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Case Study: A rare case report of 39 years old Female with Polyostotic Fibrous Dysplasia

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ABSTRACT

Fibrous dysplasia (FD) is a rare bone disorder in which the affected bone is replaced by abnormal scar-like (fibrous) connective tissue. This abnormal fibrous tissue makes the bone weak, abnormally fragile and prone to fracture. Pain may occur in the affected areas. This condition was first described in 1942 by Lichtenstein and Jaffe, is also referred as Lichtenstein-Jaffe disease. Anyone can have FD and it can be diagnosed at any age. FD can affect one bone (known as monostotic FD), multiple bones (known as polyostotic FD) or the entire skeleton (known as panostotic FD). The treatment of FD depends on diagnostic testing, number of bones affected and location of affected bones. In some cases, polyostotic fibrous dysplasia can be treated with a class of drugs called bisphosphonates, which prevent bone loss and can reduce pain. Here presenting a case study of 39yrs female with Polyostotic Fibrous Dysplasia.

Key words: Fibrous dysplasia, Monostotic, Polyostotic, Panostotic, Bisphosphonates

INTRODUCTION

Fibrous dysplasia (FD) is a benign (noncancerous) bone condition in which normal bone is replaced by abnormal fibrous tissue. The severity of this disorder varies from one individual to other. Researchers believe that the disorder is caused by a mutation in a gene called *GNAS1*. Improper differentiation of osteoblasts due to mutation of the *GNAS1* gene is believed to contribute to the development of FD. FD can affect one bone (known as monostotic FD), multiple bones (known as polyostotic FD) or the entire skeleton (known as panostotic FD). Pain caused by fibrous dysplasia usually begins as a dull ache that worsens with activity and lessens with rest. Diagnosis is made by collecting general health and medical history, X-ray shows the ground-glass-like appearance of fibrous dysplasia, MRI or CT scan to further evaluate the lesion, bone scan, biopsy may be necessary to confirm the diagnosis of fibrous dysplasia. Bisphosphonates are given to treat Polyostotic FD which decreases the activity of cells that dissolve bone. They are available in oral form or intravenous (IV) infusions.

CASE REPORT

A 39 yrs old female presents in the OPD of orthopedic Surgery, Srinagar Garhwal on 09/July/2022 with chief complaints of lower back pain, chest pain, hip and legs pain. She reported that she had these complaints with less severity since 3yrs. She is a known

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case of hypothyroidism since six months. According to her she was seen for similar symptoms previously at her primary care physician and was treated with Tab. Calcium and analgesics. This management did not improve her symptoms and she has gradually worsened with time. An old X – ray revealed multiple osteolytic lesions in femur and tibia. Doctor advised opinion from oncosurgeon/ physician. She went Dehradun city for further opinion and met Orthopaedic surgeon on 14/ July/2022. She was advised X Ray of knee Joint AP/Lateral [Fig.1], X Ray of Pelvis [Fig. 2], routine investigations such as complete hemogram, RBS, albumin, globulin, total protein, serum bilirubin, SGOT, SGPT, ALP, Blood urea, serum creatinine, serum uric acid, CRP – Reactive protein, 25 – Hydroxy vitamin –D, serum Protein Electrophoresis, TSH and Urine examination. All parameters were within normal limits except ALP, serum uric acid, CRP – Reactive protein, 25 – Hydroxy vitamin–D and TSH. ALP was raised to 126U/L, serum uric acid was raised to 6.4mg/dl, CRP was raised to 19.0mg/L, 25 – Hydroxy vitamin –D was found deficient i.e.

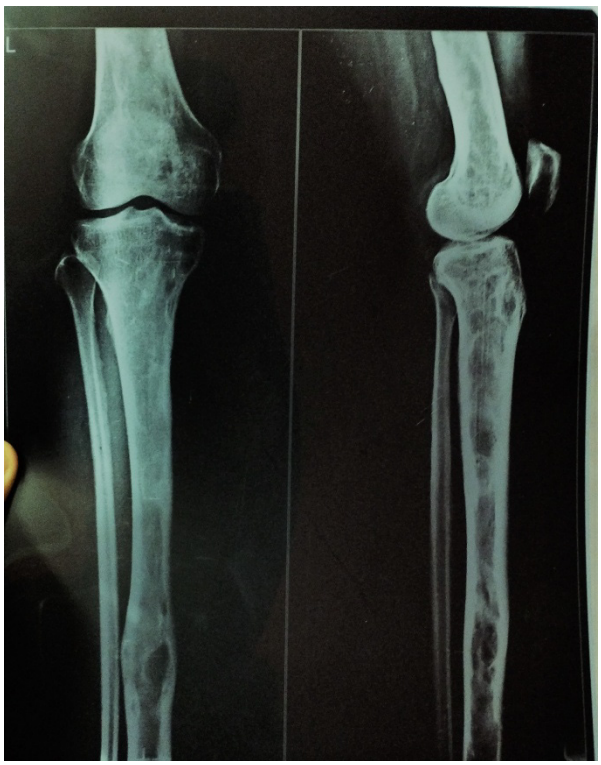


Fig 1: X - Ray Knee joint AP/Lateral 14/July 2022

18.0 ng/ml and raised TSH of 4.80 µlU/ml, mild elevation of alpha 1 and beta 1 regions was observed in Serum Protein electrophoresis [Fig. 3]. MRI of left lower limb showed Multifocal intramedullary T2/ STIR hyperintense enhancing lesions with surrounding sclerosis involving the femur and tibia causing medullary expansion, endosteal scalloping and cortical thickening. Findings may be secondary to chronic osteomyelitis. After getting the reports doctor suggested to take opinion from higher centre to confirm the diagnosis.

On 16/July/2022 she went to OPD of Department of Orthopaedics at tertiary hospital of Dehradun. Clinical examination done by the doctor revealed tenderness over middle third tibia in left leg. Doctor advised for investigations i.e., Serum ALP, Calcium, Phosphorus, Parathyroid hormone, ESR and CRP. All parameters were within normal limits except CRP which was raised to 11.31mg/L. X- Ray Chest PA view, X- Ray of both knee with thigh: AP and lateral view, X – ray of both knee with leg: AP and lateral view were advised. X- Ray Chest PA view showed normal study. X- Ray of both knee with thigh: AP and lateral view revealed Left femur showing multiple lytic areas with sclerosis- ? Osteomyelitis?? Fibrous dysplasia. Right knee with thigh appears to be normal. X – Ray of both knee with leg: AP and lateral view showed Lytic areas with sclerosis seen in mid shaft of left Tibia - ? Osteomyelitis?? Fibrous dysplasia. Right knee with leg appears to be normal.

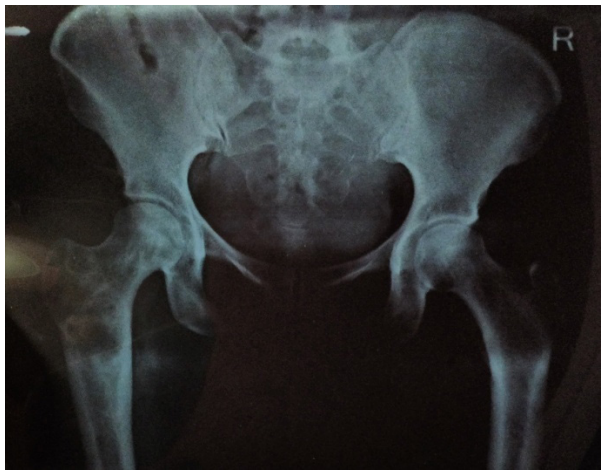
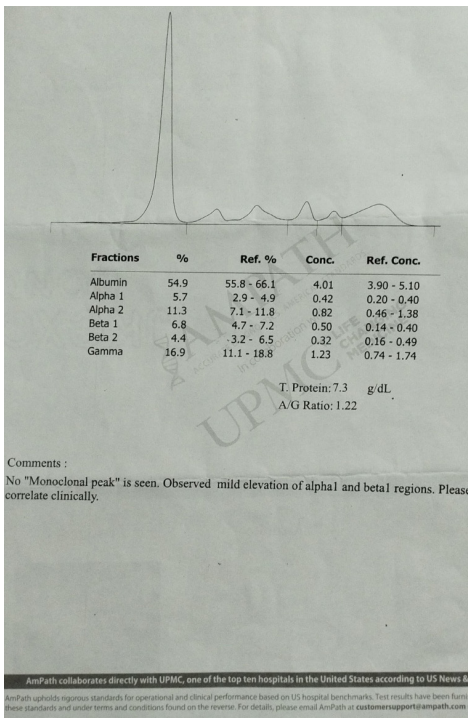


Fig 2. Pelvis with both hip joint 14/July/2022



Fractions	%	Ref. %	Conc.	Ref. Conc.
Albumin	54.9	55.8 - 66.1	4.01	3.90 - 5.10
Alpha 1	5.7	2.9 - 4.9	0.42	0.20 - 0.40
Alpha 2	11.3	7.1 - 11.8	0.82	0.46 - 1.38
Beta 1	6.8	4.7 - 7.2	0.50	0.14 - 0.40
Beta 2	4.4	3.2 - 6.5	0.32	0.16 - 0.49
Gamma	16.9	11.1 - 18.8	1.23	0.74 - 1.74

T. Protein: 7.3 g/dL
A/G Ratio: 1.22

Comments :
No "Monoclonal peak" is seen. Observed mild elevation of alpha1 and beta1 regions. Please correlate clinically.

Fig.3. Serum protein electrophoresis

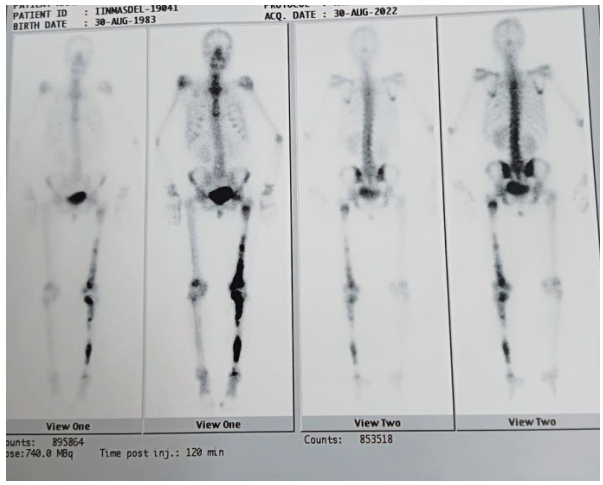


Fig.4. Skeletal Scintigraphy

After seeing the reports doctor decided to perform J Needle open biopsy from left leg on 24/July/2022. One day prior she was admitted in the hospital for Pre anesthetic check up and few investigations were done i.e., PT/INR, 12 lead ECG which were found normal. Histopathology report of Left tibia Mid shaft biopsy showed few necrotic bony bits with mild neutrophilic infiltrate. Suggestive of acute osteomyelitis. Furthermore, bone marrow examination was advised by the doctor to rule out involvement of blood cells which was performed on 10/August/2022 and the report was normal. On 17/August/2022 doctor started the treatment with Tab Alendronate Sodium 70mg, once in a week for 4 weeks and instructed not to lie down for half an hour after taking the dose. Tab Shelcal 500mg BD x 1 month was also started.

As she was not satisfied with the treatment so went to OPD of Ortho Department, Tertiary care and Research Centre, Delhi on 29/8/2022. Doctor advised for whole body bone scan. Skeletal Scintigraphy done with 20mCi of 99mTc-MDP intravenously and Scintiphotographs taken in 3 phases [Fig. 4]. In the first phase (Flow phase) immediate post injection there was increased flow in the region of Left femur and Left tibia. Second phase (Blood pool phase) 5 min post injection revealed increased pooling in the region of Left femur and Left tibia. Third phase (Delayed phase) 3 hrs post injection showed increased tracer uptake in the region of Left femur and Left tibia. Rest of the axial and appendicular skeleton showed physiological tracer uptake. After report she was diagnosed with Polyostotic Fibro Dysplasia. Doctor advised Inj. Zoledronic acid to be given in OT after adequate hydration and Serum Calcium, Phosphorus, ALP and KFT to be done. On 3/9/2022 Inj Zoledronic acid was given and she was prescribed Tab CCM – OD, Sachet Vit D3 60K IU/week and review after three weeks. On 26/9/2022 she went for follow up and doctor advised Serum Calcium, Phosphorus, ALP and KFT to be done and Inj. Zoledronic acid to be given in OT tomorrow. On 27/9/2022 Inj. was given in 50 ml NS over 1 – 2 hour slowly with 500ml NS pre hydration. Doctor advised Tab. Arkbone CT 1 x BD and Cap Lumia 60K weekly for 4 weeks. Tab PCM 650 mg SOS and review after 3 weeks in Ortho OPD with fresh X – ray.

Followed by two dose of Inj. Zoledronic acid on 19/10/2022 doctor prescribed Tab. Alendronate 70mg weekly and instructed her not to sit or lie down for one hour after taking medicine. Tab Diclo 50mg BD for two weeks, Tab. Calcium 500mg OD, Tab Pantoprazole

40mg OD were prescribed and told to review after one month. She is feeling relieved with the treatment as severity of pain has been decreased.

DISCUSSION

FD was first described in 1942 by Lichtenstein and Jaffe, is also referred as Lichtenstein-Jaffe disease. FD is a benign intramedullary bone lesion in which the normal bone marrow is replaced by abnormal fibro-osseous tissue. This disorder can involve a single bone (monostotic) or multiple bones (polyostotic). The severity and specific symptoms present in Polyostotic fibro dysplasia (PFD) differs from one person to another. Patient with PFD complaints of bone pain, bony tenderness, bony crookedness, endocrine disturbance and dermatological complications. Bone pain and tenderness was present in the client and she had hypothyroidism. The exact cause of FD is not fully understood. Researchers believe that the disorder is caused by a change (mutation) in a gene called GNAS1. In present client there is no family history of PFD or personal history of injury or accident. The disorder is diagnosed at early age but in few cases it goes undiagnosed and identified at adult age. Diagnosis can be made by detailed patient history, clinical evaluation, X-Ray, CT scan, MRI, Bone biopsy, Bone scan. FD may be diagnosed incidentally when receiving an x-ray for another reason. In patient also it was diagnosed accidentally when doctor prescribed X-Ray for pain in legs. The treatment to be given according to the symptoms apparent in patient. Individuals with PFD have been treated with drugs known as bisphosphonates such as alendronate. These drugs reduce bone turnover by inhibiting bone resorption. Calcium and vitamin D may be given along with the drug. Patient also treated with two doses of Inj. Zoledronic acid followed by Tab. Alendronate 70mg weekly with Tab Diclo 50mg BD for two weeks, Tab. Calcium 500mg OD and Tab Pantoprazole 40mg OD.

CONCLUSION

FD is a benign, slowly growing bone disorder in which abnormal fibrous tissue develops in place of the normal bone. The main complications of FD are pathologic fracture, secondary aneurysmal bone cyst formation, and rarely malignant change. So proper understanding, investigations and management to diagnose such types of cases in initial stages is of prime importance to prevent any kind of complications. We report this case, of polyostotic FD as it is the rarest type and has very less percentage of occurrence of just about 20–25%, it is our attempt to add further knowledge to the literature.

Declaration of patient consent

The author certifies that she had obtained all appropriate patient consent forms. In the form the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understand that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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Conflicts of interest: There are no conflicts of interest.

Ethical clearance: Ethical committee of College of Nursing, Dehradun.

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