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A Rare Case of Fibrous Dysplasia of Distal Femur with Secondary Aneurysmal Bone Cyst in an Adult Patient

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Introduction: Aneurysmal bone cyst occurring secondary to fibrous dysplasia is rare. While both are benign lesions, pain, compression of nearby structures and risk of fracture can require treatment.
Presentation of case: In this report, we describe a 36 year old female who developed aneurysmal bone cyst secondary to fibrous dysplasia in the distal femur. She required curettage and bone grafting in the defect created.
Discussion: The diagnosis of a secondary lesion can prove difficult. It is important to exclude a malignant disease process, particularly when imaging demonstrates an aggressive appearance. In this case, excisional biopsy was done to confirm the diagnosis.
Conclusion: Treating a bone tumour must be a multidisciplinary approach involving Radiologist, Pathologist and Orthopaedic surgeon.
Keywords: Aneurysmal bone cyst, Fibrous dysplasia
List of abbrevations : FD- fibrous dysplasia , ABC- aneurysmal bone cyst

I. INTRODUCTION:

Fibrous dysplasia(FD) is an unusual hamartomatous bone disorder characterized by fibro-osseous metaplasia. It is an intraosseous neoformation of fibrous tissue and bone caused by an anomalous development of mesenchymal tissue that can be monostotic or polyostotic. It represents 5% of all benign bone tumors. Usually, it presents clinically in children and adolescents with a median onset age of 8 years. The most common sites are the femur, tibia, craniofacial bones, ribs, humerus, pelvis in decreasing frequency. It is caused by sporadic mutation of the GNAS1 gene located on chromosome 20q.(1)

Aneurysmal bone cyst (ABC) is an expanding osteolytic lesion commonly seen at the meta-diaphyseal regions of long bones. It appears as blood-filled cavernous spaces without endothelial cell lining. The cysts are separated by septa made up of fibrous tissue, which contain multinucleated giant cells and osteoid tissue(2). ABC most commonly involves the vertebrae and long bones.

Fibrous dysplasia presenting with secondary ABC is very rare, only a few cases have been reported in the literature. In this case study, we are presenting a rare case of fibrous dysplasia of the distal femur with secondary ABC in an adult female patient. Although the mechanism of the occurrence of FD with ABC is unknown, there is a report that a secondary form of ABC may arise from a disruption in the osseous circulation caused by the primary lesion.(3)

II. CASE REPORT:

A 36-year-old female patient came to the orthopedics outpatient department with the chief complaint of pain of the right knee for 5 months. Pain is sudden in onset, gradually progressive, non-radiating, aggravated by movements, relieved with rest and medication. There was no history of trauma, fever, significant weight loss, or loss of appetite. There was no history of any other swellings. Physical examination did not reveal any findings (no swelling, no local rise of temperature, no knee range of movement restriction, no lymphadenopathy, and no distal neurovascular deficits).

A plain X-ray of the affected area revealed a mixed lytic and sclerotic lesion involving the right distal femur at the meta-diaphyseal region extending into the epiphysis with ill-defined borders. There was no obvious periosteal reaction or soft tissue involvement. (Figure 1)

MRI revealed a well-defined large eccentric intramedullary soft tissue lesion with cystic areas in the distal meta-diaphyseal region of

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the right femur predominantly in the lateral condyle. The lesion measured 6.3x4.3x4.8cms in craniocaudal, anteroposterior, and transverse planes respectively. It appeared isointense to muscles on T1 and heterogeneously hypointense on T2 and hyperintense on STIR images. Few T2 and STIR hyperintense cystic areas are noted within the lesion, with a sclerotic rim. Cortical thinning was noted in the lateral condyle with a focal area of cortical bulge in the posterior aspect of the lesion in the supracondylar level. There was no extension into the femoral articular surface (Figure 2). Anterior cruciate ligament sprain, grade II tear of the lateral collateral ligament at femoral attachment, and grade III tear in anterior horn and body of lateral meniscus was noted.

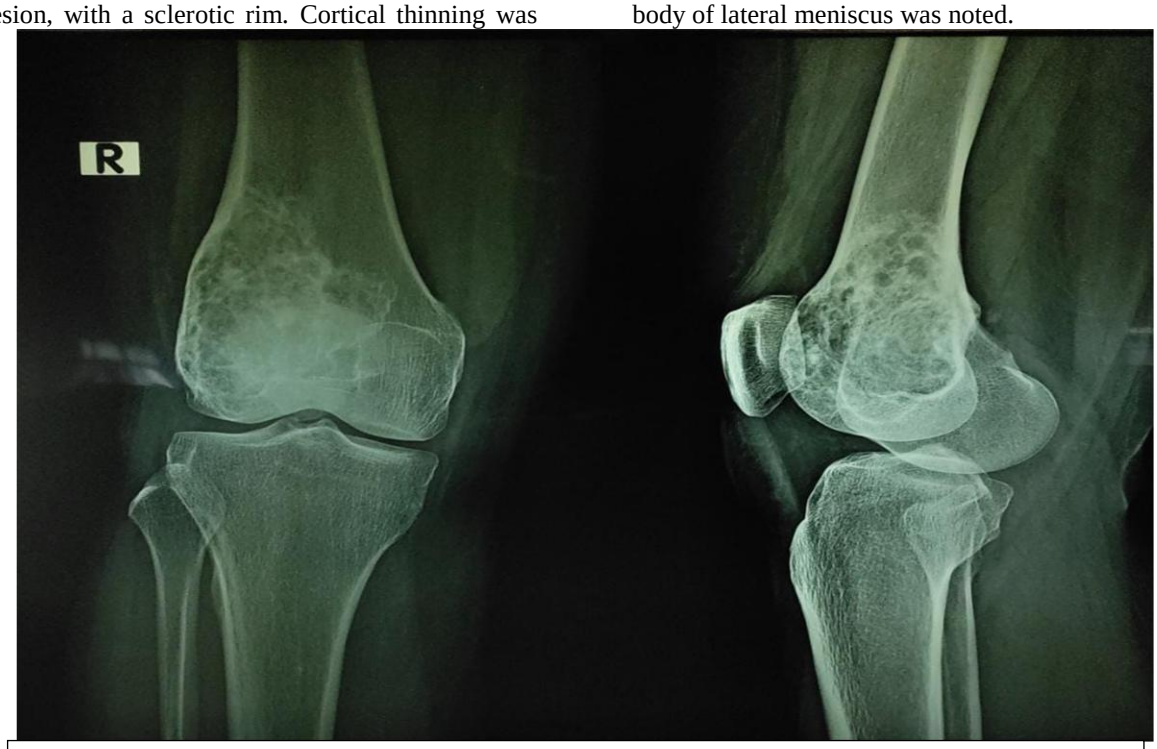

Figure 1: Preoperative X-ray – anteroposterior and lateral views showing the lesion

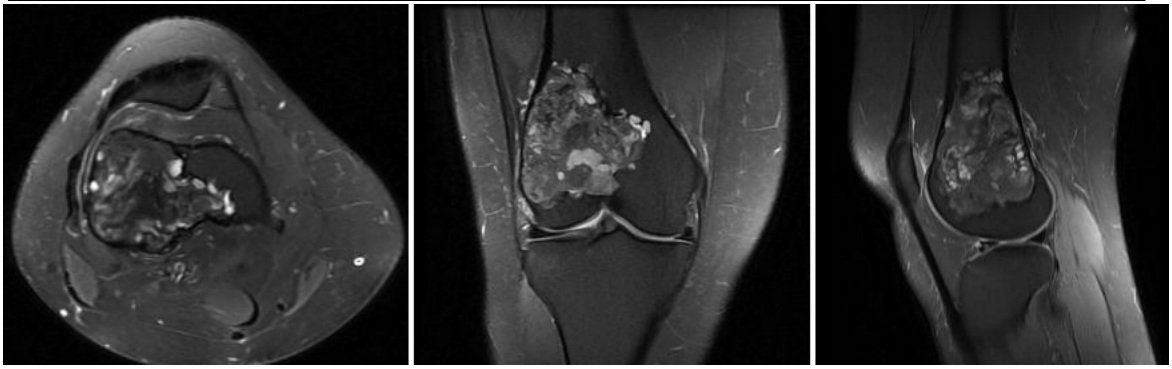

Figure 2 : MRI showing mixed lesion involving lateral condyle of distal femur

All the blood parameters were within normal levels.

We proceeded to operate upon the patient. Intraoperatively, a cortical window was created on the lateral aspect of the femur and all the tumorous tissue was scooped out under an image intensifier. The margins of the lesion were thickened and gritty. En-bloc resection of the bony lesion including surrounding healthy bone was performed for a clear resection margin. (Figure 3) Hydrogen peroxide was applied as a part of the extended curettage. The defect was filled with gel foam on the articular side and with Polymethylmethacrylate (PMMA) cement in the rest of the defect. (Figure

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4) Postoperatively patient was advised non-weight bearing on that limb and knee immobilization was done.

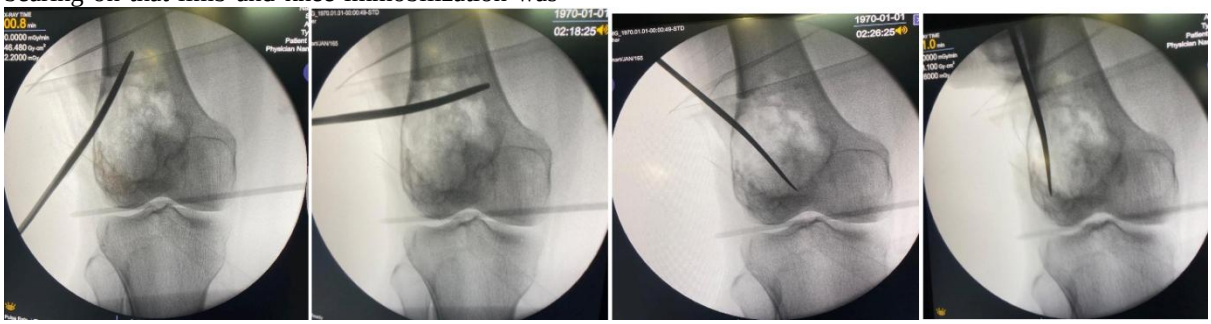

Figure 3: Intraoperative pictures showing clear margins following excision of the lesion.

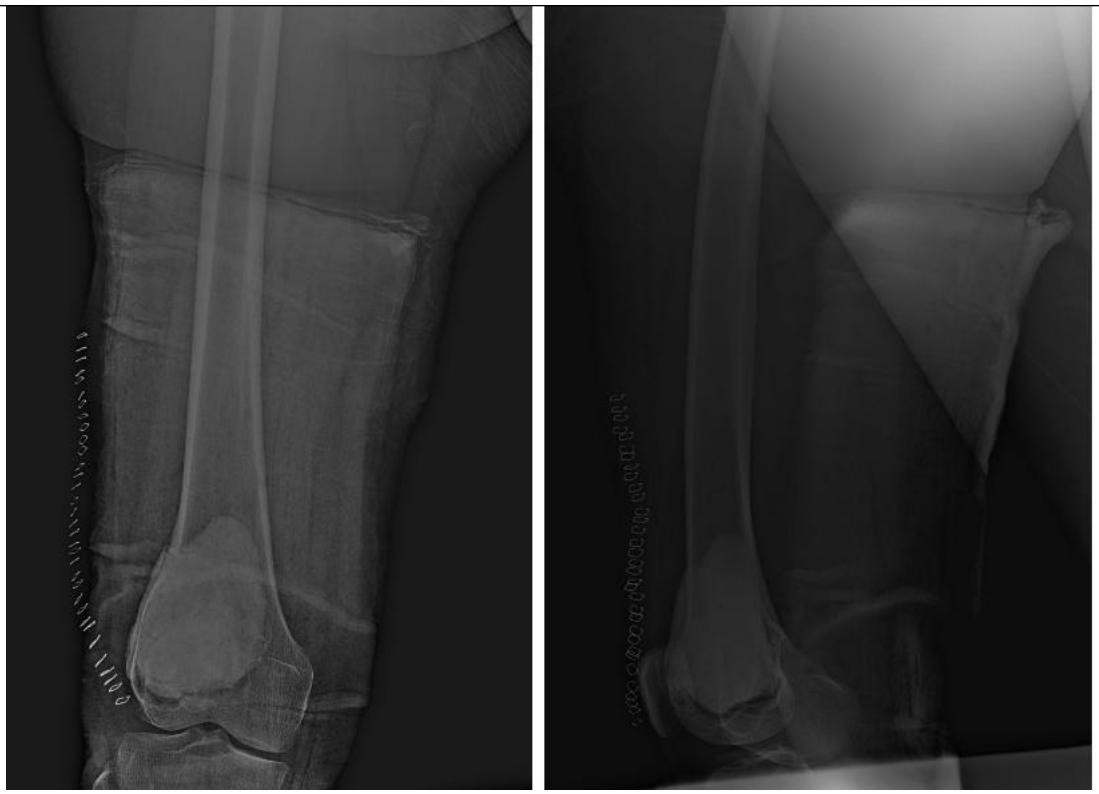

Figure 4: Post-operative radiographs showing cement filling in the defect

The histopathological report showed a lesion composed of predominantly fibrous tissue with loosely arranged fibroblasts amidst which are seen curvilinear Chinese letters type of woven bone. Foci showed large blood-filled spaces with no endothelial lining and wall of fibrous tissue with few osteoclastic type of giant cells. The histological picture was suggestive of fibrous dysplasia with secondary aneurysmal bone cyst.

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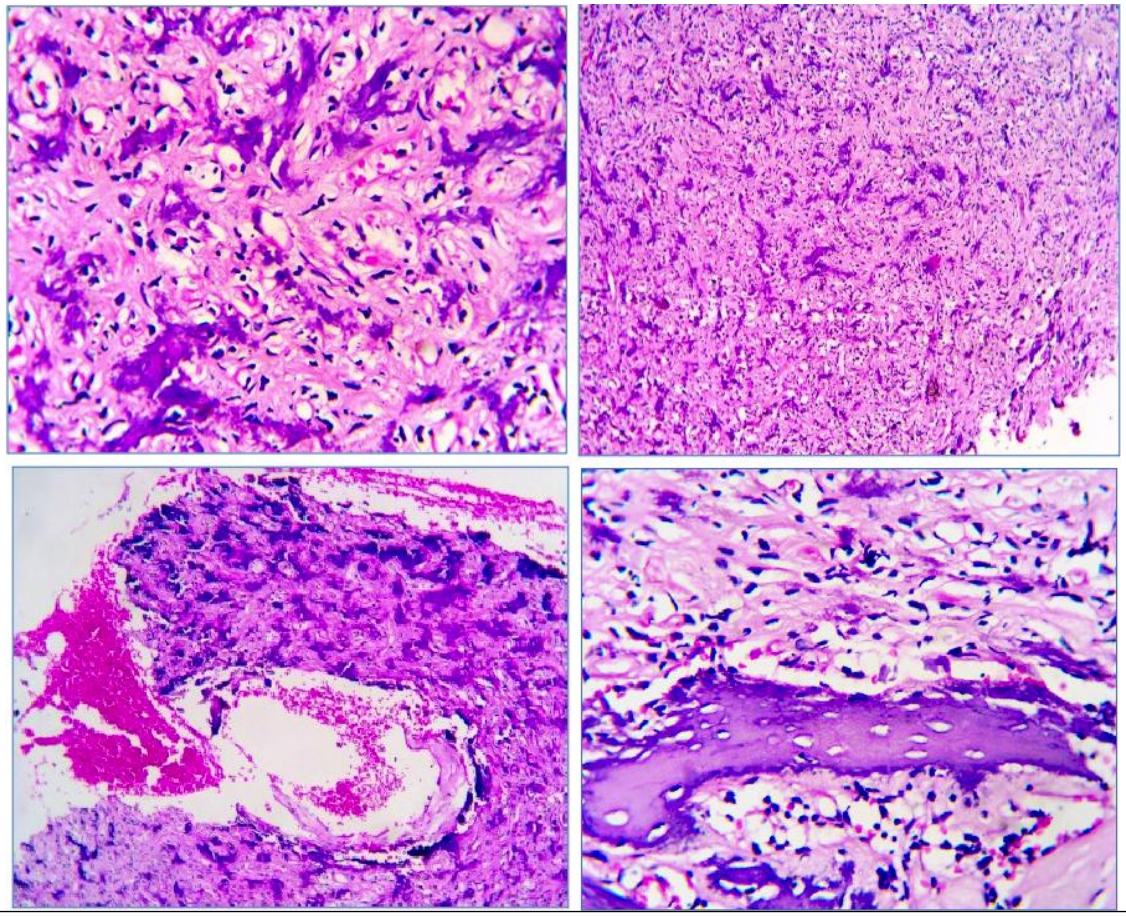

Figure 5: Histopathological images showing fibrous tissue and irregularly spaced trabecular tissue in between, vascular spaces lined by osteoclastic giant cells & fibrous tissue.

III. DISCUSSION:

FD with ABC is rare. Choi et al. reported a case affecting the proximal tibia in a toddler. Fernandez et al reported a case in an 8year old boy.(4) Our case is a 36 year old female patient .

FD with ABC has been reported in facial bones(5), skull base, orbit (6), and calvarium(7) . It has been reported in the spine.(8) Montalti et al. reported a case involving the proximal femur(9). This case is probably the first case of monostotic fibrous dysplasia with secondary ABC in an adult distal femur.

Most of the reported cases have occurred with monostotic FD. The presentation of both FD and ABC depends on the site of involvement, rate of growth, and proximity to important structures such as the brain, eye, and nerves. Pressure symptoms may also be a presenting feature. A pathological fracture may occur (10). Symptoms may include a painless or painful mass, as in our case.

The diagnostic modalities include imaging studies, especially CT /MRI and histopathological examination. In our case histopathology aided in the diagnosis of FD with secondary ABC.

Treatment of FD includes bisphosphonates with regular follow-up and surgery in some symptomatic cases. (11) ABC on its own may be treated by selective arterial embolization or by surgery. Treatment of secondary ABC is that which is appropriate for the underlying lesion.

IV. CONCLUSION:

We are reporting this case of Fibrous dysplasia with secondary ABC in view of its rarity. It may not necessarily present with the classic signs

of a bone tumor. The radiological report may be misleading as in our case and histopathology gave us the final diagnosis. So, treating a bone tumor entails the coordination of a team comprising a Radiologist, Pathologist, and Orthopaedic surgeon.

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Consent for publication – Written informed consent was obtained from the patient for publication of this case report and accompanying images.

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Dr.K.Satya Kumar – made substantial contributions to the conception or design of the work; or the acquisition, analysis, or interpretation of data; or the creation of new software used in the work , approved the version to be published

Dr.T.Sunil – conceptualization, design, acquisition of data, drafting, and review of this article

Dr.I.V.Renuka - drafted the work or revised it critically for important intellectual content

Dr.Harika – data analysis , manuscript preparation