Medical and Research Publications
International Open Access

Case Report **Journal of MAR Case Reports (Volume 6 Issue 6)**

Dramatic Response in Polyostotic Fibrous Dysplasia by Pamidronate

Abid Rasool *[1], Ajaz qadir [2], M.A.Ganie [3], Tajali Sahar [4], Omair Shah [5]

1. Senior Resident, Department of Endocrinology, Sher-I-Kashmir Institute of Medical Sciences.
2. Senior Resident Department of Endocrinology, Sher-I-Kashmir Institute of Medical Sciences.
3. Professor, Department of Endocrinology, Sher-I-Kashmir Institute of Medical Sciences.
4. Phd, Scientist B, Department of Endocrinology, Sher-I-Kashmir Institute of Medical Sciences.
5. Senior Resident Department of RADIODIAGNOSIS, Sher-I-Kashmir Institute of Medical Sciences.

Corresponding Author: Abid Rasool, Senior Resident, Department of Endocrinology, Sher-I-Kashmir Institute of Medical Sciences.

Received Date: January 27, 2023

Published Date: February 10, 2023

Fibrous dysplasia is uncommon and is picked up incidentally when patient complaints fractures, or deformity or bony pain. It is divided into monostotic and polyostotic forms. We describe a 16-year-old female who started with deformity of right ring and little finger following trivial trauma at the age of 4 years with bony pains. Historically, did not have any features suggestive of precocious puberty. Investigations were normal but skeletal survey was showing multiple hyperlucent bony cyst involving right upper and lower limb. She was diagnosed polyostotic fibrous dysplasia and started on injection pamidronate 90mg for 3 days and repeated after six months. She showed dramatic response in pain and imaging also showed improvement.

Introduction

Fibrous dysplasia (FD) is a sporadic, tumour-like, benign skeletal disorder originally described by Lichtenstein in 1938 [1] and by Lichtenstein and Jaffe in 1942 [2]. FD is often asymptomatic. However, FD often leads to bone pain, deformities, or pathological fractures. FD is classified into two types: monostotic FD affects only a single bone, and polyostotic FD presents the involvement of multiple bones. While small monostotic FD lesions occur more frequently than polyostotic FD, they result in fewer complications. Conversely, polyostotic lesions tend to enlarge and result in severe complications [3, 4]. We describe a case of polyostotic fibrous dysplasia who responded dramatically by pamidronate injection.

Case

16 years female fourth in birth order product of non-consanguineous marriage born by full term vaginal delivery with uneventful perinatal and postnatal history with normal developmental milestones, presented history of bony pains and asymmetrical swelling of right ring and little finger along with swelling around 4th and 5th metacarpal and deformity of right forearm post trivial trauma with fracture 10 years ago. It was associated with pain, redness and limitation in mobility. There is also history of trivial trauma of right leg, followed by fracture and swelling and limp on walking six years ago. There is no family history of similar disease. Patient achieved menarche at the age of 15 years. Clinical exam: SMR: A2B2P2, Height 163 cm (50-75th percentile), Weight 35 kg (3rd percentile). Deformity of right little and ring finger, bony hard in consistency, mild tenderness and redness. Deformity of right leg with circumferential swelling, hard in consistency, no tenderness or redness, Spine mild scoliosis, no

café lait spots or features of precocious puberty. Patient was admitted and on evaluation investigations revealed following.

Pre-treatment investigation							
Hb (mg/dl)	TLC (1000/micr/li)	PLT (1000/microlit)	Urea (mg/dl)	Creat (mg/dl)	Calcium (mg/dl)	Phosphorus (PO4) mg/dl	Alkaline phosphate(ALP)
13.8	5.4	185	28	0.36	9.1	4.56	408
Albumin (ALb)	TSH	T4	T3	FSH IU/L	LH (IU/L)	iPTH (pg/ml)	Vitamin D (ng/ml)
3.66	3.77	7.02	1.02	8.22	9.41	29.8	17.6
Post treatment investigation							
Hb	TLC	PLT	Urea	Creat	Calcium	PO4	ALP
12.6	4.3	150	33	0.34	8.8	3.7	289

USG pelvis: uterus 6x 2.2x 3 cm in size, ET =7mm, rest normal.

Skeletal survey revealing multiple hyperlucent bony cystic lesions at various sites including right upper limb, right lower limb suggestive of polyostotic fibrous dysplasia.

Bone scan was also consistent with polyostotic fibrous dysplasia. Patient was subjected to Pamidronate 30mg/ day for 3 days on 24/5/21(1ST cycle). Patient is now in our follow up and post 1st cycle there is marked improvement in pain from initial VAS (visual analogue scale) 8/10 to 5/10.

DXA scan:

	L1-L4	Femoral Neck	Total Hip
Z score	-3.3	-1.5	-1.0

Fig 1 showing deformity of right ring and little finger

Fig 2. Showing no Café lait spots

Pre-treatment imaging

Fig 3. Showing multiple bone cysts over hand and arm

Fig 4. Showing multiple cystic lesions over right leg

Post treatment imaging

Fig 5. Showing decrease in number of bone cysts and new bone formation

Discussion

Fibrous dysplasia (FD)is very rare disease which is often associated with bone pain, deformity and fractures. Histologically there is excessive fibrous tissue in bone marrow due to mutated osteoblasts and high number of osteoclasts and evidence of bone resorption [5]. Thus, increased bone resorption may play a role in development of fibrous dysplasia.

This justifies use of bisphosphonates as treatment for FD. Mostly commonly pamidronate has been used as a bisphosphonate and promising result has been obtained in terms of improvement in bone pain.

Liens et al [6] studied nine patients with symptomatic and severe fibrous dysplasia who were treated with intravenous pamidronate (60 mg per day for 3 days every sixth month) and were followed up for as many as 4 years. Major effect they saw was decrease in bone pain.

Chapurlat et al [7] who assessed the long-term effects of intravenous pamidronate in patients with fibrous dysplasia. In their study, 20 patients (11 males and nine females; mean age, 31 years) received courses of 180 mg of intravenous pamidronate every 6 months (60 mg/day during 3 days by infusion). The mean duration of follow up was 39 months (range, 18–64 months). Severity of bone pain, number of painful skeletal sites per patient, radiographs of all involved areas, serum alkaline phosphatase, fasting urinary hydroxyproline, and urinary Type I collagen C-telopepide were assessed every 6 months. The severity of bone pain and the number of painful sites seemed to be reduced significantly. All biochemical markers of bone remodelling were lowered substantially. The authors reported a positive radiographic response in nine patients because they observed refilling of osteolytic lesions. They suggested that intravenous pamidronate alleviated bone pain, reduced the rate of bone turnover as assessed by biochemical markers and improved radiologic lesions of fibrous dysplasia

A recent report by Pfeilschifter and Ziegler [8] reported the results of treatment of eight patients (three with fibrous dysplasia and five with McCune-Albright syndrome) with intravenous infusions of 60 mg of pamidronate. All patients had significant improvement of pain whereas two patients also had a reduction in size of some of the osteolytic lesions.

In our case report there was reduction in bone pain and improvement in radiology. So, it may be suggested to use bisphosphonate especially pamidronate in fibrous dysplasia for pain management and fracture reduction.

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