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Clinical outcome of monosteotic fibrousdysplacia treated with intramedullary nailing and bisphosphonate therapy.

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ABSTRUCT-

Background-Fibrous dysplasia of bone is an enigma with no proper guideline.Treatment currently consists of curettageand bone-grafting in an attempt to eradicatethe lesion and to prevent progressive deformity. No definite criteria have been established to identify patients at high risk of presenting pathological fracture . Clear guidelines for orthopaedic management of fibrous dysplasia.

In the current study, the combination bisphosphonate therapy diminished pain, prevented fractures, resolution of fibrous dysplasia lesions.

Material and method –

At Medical college Kolkata,Ten patients with monostotic fibrous dysplasia in lower extremities treated between 2017 to 2021 were included in the study. All patients underwent full skeletal survey followed by core needle biopsy with the help of MRI and C-ARM guidance ,after confirmation , Closed intramedullary nail without reaming was used in all cases. Bone grafting was not performed. zoledronic acid, given intravenously at the dose of 4 mg every 6 months.

Patients were allowed full weight bearing on the affected extremities on the second postoperative day

Result -

We get good outcome and .Clinico-radiological Improvement of all cases.

Conclusion-As a result of this study, we believe intramedullary fixation can be performed successfully. in cases of monostotic fibrous dysplasia with adjuvant bisphosphonate therapyproven increase functional activity and controle pain. This will avoid problems that may occur following pathological fractures.

Keywards-monosteoticfibrousdysplacia, intramedullary nailing, zoledronic acid.

I. INTRODUCTION-

Fibrous dysplasia (FD) is a challenge to the orthopaedicsurgeon .Fibrous dysplasia, first identified by Lichtenstein in 1938 (11), is an anomaly characterised by widening of of the affected bone with cortical thinning and presence of fibro-osseous tissue inside the bone. There may also be areas with islands of cartilage or cysts, and some lesions may be expansile. It may present under a monostotic or polyostotic form. Fibrous dysplasia of the proximal femur is difficult to treat due to the varied presentations like pain, pathological fractures, severe deformity, and high chances of recurrence .Lesions have a tendency to recur and may result pathological fractures following curettage and grafting (5) . No definite criteria have been established to identify patients at high risk of presenting pathological fractures (1). Clear guidelines for orthopaedic management of fibrous dysplasia in long bone have not been established.

We reviewed published data on the treatment of FD with bisphosphonates, calcium, vitamin D, and phosphorus.

We want to present our results with intramedullary nailing along with intravenous zoledronic acid 4mg in every 6 months, in10

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patients with monostotic fibrous dysplasia, pain increasing with movement. To our knowledge, no previous study has focused on intramedullary nailing of the proximal femur along with bisphosphonate therapy over monostotic fibrous dysplasia in symptomatic patients.

The aim of our study was to analyze the various presentationsof fibrous dysplasia in long bone like pain, fractures, shepherd crook deformity, and describe the results of the various treatment modalities for the same.

II. REVIEW OF LITERATURE:

FD is an orthopaedic condition with a wide spectrum of presentation. The treatment of the dysplastic lesions in the proximal femur region is still somewhat unclear, and varies widely. Fibrous dysplasia of the bone can present as three clinical forms: monostotic, polyostotic and as a part of a McCune-Albright
syndrome.
Lichtenstein (11) is credited with having coined the term fibrous dysplasia in 1938 ; in 1942,
Lichtenstein and Jaffe (12) reviewed all known cases of this entity. Those authors established that fibrous dysplasia of bone was a distinct pathological and clinical condition. Fibrous dysplasia may occur due to a failure in remodelling of primitive bone into mature lamellar
bone, which negatively affects the mechanical properties
of the affected bone. Thus pain, deformities, and pathological fractures may occur.Fibrous dysplasia has traditionally been divided into three clinical forms : monostotic, polyostotic
and endocrinopathic (McCune-Albright syndrome) (7). It is generally accepted that monostotic lesions are easier to treat, are associated with better
outcomes, necessitate fewer operations and result in fewer fractures (4).
Healing after pathological fractures in dysplastic bones is comparable with that of normal bone.
However, the callus includes dysplastic bone tissue (2). The lesion persists despite healing of the
fracture. The accepted principle in the treatment of lesions that are painful or at risk for fracture, even if asymptomatic, is curettage and grafting (6). However, according to our review of the literature, it is uncertain whether this form of treatment offers a definitivesolution (7). It has also been reported that curettage or biopsy of an isolated lesion may predisposethe bone to pathological fracture or progression of the lesion (5). There is no accurate indication of therate of success of curettage and bone grafting. In their study on patients with fibrous dysplasialocalised in the neck of the femur, Guilleet al (6) have shown that the lesion was not eradicated withcurettage and grafting, and the bone was further weakened due to deformation of the trabecularstructure in dysplastic bone as a result of curettage.In the present series, we did not perform curettageand bone grafting.
Resorption and recurrence secondary to grafting after curettage are other problems. Guilleet al (7) have shown in their study that all cancellous or cortical grafts they used, in addition to autogenous fibular strut grafts, were resorbed. In addition,according to DiCaprio and Enneking (1), cortical grafts are more durable compared to cancellous grafts, as they are only partly replaced by dysplastic
host bone : only their osteonal portion (about 50% of the graft) is replaced by dysplastic bone,
whereas the interstitial lamellae are not replaced and persist .

Therefore, we recommend prophylactic intramedullary fixation in patients with monostotic fibrous dysplasia. This prophylactic therapy avoids complications such as delayed union and deformities following fracture. A vascularised fibula has been used in some cases following fracture (10). It appears more reasonable to take the necessary steps to prevent fracture, considering the technical difficulty, delayedweight bearing, risks of graft resorption and re-fracture in addition to high costs, if the affected bone is not strengthened and fractures.

III. METERIALS AND METHODS-

Study Area:
MEDICAL COLLEGE, KOLKATA

Study Period:
November 2017 to OCTOBER 2021

Sample Size ;10 patients total.

Sample Design:
1) Patient Selection :
The study will be conducted among the adult patients attending Orthopaedics out-patient department with fibrous dysplacia
2) Inclusion Criteria :

- Impending pathological fracture.
- Pathological fracture of long bone

3) Exclusion Criteria :

- Renal impairment
- Previously surgical intervention done
- Multiple comorbidity

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Study Design:
Institution based prospective observational study.

Study Tools:
- Roentgenogram
- General internal fixation instruments for long bone.

Parameters to be studies:
Primary outcome
- bone healing and remodelling

Operative details
- duration of operation.
- amount of blood loss.

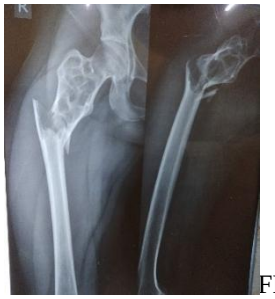
Perioperative complications
- radiological evaluation of bone loss and fracture

Post operative complications
- infection.
- range of movement

Method- All patients underwent full skeletal survey followed by renal function test,in case of pathological fracture we will perform core needle biopsy with the help of MRI and C-ARM guidance.
Patients who had been diagnosed with fibrous dysplasia with functional pain or pathological fractures were included in the study and undergone biopsy confarmation and nailing, whereas patients who had no functional pain , had been incidentally diagnosed were excluded from the study.

After taking written consent form every patient and their family member we had perform core needle biopsy confirmation for all cases.
Under c arm guide we had performed intramedullary nail -titenium ,preferably 3dr generation long gamma nail for better fixation of proximal femur ,as it's a weight bearing bone.
We put all patient under medical therapy of intravenous zoledronic acid 4mg dissolved with 100ml NS. Over 30 minutes, In every 6 months.
We avoid localcurratage and bone grafting because it FD is notorious for its recurrence and local fibrous tissue prevent the activity of bone grafting.As well as it's an open procedure ,hence post operative morbidity is higher. Resorption and recurrence secondary to grafting after curettage are other problems. Reaming was not used prior to nailing in our patients, as it was deemed unnecessary for nailing of non-fractured long bones, added to the fact that it might have contributed to weakening the bone to some extent
we plan for adjuvent medical therapy as we don't use bone graft.radiological follow through we want to see the effect of zoledronic acid over pathologic fibrous tissue ,can't undergo calcification in normal process.
Functional pain, size of the lesion on radiographs and stability of prophylactic fixation were evaluated in follow-
up visits every 6 months. A visual analogue scale (VAS) was used in assessing functional pain.

Funding/sponsor: None
Conflict of interest statement: None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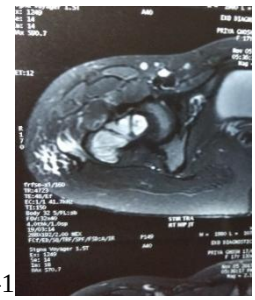
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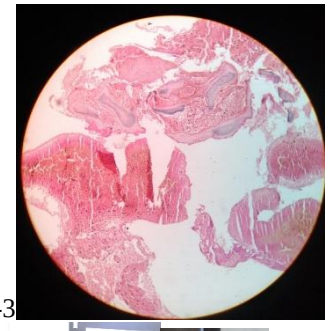
FIG-3


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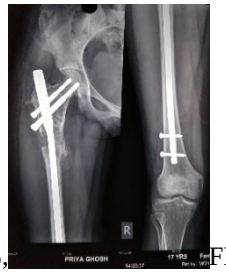
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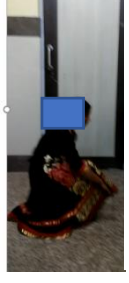
FIG-6

fig-7

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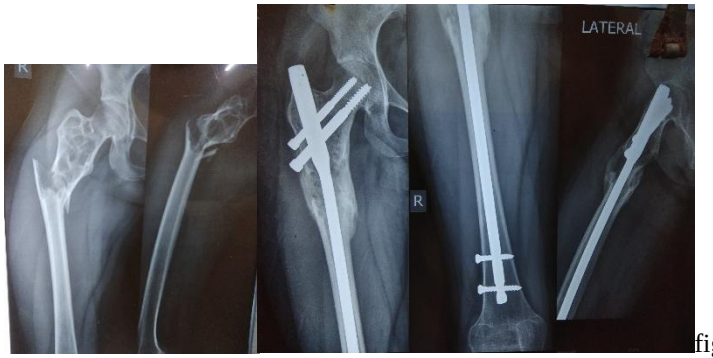
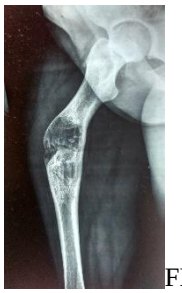


fig-8

Fig-1 pre operative xray.,fig-2-MRI . fig-3 Core needle biopsy,fig- 4-histopatho

Fig-5-post op fig-5,8 -post zoledronic acid therapy 6months ,1 year

Fig-7 post operative rehav pain less squatting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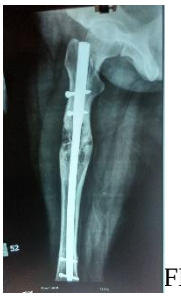
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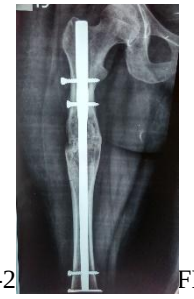
FIG 3

FIG-1 -PRE OP IMPENDING PATH #.
FIG-2,3 – post op
Healing of osteolytic area

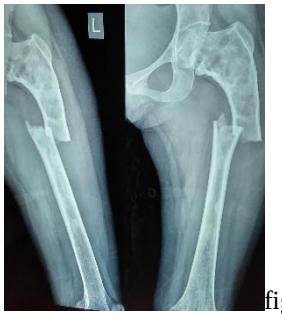

fig-A
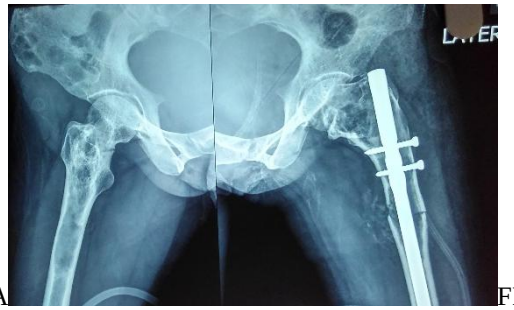
FIG-B FIG A-PRE OP PATH # & B- POST OP

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GRAND CHART FOR DATA ANALYSIS

cases	sex	Age years	site	Indication Of intervention	Follow up(months)	Preop VAS	POST OP VAS
1	M	23	PROXIMAL FEMUR	PAIN. pathological #	32	5.8	2.4
2	F	30	PROXIMAL FEMUR.	Pathological#	30	4.9	1.7
3	F	19	PROXIMAL FEMUR	PAIN.	26	6.4	1.8
4	F	29	PROXIMAL FEMUR	PAIN.	32	5.7	2
5	M	23	PROXIMAL FEMUR	PAIN.	27	4.9	1.6
6	F	23	PROXIMAL FEMUR	PAIN.	29	6.1	2.5
7	F	30	PROXIMAL FEMUR	Pathological#	25	5.2	3
8	M	24	PROXIMAL FEMUR	PAIN.	25	5.3	2.5
9	F	25	PROXIMAL FEMUR	PAIN.	28	5.7	2.9
10	M	26	PROXIMAL FEMUR	Pathological#	29	4.5	1.8

During screening of the subjects, all potential subjects who fulfilled the study selection criteria will be informed by the investigator, verbally, in vernacular, about the study in details (including the rationale, aims and objective of the study, study related procedures, potential discomfort and benefits of participation). Following this copy of Informed Consent Form and Patient Information Sheet will be provided to the subjects and they will be requested to go through them. The investigator will also answer any study related queries raised by the subject. After the above mentioned procedure only those subjects who are willing to participate will be asked to sign and date the written informed consent form expressing their voluntary participation in the trial. All study related activity will start only after such consent is obtained.

The investigator will ensure the confidentiality of the study participants. The case record forms, study documents and biological samples collected will be untitled and anonymous. All study related documents will be kept under the strict supervision of the principal investigator at a designated place in Medical College, Kolkata. Sterility and universal precaution will be maintained during the process.

Citation of references in the text: Cited

Case report form/ data collection form: Attached
Informed consent documents: Attached

IV. DISCUSSION

FD is an orthopaedic condition with a wide spectrum of presentation. The treatment of the dysplastic lesions in the proximal femur region is still somewhat unclear, and varies widely. Fibrous dysplasia of the bone can present as three clinical forms: monostotic, polyostotic and as a part of a McCune-Albright syndrome.
Lichtenstein (11) is credited with having coined the term fibrous dysplasia in 1938 ; in 1942,

Lichtenstein and Jaffe (12) reviewed all known cases of this entity. Those authors established that fibrous dysplasia of bone was a distinct pathological and clinical condition. Fibrous dysplasia may occur due to a failure in remodelling of primitive bone into mature lamellar
bone, which negatively affects the mechanical properties
of the affected bone. Thus pain, deformities, and pathological fractures may occur.Fibrous dysplasia has traditionally been divided into three clinical forms : monostotic, polyostotic
and endocrinopathic (McCune-Albright syndrome) (7). It is generally accepted that monostotic lesions are easier to treat, are associated with betteroutcomes, necessitate fewer operations and result in fewer fractures (4).
Healing after pathological fractures in dysplastic bones is comparable with that of normal bone.
However, the callus includes dysplastic bone tissue (2). The lesion persists despite healing of thefracture. The accepted principle in the treatment of lesions that are painful or at risk for fracture, even if asymptomatic, is curettage and grafting (6). However, according to our review of the literature, it is uncertain whether this form of treatment offers a definitive
solution (7). It has also been reported that curettage or biopsy of an isolated lesion may predisposethe bone to pathological fracture or progression of the lesion (5). There is no accurate indication of therate of success of curettage and bone grafting. In their study on patients with fibrous dysplasialocalised in the neck of the femur, Guilleet al (6) have shown that the lesion was not eradicated withcurettage and grafting, and the bone was further weakened due to deformation of the trabecularstructure in dysplastic bone as a result of curettage.In the present series, we did not perform curettageand bone grafting.
Resorption and recurrence secondary to graftingafter curettage are other problems. Guilleet al (7) have shown in their study that all cancellous or cortical grafts they used, in addition to autogenous fibular strut grafts, were resorbed.
In addition,according to DiCaprio and Enneking (1), cortical grafts are more durable compared to cancellous grafts, as they are only partly replaced by dysplastic
host bone : only their osteonal portion (about 50% of the graft) is replaced by dysplastic bone,
whereas the interstitial lamellae are not replaced and persist .The size of the lesion may change with skeletal growth ; however it is difficult to differentiate whether this is secondary to skeletal growth or to progression of the lesion (4). Since fibrous dysplasia is a genetic disorder which is not curable, the treatmentmodality should be long lasting. No definite criteria are available to state in which cases pathological fracture will occur (1). In the multicentric study of the European Paediatric Orthopaedic
Society, fractures had occurred in 47% of patients with monostotic fibrous dysplasia (8).
Therefore, we recommend prophylactic intramedullary fixation in patients with monostotic fibrous dysplasia. This prophylactic therapy avoids complications such as delayed union and deformities following fracture. A vascularised fibula has been used in some cases following fracture (10). It appears more reasonable to take the necessary steps to prevent fracture, considering the technical difficulty, delayed
weight bearing, risks of graft resorption and re-fracture in addition to high costs, if the affected bone is not strengthened and fractures.
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V. CONCLUSION-

As a result of this study, we believe intramedullary fixation can be performed successfully. in cases of monostotic fibrous dysplasia with adjuvant bisphosphonate therapy proven increase function activity and control pain.
This will avoid problems that may occur following pathological fractures.

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