femur and extending upto the articular surface- probable Giant cell tumour (Figure 1). The patient underwent wide resection right distal femur and CM prosthesis. The specimen was received in the histopathology department. Coronal section through the femur revealed multiple cystic spaces with mucoid to serous fluid with intervening firm areas spanning 20x14.5x9cm involving epiphysis, metaphysis and part of diaphysis. A pale white, solid and fleshy nodule was seen in the diaphysis measuring 4x3.1x2.8cm located 6.2cm from the bone resected end (Figure 1). Microscopic examination revealed a variable cellular lesion with areas of cystic change and dilated congested vascular spaces devoid of epithelial lining. The spindle cells are arranged as fascicles and storiform pattern amidst trabeculae of mature bone. In some areas, there is increase in cellularity of spindle cells exhibiting moderate nuclear pleomorphism, nucleomegaly, vesicular to hyperchromatic

nuclei and small nucleoli. Mitotic count was 4-5 per10 high power fields in these areas(Figure 2). Necrosis, malignant osteoid, malignant chondroid, bizarre tumour cells or giant cells were not evident. The tumour extended upto the periosteum. All round margins were free of tumour. The diagnosis of Secondary Fibrosarcoma, Grade 2 arising in a Fibrous Dysplasia was rendered.

### DISCUSSION

Malignant transformation of FD is very rare. Osteosarcoma is the most common malignancy rising in a FD followed by fibrosarcoma and chondrosarcoma<sup>3</sup>. Ruggieri et al studied 1122 cases of FD of which 28 cases showed sarcomatous transformation- osteosarcoma (19 cases), fibrosarcoma (5 cases), chondrosarcoma (3 cases) and malignant fibrohistiocytoma (1 case)<sup>4</sup>. The most common bones involved are the craniofacial bones. In our patient, adequate sampling was done to look for malignant lace like osteoid, atypical chondroid tissue and large bizarre atypical cells. Multiple sections did not reveal the same, thereby ruling out an osteosarcoma, chondrosarcoma and malignant fibrohistiocytoma .

In a case series done by Qu N et al on malignancies arising in monostotic FD, 50% of the patients had history of surgery, none had history of radiation, 70% developed osteosarcoma and 20% developed fibrosarcoma.<sup>5</sup> Imaging studies revealed poorly marginated, mineralized, and osteolytic lesions in these patients. A study by Xu D et al observed that pain, swelling and osteolytic lesions are useful clinical and radiological features that favour a sarcomatous transformation in a FD<sup>6</sup>. Our patient with long standing monostotic FD of right femur had history of surgery, subsequent development of pain in the right knee after 10 years. Imaging revealed osteolytic lesion in the right distal femur.

Fibrosarcomas of the bone are rare and account for less than 5% of bone sarcomas<sup>7</sup>.Clinically they present as swelling and restriction of

movement. Imaging studies show lytic lesions, with a geographic, moth-eaten or permeative pattern of bone destruction. The important factors affecting prognosis are age, tumour site and tumour grade<sup>7,8</sup>. Tumour grading is based on cellularity, atypia and mitosis. Grade 1 tumours show collagen bundles amidst the spindle cells, mild nuclear atypia and rare mitosis. Grade 2 tumours show more cellularity, atypia and mitosis compared to grade 1 tumours. Grade 3 tumours show increase in cellularity and mitosis. Necrosis may be present. Collagen is minimal. Grade 4 tumours show abundant prominent mitosis and necrosis, smaller cells arranged compactly with no intervening collagen<sup>9</sup>. Recent studies suggest that the term Fibrosarcoma should be reserved for grade 1 and grade 2 tumours and those with grade 3 and grade 4 should be classified as Malignant Fibrous Histiocytoma<sup>10</sup>. Our patient was diagnosed as grade 2 Fibrosarcoma.

We are reporting this case for its rarity in two aspects- fibrosarcoma of the bone and malignant transformation of FD. With this case we would like to highlight that development of pain, swelling and osteolytic lesions in a longstanding case of FD should be dealt with a high index of suspicion for malignant transformation.

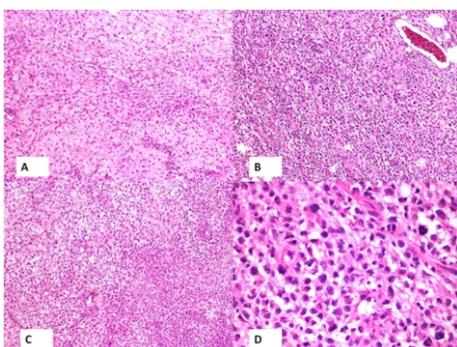
#### Patient Declaration Statement

"The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed."

**Figure 1:** (A) Plain radiography showing an expansile lesion in distal end of right femur with an insitu plate and screws; (B) and (C) Magnetic Resonance Imaging showing a large expansile osteolytic lesion with fluid levels involving the distal third of right femur; (D) Gross photograph showing coronal section through the femur revealing multiple cystic spaces and a fleshy nodule in the diaphysis.



**Figure 2:** (A-C) Photomicrograph showing a cellular tumour composed of spindle shaped cells arranged in fascicles and storiform pattern (H&E X 100); (D) Photomicrograph showing moderately pleomorphic cells with nucleomegaly, hyperchromatic nuclei and mitotic figures (H&E X 400)



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