**Introduction**

Fibrous dysplasia is a benign intramedullary disease usually present in childhood, that is, monostotic or polyostotic form, with the former being the most common form ranging from 75% to 80% [1]. It is not a true bone tumor, rather a developmental disorder with the proliferation of primitive fibrous tissue within the medullary canal, which on expansion causes thinning of the cortex [2]. True incidence and prevalence of fibrous dysplasia are difficult to estimate; though not rare, it accounts for 5–7% of all benign bone lesions presentation at adolescence and early adulthood without any gender predilection [3].

**Case Presentation:** In this retrospective case series, six children aged 7–12 years with monostotic fibrous dysplasia with or without fracture were treated with different treatment modalities from 2015 to 2020. Extended curettage and bone grafting and stabilization were done with extramedullary implants such as locking plates and DHS. Autograft alone or combination with allograft was chosen according to size of lesion. Patients without fracture were treated with curettage and artificial bone graft substitute with hip spica. Patients were followed up to 12 months. Revised Musculoskeletal Tumor Society (MSTS) score at each follow-up and Toronto Extremity Salvage Score (TESS) at final follow-up.

**Results:** Mean fracture healing time was 14.8 ± 2.28 weeks in patients with fracture. Full weight-bearing was started at average 15.67 ± 2.94 weeks. One patient had shortening of 1 cm but none had any surgical site infection, loss of correction, or varus collapse more than 5°. Mean revised MSTS at 12 months was 24.2 ± 2.28 in patients with curettage, bone grafting, and internal fixation, while it was 27 in patients with curettage, grafting with hip spica. Mean TESS was found to be 90 ± 9.41 in internal fixation group, while it was 95 in patient with curettage, grafting with hip spica.

**Conclusion:** Treatment modality should be chosen wisely after analyzing pre-operative radiograph, lesion size, presence of pathological fracture, and each patient profile. Extramedullary fixation devices can be an alternate choice of implant in children and adolescents for monostotic fibrous dysplasia. Long duration follow-up and patient counseling should be done for recurrence of lesion and deformities.

**Keywords:** Monostotic fibrous dysplasia, pediatric pathological fracture, extramedullary device, allograft, autograft.
AP and lateral radiographs of pelvis with both hips and thighs were taken. All the patients underwent a skeletal survey. Polyostotic lesion patients were excluded from the study. The femurs were classified radiologically according to the classification by Zhang et al. [14]. At least one of the followingThere seems to have knowledge gap in the ideal treatment for fibrous dysplasia in pediatric patients as well as there remains controversy regarding the use of type of bone graft with or without fixation device. We treated six pediatric patients with monostotic fibrous dysplasia with different treatment modalities according to the size and type of lesion in our tertiary care center. We evaluated their functional outcome using the revised musculoskeletal tumor society scoring system (MSTS) [12] and TESS [13].

Preoperative evaluation
AP and lateral radiographs of pelvis with both hips and thighs were taken. All the patients underwent a skeletal survey. Polyostotic lesion patients were excluded from the study. The femurs were classified radiologically according to the classification by Zhang et al. [14]. At least one of the following

Figure 1: The figure shows institutional management protocol.
Figure 2: The image shows radiographs of patient no. 2 treated with locking plates and autograft (a) pre-operative image, (b) at 3-month post-operative, and (c) at 52 weeks follow-up shows complete healing without any varus collapse or implant failure.
Figure 3: The image shows radiographs of patient no. 1 treated with DHS and both autograft and allograft (a) pre-operative CT imaging shows a large void, (b) pre-operative X-ray, (c) at 3-month post-operative shows callus formation, and (d) at 2 weeks follow-up shows healing with no varus collapse or implant failure.
Figure 4: The image shows radiographs of patient no. 5 treated with curettage and ceramic bone substitute and hip spica cast without internal fixation (a) pre-operative X-ray, (b) at 3-month post-operative, and (c) at 60 weeks follow-up.
features had to be present to be considered as fibrous dysplasia (a) grayish ground-glass appearance, (b) endosteal scalloping, (c) shepherd crook deformity, (d) intramedullary expansile lesion with a smooth sclerotic margin, and (e) rind sign and parrot beak sign [14, 15, 16]. A pre-operative CT scan was done to evaluate (a) the location and size of the lesion, (b) extension of fracture line, and (c) reduced bone strength (thickness of remaining cortical bone <50% of original thickness on axial CT). As radiological features were highly suggestive of a benign pathology like fibrous dysplasia, pre-operative histological confirmation was not done in our study. However, post-operative histological study was done and confirmed features suggestive of fibrous dysplasia in all cases.

Surgical modalities

Two patients came to us with moderate to severe pain, limping, and difficulty in weight-bearing. Radiographs and CT scan showed that they had a Type 2 lesion with typical intramedullary location and ground-glass appearance suggestive of fibrous dysplasia [14]. They were advised curettage with bone substitute like G-BONE (synthetic granules of calcium hydroxyapatite in crystalline form) and post-operative hip spica cast for 8 weeks and then were given a hip orthosis. One of them (patient number 3; Table 1) was not willing for surgery and started on conservative measures like protected weight-bearing and calcium Vitamin D supplements. After 2 months, this patient returned with an insufficiency fracture at the femoral neck and was taken for curettage and bone grafting along with internal fixation with dynamic hip screw.

Four patients had a history of trivial trauma and presented with pathological fracture of proximal femur with lytic lesion. One of them had a previous history of pain around the hip, which was relieved after taking NSAIDs. Rest four patients did not have any previous significant history.

Patients with pathological fracture were placed in skin traction and medically optimized for surgery. Surgical plan was formulated and individualized for each patient with aim of (a) local control of disease by extended curettage with phenol and sending tissue for histopathological examination, (b) filling of void with bone graft/graft substitutes, (c) neutralization of fracture with extramedullary implants, and (d) correction of deformity of neck or proximal femur and achieving mechanical alignment. We chose a surgical plan according to the institutional protocol (Fig. 1).

Intraoperative curettage was done, and tissues were sent for histopathologic confirmation followed by extended curettage using 85% phenol and then washing it with 96% ethanol [17] followed by bone grafting. An appropriate extramedullary implant was selected, as we did not have access to growing nails. Two patients were fixed with dynamic hip screw and two with locking plates (one PHILOS plate and one distal femur locking plate) according to availability of implants. To get a better hold with the implant, we had to cross the physis in all of our patients. Small cavities (<60 cc) were filled up with autogenous cancellous iliac bone graft [18] (Fig. 2a-c). The larger cavities were filled with autogenous cancellous graft and then supplemented with freeze-dried allogenic cancellous bone graft after 20 min of rehydration with normal saline (Fig. 3a-d). Operative time, blood loss, and any other complications were recorded.

After care

Postoperatively, the patient without internal fixation (patient number 5) was kept on hip spica cast for 8 weeks and then cast
was removed and started on partial weight-bearing till radiological signs of healing (Fig. 4a-c).

For patients with pathological fracture and internal fixation, knee range of motion, ankle pumps, static quadriceps strengthening, and bedside mobilization were started immediately. After check dress, patients were discharged on an average of the 5th post-operative day with hip orthosis. Suture removal was done on the 15th post-operative day. Passive hip adduction and abduction were started after suture removal. Passive straight leg rising test (SLRT) followed by active-assisted SLRT was started at 3 weeks. Active SLRT was started at 4 weeks. After achieving satisfactory hip and knee ROM, patients were mobilized non-weight-bearing with walker and hip orthosis. Patients were kept non-weight-bearing for at least 12 weeks or radiological signs of healing whichever are earlier. Full weight-bearing was started after radiological confirmation for adequate callus formation. As we had to get better hold in the neck, we had to cross physis during internal fixation, we counseled the patient regarding limb length discrepancy.

Follow-up

All six patients were followed up at 1, 3, 6, and 12 months. At each follow-up, each patient was evaluated radiologically for fracture union, mechanical axis alignment and implant position, any varus collapse and clinically for pain, range of motion, shortening, development of any deformity, gait, and patient compliance toward the treatment protocol. All the data were noted and produced as revised MSTS scores. At the final follow-up of each patient, mean 54.33 ± 3.2 weeks (range, 52–60 weeks), TESS was calculated for the patient using the pre-determined questionnaires.

Results

Each patient was given a serial number according to the chronologic order of presentation (Table 1). Patient nos. 1, 2, 4, and 6 presented with pathological fracture. Patient 3 had failed conservative therapy, presented with insufficiency fracture at 2 months follow-up, and was later internally fixed with grafting. Patient no. 5 was treated with simple curettage and calcium hydroxyapatite granules. In the patients with pathological fracture, the mean surgical duration was 112.29 ± 20.25 min. In the non-fracture patient, surgical time was found to be 100 min. The average blood loss during surgery was 201.43 ± 33.88 ml. The mean follow-up of all the patients was 54.33 ± 3.2 weeks. Patient no. 5 with artificial bone graft (calcium hydroxyapatite granules) showed healing at 14 weeks. Patient nos. 1 and 6 received both autograft and allograft, and fracture site healing was seen at 14 and 16 weeks, respectively. Patient nos. 2, 3, and 4 received only autograft and healing seen at 12, 14, and 18 weeks, respectively. In our study, mean fracture healing was seen at 14.8 ± 2.28 weeks. However, our study sample is small to reach a definitive conclusion about the effects of various grafts on healing time. Full weight-bearing was started in all the patients at an average of 15.67 ± 2.94 weeks. None of the patient had any surgical site infection. However, one patient (pt. no. 4) had shortening of 1 cm. None had significant varus collapse (>5°) at fracture site. The intraoperative tissue curettage histopathology report supported our diagnosis. Histopathological reports showed woven structure of bone lacking osteoblastic rimming, Sharpey’s fibers along trabeculae edges, and characteristic “alphabet soup” appearance. During follow-up visits, the functional outcome was studied using revised MSTS score and TESS for every patient (Table 2).

Discussion

Fibrous dysplasia of bone is a genetic, non-inherited orthopedic condition resulting from the activating mutation of the GNAS1 gene. However, fibrous dysplasia rarely undergoes malignant transformation [5, 7]. Most of the studies in the literature have shown adult patients presenting with pathological fracture, though from our study, it can be said that pathological fractures can occur in children and adolescent group which needs special attention. Most of the lesions are small and silent or with minimal symptoms such as pain, difficulty walking, and swelling. Some are large lesions with moderate-to-severe symptoms such as severe pain, limp, deformity, and limb length discrepancy. Some may present with a fracture with trivial trauma and diagnosed with pathological fracture during fracture films [18]. Treatment of the lesions located at this site makes the treatment difficult. Increased stress concentration and muscle pull

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**Table 1: Functional outcome of patients with bone grafting with internal fixation.**

| Patient no. | MSTS score | Pre-operative | 1 month | 3 months | 6 months | 12 months |
|-------------|------------|---------------|---------|----------|----------|-----------|
| 1           | 20         | 4             | 12      | 22       | 26       | 96        |
| 2           | -          | 3             | 18      | 24       | 27       | 99        |
| 3           | 15         | 2             | 8       | 18       | 22       | 85        |
| 4           | -          | 3             | 14      | 20       | 24       | 94        |
| 6           | -          | 6             | 10      | 16       | 22       | 76        |
| Mean±SD     | -          | 3.61±1.52     | 12.4±3.85 | 20±3.16 | 24.2±2.28 | 90±9.41  |

*Patient no. 5 did not receive any internal fixation whose mean MSTS preoperatively was 13 and MSTS at 1, 3, 6, and 12 months post-operative was 2, 17, 24, and 27, respectively. TESS at final follow-up was 95.*
proportionately increase the chances of fracture at the proximal femur. The age of the patient, lesion size, presence of fracture, and behavior of the patient influence the treatment choice [19]. Various treatment modalities such as bisphosphonate therapy, curettage and bone grafting, internal fixation with nailing or plate-screw system, and valgus osteotomy have been described in the literature [11, 20, 21, 22]. Baghdadi and Arkader had described about “total bone fixation” with intramedullary fixation devices in Ippolito type 3 and 4 proximal femoral fibrous dysplasia and pre-operative 3D printing to determine accurate osteotomy sites in difficult cases of Shepherds crook deformity [23]. Various bone grafts are used in the treatment of these lesions. Ippolito et al. and Leet and Collins showed that cortical bone grafts are frequently resolved and do not hold disease progression [5, 24]. Glancy et al. showed no difference in success rates between allograft and autograft in lesions <60 ccs. Whereas lesions more than 60 ccs autografts appeared to be superior [25]. All the surgical patients in our study showed fracture site healing radiologically and clinically at around 14 weeks postoperatively. Follow-up radiographs showed increased cortical thickness, maintained neck-shaft angle, and no recurrence of lesion.

Lala et al. showed that bisphosphonate therapy in children and adolescents with McCune Albright syndrome increased bone density and decreased pain [26]. Leet and Collins suggested that bisphosphonate does not change the natural history of the disease but relieves pain [24]. However, in all these studies, size or type of the lesion was not taken into consideration. Bhadada et al. in their case report described a novel therapy of injecting intralesional bisphosphonate, which at follow-up showed improved pain scores and decreased uptake on bone scintigraphy [27]. In our study, we avoided bisphosphonate therapy as most of our study population came with pathological fracture and underwent surgery. One of the two patients (patient number 3) with type 2 femur was started on conservative therapy but progressed into an insufficiency fracture after trivial trauma at medial calcar after 2 months of therapy and needed bone grafting and fixation. The other patient with a similar lesion was treated with curettage and ceramic hydroxyapatite bone substitute. Both patients showed radiological and clinical signs of union at an around 14 weeks postoperatively. However, in our study, the healing of the lesion cannot be attributed to the use of ceramic bone substitute in patient number 5.

Very little literature assessed the functional outcome after treating monostotic fibrous dysplasia in the proximal femur in children using revised MSTS score and TESS. A study by Nishida et al. showed that all patients had improved MSTS scores at final follow-up after using cortical autograft and internal fixation [15]. A study by Ebeid et al. showed that mean MSTS was 27.63 when they used internal fixation without grafting [20]. Despite different treatment modalities, we found that improved MSTS score from 1-month post-operative period to final follow-up. At 1 month, the MSTS score decreased initially because of post-operative pain, weight-bearing restrictions, and emotional factors. However, the MSTS score gradually improved at subsequent follow-ups. Similarly, MSTS score and TESS of curettage and artificial grafting patient were 27 and 95, respectively, which are comparable with the patients with different treatment modalities in our study. The decreased MSTS score in these two patients during early follow-up was due to prolonged immobilization and delayed weight-bearing compared to other patients.

**Conclusion**

In this case series of six patients with monostotic fibrous dysplasia of proximal femur, we found that the ideal treatment modality should be chosen wisely and individualized after analyzing pre-operative radiograph, lesion size, presence of pathological fracture, and each patient profile. Furthermore, the type of graft should be decided preoperatively according to the size of the lesion. Extramedullary fixation devices can be an alternate choice of implant in children and adolescents for monostotic fibrous dysplasia. According to our study, it is crucial for the patients to follow the pre-determined treatment protocol for early and better functional outcomes.

**Clinical Message**

Proximal femur monostotic fibrous dysplasia in children can be challenging due to lack of graft and intramedullary implant options. Our institutional treatment protocol of size and location-based treatment, use of allografts, and extramedullary implant options have shown promising primary results both in terms of radiology and functional outcomes.

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