Case report of a Young Girl with Fibrous Dysplasia (FD) Concomitant with Secondary Aneurysmal Bone Cyst (ABC).

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Authorship and contribution Declaration:
Each author of this article fulfilled ALL 04 Criteria of Authorship:
1. Conception and design of or acquisition of data or analysis and interpretation of data.
2. Drafting the manuscript or revising it critically for important intellectual content.
3. Final approval of the version for publication.
4. All authors agree to be responsible
for all aspects of their research work.

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ABSTRACT

Background: Fibrous dysplasia (F.D.) is a developmental bone disorder in which normal matrix of bone is replaced by fibrous tissue. Most common bones effected are ribs, long bones and pelvis. A benign type of bone lesion known as Aneurysmal bone cyst (ABC). Common bones affected by ABC are tibia and femur. Concomitant Fibrous dysplasia with secondary ABC is long bones is very rare.
Case Report: We are presenting a case report of 22 years old girl, who presented to our outpatient department with complain of pain right thigh and leg. Work up was done and based on radiological findings she was diagnosed with Fibrous dysplasia (F.D). Surgery was planned for curettage and bone grafting. Per op there was blood filled sinus and fibrous septa, Biopsy was sent, it turned out to be fibrous dysplasia concomitant with secondary ABC.
Discussion: Concomitant FD with Secondary ABC is very rare. After through literature review, we could identify two cases of concomitants FD with ABC in long bones. So, it was a rare case presentation.
Keywords: Fibrous Dysplasia (FD), Aneurysmal bone cyst (ABC), Benign bone lesions and Bone grafting

This article may be cited as: Wahab H, Birlas A, Ibrahim M, Khattak N S, Qadir R I. Case report of a Young Girl with Fibrous Dysplasia (FD) Concomitant with Secondary Aneurysmal Bone Cyst (ABC). J Pak Orthop. Assoc. (JPOA), Vol 35 (2) June, 2023

BACKGROUND

Fibrous dysplasia (FD) is a bone growth disorder in this condition normal bone matrix is replaced by poorly organized, inadequately mineralized immature bone and fibrous tissue as a result of abnormal proliferation of fibroblasts. More resorption of bone is seen in fibrous dysplasia lesion.

Fibrous dysplasia can involve any bone in the skeleton. The most commonly affected bones are long bones, ribs, pelvis and skull. [1]

FD has two type either can be monostotic (involving single bone) or polyostotis (involving multiple bones). Monostotis is more common. [1]

Most common presentation of FD is pain, and when present in maxillofacial or cranium it presents as painless mass. Most FDs are self-limiting, with stabilization after puberty. [2]

Aneurysm cysts (ABCs) are benign bone lesions, composed of multiple thinned walled interconnecting cystic cavities containing blood. They are relatively rare lesions, representing only 1 to 2 % of bone tumors. These are expansile lytic lesions with aggressive bone destruction. [3,4]

Fibroblasts, osteoclasts, and reactive woven bone line and surround these numerous thin-walled cystic spaces, which are bordered by osteoblasts.[2,3]

There are mentioned numerous instances of polyostotic FD in literature but very few cases with ABC. Planning and surgical intervention is varied, symptomatic lesions should be given priority followed by asymptomatic lesions. [5]

ABC is more common in patients less than 20 years; most commonly affected bones are tibia and femur.

Secondary type of ABC is linked with well-organized already present bone lesions, like ossifying fibromas, osteoblastoma and fibrous dysplasia. [1]

Management of ABC is intralesional curettage and bone grafting. There are chances of local recurrence after curettage up to 8 to 25 %. [3,6,7]

CASE PRESENTATION

A 22 years old girl, resident of Swat district (Pakistan) presented to our outpatient department with complaints of right thigh and right leg pain for last 10 years. Pain has progressively increased over time. Now for last few months it is associated with limping. There was no history of trauma or fever. The patient has visited multiple doctors during last 10 years and she was given painkiller with which her pain improved.

On Physical examination the patient was alert, cooperative. Average height and built girl. There was no limb length discrepancy. Right lower limb was normal on inspection but was slight tender on deep palpation, and also range of motion of her hip and knee were with in normal range, except that extreme of movements were tender. Her Vitals were with in normal limit. General examination of the patient revealed no significant abnormal findings.

Baseline Laboratory tests including Complete blood count, ESR, CRP, Renal Profile and RBS were within normal limits.

X ray showed ground glass appearance in femur and tibia as shown in Picture 01. Based on radiological findings she was diagnosed with fibrous dysplasia and she was planned for staged procedures. 1st stage was planned for curettage biopsy and bone grafting and DHS right femur, in second stage she was planned for curettage of tibia lesion and bone grafting. She underwent her Right femur curettage and DHS and bone grafting and Right Tibia curettage and Bone grafting was done 5 weeks later in second stage.

Allograft bone was used for bone grafting, which is available in our hospital, North West General Hospital.

Patient continues to do well on 1 month follow up.

Radiology: On x ray the appearance of fibrous dysplasia is usually homogeneous with thinning of cortices. Mostly the cortex is intact, as the lesion is expansile the cortex is thinned. The lesion may appear ground glass, completely lucent (cystic) or sclerotic, no periosteal reaction.

Work up was done, X ray showed ground glass appearance in femur and tibia as shown in Picture 01.

Radiographs:

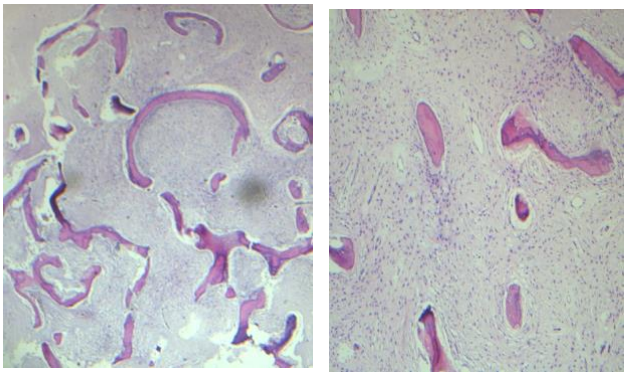
X ray Femur and tibia: Multiple lytic foci of variable sizes with thin sclerotic border rim and narrow zone of transition no adjacent soft tissue involvement seen. No definite fracture seen.

There is increased density with ground glass appearance of the medullary cavity with associated mild expansion. There is suggestion of subperiosteal bone ridges, thinning and endosteal scalloping of the overlying cortex intervening with the normal cortex at places

Similar appearances are evident in right femur with lytic expansile lesion in the proximal femoral metaphysis having a narrow zone of transition and mildly scalloping the overlying cortex without definite cortical break or periosteal reaction.

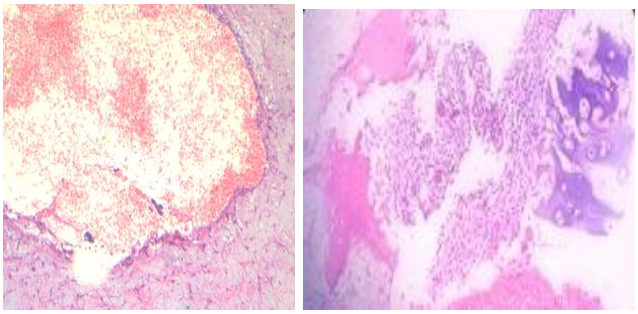
Post op imaging of right femur shows the radiolucent regions have been filled with bone grafts in femur and tibia.

Histopathology: Histologic examination of the specimen from Osteolytic lesion right neck and head of femur demonstrated areas with a fibro-osseous appearance reminiscent of fibrous dysplasia with irregular trabeculae of woven bone that were without lining osteoblasts and without atypia or mitotic activity in the intervening spindle cells (Fig.1 A and B). However, sections from Osteolytic lesion in right femur shaft reveal areas dominated by foci of hemorrhage and dilated pseudo vascular spaces with associated multinucleated giant cells (Fig.2A and B). A focus of solid spindle cell component without woven bone was identified. Fibrous dysplasia, like many bone lesions, can undergo secondary changes, including ABC formation.



A B

Fig 1: Osteo lytic Lesion right neck and head of femur: A. Branching and anastomosing irregular trabeculae of woven bone ("C" and "S" shapes) with no conspicuous osteoblastic rimming.B. Intervening fibrous stroma containing cytologically bland spindle cells.



A B

Fig 2: Osteolytic lesion in right femur shaft: A. Large blood filled cystic spaces separated by cellular septa.B septa contains fibroblasts, giant cells and Calcified, basophilic material

Follow up: Based on radiological findings she was diagnosed with fibrous dysplasia and she was planned staged procedure. She underwent stagged surgical procedure. 1st stage was planned for curettage biopsy and bone grafting and DHS right femur, in second stage, 5 weeks later she was planned for curettage of tibia lesion and bone grafting.

Allograft bone from bone bank was used for bone grafting, which is available in our hospital, North West General Hospital and Research center.

Post operation the results came out to be great with patient being mobilized on the 2nd post op day with great recovery and physical activity, patient was discharged on home medication. On 3 months follow up patient showed improvement is physical activity and pain.

DISCUSSION

It is difficult to determine the exact incidence of the FD due to majority of the asymptomatic cases. But still it is estimated that FD consists 5-7 % of all primary bone tumors.[8] Both genders are affected equally and affects mostly adolescents and young adults.

According to a multicenter study conducted by European Pediatric Orthopedic Society the most commonly affected bone is Femur followed by tibia.[9]

According to a study published in 2018 by Mine Ozsen et al, femur bone was involved in 15 out of 36 cases.

Pain is the most common presenting complain, followed by swelling and bone fracture (pathological).[10]

Reported by Chapurlat, that as compared to pediatric population in adult patients' pain was the most common presentation, pain at the involved site was presenting complain in 67 percent patients. Most

patients with Fibrous dysplasia (FD) are asymptomatic especially monostatic type.[9,10,11]

Polyostotic form of FD presents with deformity in craniofacial bones and femur (i.e. the Shepherd's crook) and pathological fracture. Deformity and fractures are more common in weight bearing bones.[10, 12]

Concomitant FD with ABC, lesion is an extremely rare entity. Concomitant ABC with FD was first reported in 1971 (Buraczewski et al).[13] The primary diseases known to be associated with ABC are osteoclastoma, osteosarcoma, osteoblastoma and hemangioma. The report of ABC associated with FD is very rare.

A few studies have reported concomitant FD and ABC affecting the skull bones. Our literature search performed in the PubMed/MEDLINE/Google Scholar databases and the gray literature revealed 46 cases (Table 1), and most cases described involvement of the skull base. To our knowledge we have not come across any long bone having concomitant FD with secondary ABC in last 30 years.[14,15]

Our patient displayed with a greatly uncommon pathology of concomitant polyostotic FD with at the same time ABC of the femur bone. In writing, there exists numerous cases of polyostotic FD, but as it were a modest bunch of FD cases with ABC. Eminently, surgical intervention and planning ought to be prioritized by symptomatic lesions to begin with, followed by asymptomatic lesions.[1]

The first case of secondary ABC with FD in long bone (Femur) was reported by RL Diercks et al in 1986 in a 23 years old female. This girl had polyostotic FD, in right lower limb. Above knee amputation followed by hip arthrodesis was eventually performed. 5 years later she developed sudden swelling of the stump, biopsy was done which showed ABC developed in association with FD.[15]

Previously one case of ABC in association with FD was reported in tibia (long bone) by Buraczewski at al in 1971.[13]

Publication year	Author	Patient details	Bone involved
1971	Buraczewski at al	F/28	3rd Rib, Tibia
1971	Buraczewski at al	M/26	Mandible
1973	Oliver et al	F/20	Mandible
1980	El-Deeb at al	M/19	Mandible
1986	Branch CL,	F/9	Parietal / Frontotemporal
1986	RL Diercks	F/23	Femur
1986	Branch CL,	M/19	Parietal
1988	Yokoyama	M/29	Parietal, occipital
1989	Rappaport ZH,	M/25	Occipital
1994	Wojno KJ,	F/14	Temporal, middle cranial fossa
1994	Wojno KJ,	M/40	Frontal
	Same above	F/22	Maxillary and ethmoid sinus
	Same above	M/19	Mandible
	Same above	M/22	Sphenoid
1995	Lucarelli et al	M/19	Orbit
1997	Arden et al	F/19	Mandible, ethmoid, sphenoid
1998	Haddad et al	F/ 6	Skull base
1998	Saito et al	M/11	Nasal bone and skull base
2001	Suzuki et al	M/23	Maxillary sinus
2002	Itshayek E,	M/19	Occipital
2002	Yuen et al	M/22	Tempora, orbital, frontal and sphnoid
2002	Pasquini et al	M/5	Skull base
2004	Lin WC,	M/18	Frontal
2005	Mattei TA,	F/19	Occipital
2005	Iseri et al.	F/35	Occipital
2006	Malik et al	M/13	Anteriorcranial fossa, nasal cavity and orbit
2008	Skladzirerin et al	M/16	Nasal cavity, paranasal sinuses, orbit, cranial fossa
2010	Lee JW,	F/18	Frontoparietal
2010	Hadidy et al.	M/15	Frontal bone of skull

2011	Terkawi et al.	F / 7	Sphenoid bone & ethmoid bones
2011	Sankaranarayanan et al.	F/ 6	Maxillofacial
2011	Salmasi et al	M/16	Mandble, sphenoid, sella turcica, clivus, nasal cavit
2013	Manjila et al	M/10	Nasal bone, Skull base
2014	Lee et al	M	Zygomatic bone orbit and maxilla
2015	Hnenny et al.	F/28	Nasal cavity and skull base
2015	Moorthy et al	F/19	Ethmoid, frontal, maxillary and sphenoid
2015	Ngo et al	F/15	Sphenoid and cavernous sinus
2016	Urgun et al.	F/14	Skull base - C2 vertebra
2016	Elluwuch et al	F/16	Parietal
2016	Dani et al	M/23	Frontal and ethmoid
2017	Gotmare et al.	M/ 8	Maxillofacial
2017	Brik et al	M/23	Parietal
2017	Yollu et al	M/27	Paransanal and maxillary sinus
2018	Kaloostian et al.	M/ 20	Skull and skull base, rib, and humorous
2018	HSLee	25/F	Frontal, Sphenoid and ethmoid bone
2019	LH Rau	M/11	Zygomatic bone

On literature review we came across 46 such cases involving different bones in the body, mostly occurring in the skull bones. Only 2 cases with long bones, accounting about 4.3% of the cases with patient presenting with wide variety of signs and symptoms but mostly with pain complain in the respective body part involved with the disease.

Conflict of Interest: None

Grants/Funding: None

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