

Monostotic Fibrous Dysplasia of the Hip Treated with Autologus Non-Vascularized Fibular Graft

Farid Murfuadz^{a*}, Otman Siregar^b, Andriandi^c

^a faridmurfuadzsitepu@gmail.com

^aResident of Orthopaedic & Traumatology, Faculty of Medicine, Universitas Sumatera Utara/ H. Adam Malik General Hospital-Medan, Indonesia

^bStaff of Orthopaedic and Traumatology, Spine Division, Faculty of Medicine, Universitas Sumatera Utara/ H. Adam Malik General Hospital-Medan, Indonesia

^cStaff of Orthopaedic and Traumatology, Oncology Division, Faculty of Medicine, Universitas Sumatera Utara/ H. Adam Malik General Hospital-Medan, Indonesia

Abstract

Fibrous dysplasia represents a dysplastic disorder of bone characterized by solitary or multifocal polyostotic intramedullary lesions composed of proliferations of fibroblast-like spindle cells with a characteristic whorled pattern in which trabeculae of immature woven bone. A 20 years old female with pain and limping on his left hip has been suffered for about 1 month. The pain was localized on the left hip with dull characteristics, and worsen when she moves and walks. The pain intensity also decreased with rest. Then she came to hospital and performed X-ray with shepherd crook deformity on her left hip and performed ipsilateral non-vascularized fibular strut graft on her left hip. Right after post-operative wound has been healed the chief complain was marked decrease. For about 2 years after the surgery follow up by X-ray and also MRI examination with contrast the condition of the fibular strut graft was viable, the complaint was gone and the progression of the disease has been stopped.

Keywords: Monostotic Fibrous dysplasia, Autologous non-vascularized fibular strut graft, impending fracture

1. Introduction

Fibrous dysplasia is a rare benign bone disease caused by post zygotic, activating mutation of the GNAS gene, which alters the signaling of G-Protein at the cellular level that characterized by local replacement of healthy bone by fibrous tissue which is produced by poorly differentiated osteoblasts, osteoclast activation, and local increase in bone turnover (Majoer et al, 2017).

Fibrous dysplasia has two basic clinical form: monostotic and polyostotic. Monostotic fibrous dysplasia occurs more frequently than the polyostotic variant. The lesion may occasionally be heralded by mild to moderate pain of long duration. Pathologic fracture, particularly in the long tubular bones, may also be a presenting symptom (Czerniak, 2016)

Management of fibrous dysplasia of the proximal femur is a progressive, often recurrent condition of bone that can cause skeletal deformity, fractures, and pain. cortical strut grafting to minimize the risk of fracture or as part of fracture treatment is a promising treatment option (Ghosh et al, 2015).

The difference between autograft and allograft reconstructions, that can be done either as vascularized or non-vascularized graft. The advantage of using vascularized graft is rapid biological incorporation, good growth potential, and the ability to thrive in compromised soft tissue. However, these require technical expertise, specific tools for operation and complete facilities. The long term follow-up of non-vascularized

fibular graft also gives good result (Masquelet et al, 2000).

The optimal management for fibrous dysplasia of proximal remains debatable, internal fixation alone without grafting also has satisfactory functional outcome (Ozsen et al, 2016). For carefully selected patient (symptomatic dysplastic lesion without varus deformity of proximal femur) cortical bone grafting with fibular strut graft is an excellent procedure which provide strong structural support to biomechanically weakened bone (Ghosh et al, 2015).

Fang et al. in their study recommended curettage, bone grafting, and internal fixation for large lesions with deformity or high pathological fracture risk and showed good clinical outcome (Fang et al, 2018). Nishida et al. showed Autogenous fibular cortical strut grafting and compression hip screw fixation achieved good post-operative function and provided an early return to work for adult patients (Nishida et al, 2015). Nevertheless, Ebeid et al. in their study suggested that internal fixation alone without grafting had a good local control and satisfactory functional long-term outcome, but 4 out of 19 patients had post-operative complications such as mild limping, residual varus deformity, and infection (Ebeid et al, 2018).

The purpose of this report is to share our experience to treat and prevent fracture on monostotic fibrous dysplasia of hip using non-vascularized fibular autogenous grafts and also 2 years follow-up after surgery that show no progressive deformity on the affected bone.

2. Case reports

A 20 years old female with pain and limping on his left hip has been suffered for about 1 month. The pain was localized on her left hip with dull characteristics, and worsen when she moved and walked. The pain intensity also decreased with rest. Then she came to hospital with antalgic gait appearance, from physical examination there's no significant lump or even shortening on her lower extremity and then pelvic X-ray was performed. From her pelvic X-ray we found shepherd crook deformity with ground glass appearance on her left hip (Fig 1).



Fig 1. Pelvic X-ray

After that this patient was planned to perform non vascularized autologous fibular graft as strut graft on her left hip and the graft was taken on ipsilateral leg along with biopsy on her left hip. The patient was positioned with right lateral decubitus on operating theatre (Fig 2), and the greater trochanter was marked with skin marker and the incision and approach was made according to direct lateral approach of the hip. Along with approach to the hip we also take the fibular graft using direct lateral approach and the fibula was taken above 7 cm from syndesmosis and the graft was collected with 9 cm in length. After the graft was taken on the hip,

we performed drilling along femoral neck with C-Arm Guided and also curettage for histopathology examination, then we insert the fibular graft until the proximal anchoring was achieved then the excess graft was cut (Fig 3).



Fig 2. Patient position on operating theatre

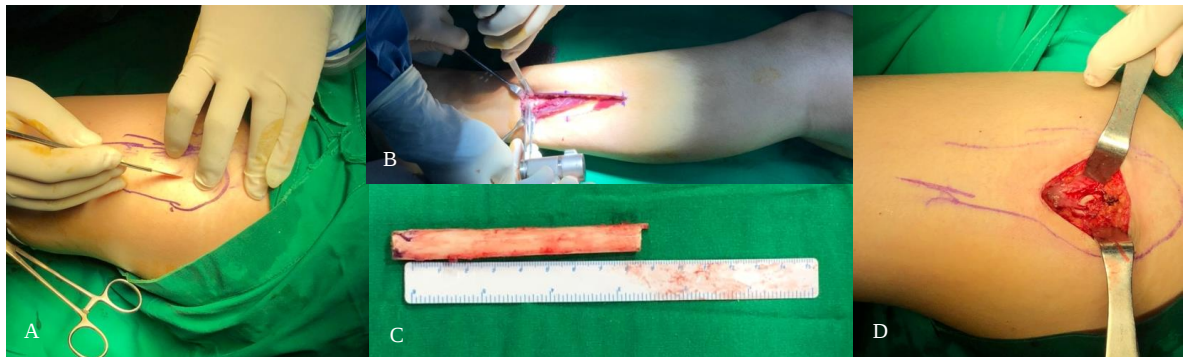


Fig 3. (a) Incision and design; (b) Collecting Fibular Graft; (c) Non vascularized fibular graft; (d) Graft Inserted along the femoral neck

Then after surgery patient was performed pelvic Xray (Fig 4) and hospitalized for a short time then discharge home and advice to non-weightbearing for 2 weeks, and after 8 weeks partial weight bearing was allowed, and after 12 weeks the patient was allowed for full weight bearing to allow adequate healing, and integration of the bone graft. After the post-operative wound has been healed, the patient routinely control to polyclinic for serial X-ray (Fig 5).



Fig 4. Post-Operative Xray



Fig 5. Pelvic X-Ray (a) 6 months after operation; (b) 12 months after operation; (c) 24 months after operation

Then the patient was performed MRI of her left hip with contrast to make sure that the graft that we put avascularly on the proximal femur as a strut graft is viable (Fig 6). Final follow-up found in 24 months radiographs (Xray and MRI) showed consolidation and the strut bar of fibular graft stop the progression of the disease and prevent impending fracture. On the MRI with contrast there is an angiogenesis infiltrating the avascular fibular graft (Fig 7).

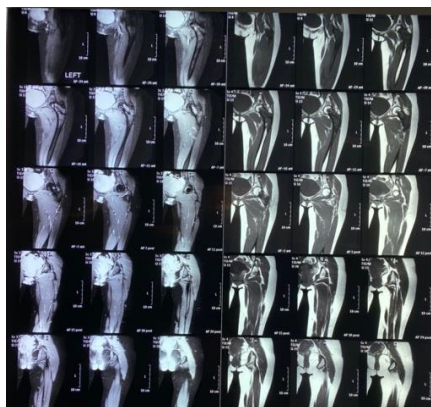


Fig 6. MRI on coronal section without contrast

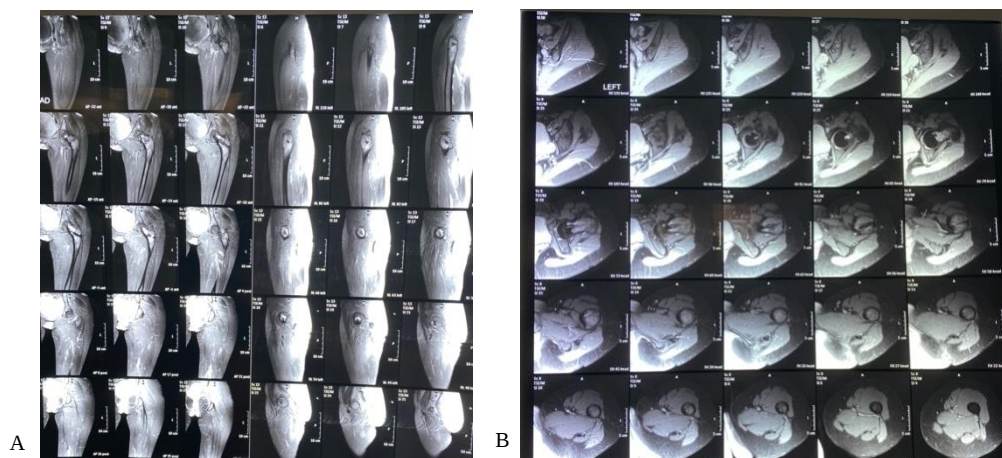


Fig 7. (a) MRI on coronal and sagittal section with contrast; (b) MRI on axial section with contrast

The Hip Outcome Score (HOS) is an instrument used to assess patients with hip disorders who are young, physically active, or both but who do not have severe degenerative abnormalities. Other hip assessment instruments do not address this population with the same degree of specificity (Costa et al, 2019). The instrument was then applied for the evaluation of this patient. Activity of daily living (ADL) the patient showed the score of 91.2%.

2 years post-operatively the patient had a good result, no progression of the disease, with good functional outcome after using a non-vascularized autologous fibular strut graft for monostotic fibrous dysplasia of the hip. The HOS was then retaken and the ADL showed improvement compared to the pre-operative assessment with the score of 95%.

3. Discussion

Fibrous dysplasia is a developmental dysplastic disorder of bone in which the normal bone matrix is replaced by fibroblastic proliferation. In 1938 Lichtenstein and jaffe described that fibrous dysplasia is a benign intramedullary fibro-osseous lesion (Srivastava et al, 2016).

The prevalence of fibrous dysplasia are not rare benign bone tumor, they are reported to represent approximately 5-7% of benign bone tumor, with equal female to male ratio. Fibrous dysplasia most frequently found on the metaphysodiaphyseal region of long bone, such as on rib (28%), followed by femur (23%), tibia and craniofacial bone (10-25%) (Srivastava et al, 2016).

The patient in this report underwent non-vascularized fibular strut graft on her left hip. The proximal femur is a common site of fibrous dysplasia. Surgical management in these cases should therefore

be as minimally invasive as possible, with the aim for the patient to make a complete return to the rigorous daily activities and hobbies of youth as soon as possible, as well as to prevent pathological fractures. The tumor has benign non aggressive nature and an extremely low risk of malignant transformation. The procedure in managing younger patients with fibrous dysplasia should also consider the prevention of any of the risks and complications of instrumentation (Yung et al, 2018). This was the reason for choosing the treatment of this patient.

Although the definitive choice of surgical treatment of fibrous dysplasia on the proximal femur is still debatable, there are some indications for performing this type of procedure such as an impending fracture of the proximal femur, persistent pain, or an actual nondisplaced femoral fracture (Majoor et al, 2017).

Non-vascularized fibular strut graft has several advantages. Bone grafts are the ideal filler agent following curettage of the proximal femur. They are osteoinductive, osteoconductive, and osteogenic, which facilitates the organic incorporation of the graft at the transplant site. Bone grafts also achieve sufficient biomechanical strength in the long term. Non-vascularized cortical bone grafts are the choice graft for transplantation in proximal femoral fibrous dysplasia. They provide excellent initial structural support, while retaining some osteoinductive, osteoconductive, and osteogenic properties. Cancellous bone grafts are no longer used in fibrous dysplasia treatment due to the high reabsorption rate and replacement of the graft by dysplastic tissue, which results in recurrence of the disease. Allogeneous grafts have become more popular recently, but they have some disadvantages because allogeneic grafts do not have osteoinductive or osteogenic properties and, thus, are associated with a slower rate of incorporation (Yung et al, 2018).

We did not perform instrumentation to the patient. There are long-term risks associated with the retention of implants in young patients, which include the risk of peri-implant fractures, implant migration and penetration, implant failure, and infection. Further, fibrous dysplasia comes with the potential of recurrence. In the case of local recurrence, previous instrumentation complicates future surgeries due to implant embedment, as well as local fibrosis and scarring (Yung et al, 2018).

In one study by Ebeid et al reporting fibrous dysplasia of the hip treated with internal fixation without bone graft in 19 patients, 15 patients show satisfied result with complication occur in 4 patients such as limping, infection, varus deformity, shortening of the limb. For carefully selected patients especially fibrous dysplasia without varus deformity of proximal femur, cortical bone grafting with fibular strut graft is an excellent procedure which provides strong structural support to biomechanically weakened bone (Ebeid et al, 2018).

There are general perceptions in practice that a vascular supply should be used when large pieces of bone graft are used, particularly those greater than 6 cm in length for bone reconstruction, but there are still no clear sources that said if bone reconstruction surgery are performed using more than 6 cm of non-vascularized fibular graft will clearly fail.

In our case report, the good result happened in the 20 years old female with complete integration, preventing impending fracture, stopping the progression of the disease and enhancement was found on MRI examination with contrast agent of the nonvascularized fibular graft with length exceeding 7 cm.

The findings of our patient were similar to a case report by Yung et al. who also performed curettage followed by autologous fibular strut grafting to a 19-year-old female basketball player with fibrous dysplasia on the proximal femur. The report showed that the patient was able to fully return to an active and vigorous lifestyle without restriction of activities or long-term risks of orthopedic implant complications (Yung et al, 2018).

Another case report by Ghosh et al. also supported these findings. They presented a case of a 25-year-old female patient with fibrous dysplasia on the proximal femur. They performed autogenous non-vascularized fibular strut grafting to the patient and found that the fibular graft was providing excellent structural support without any need for any form of internal fixation. At routine follow-up at 8 months, the patient was found ambulating at her own without any substantial pain at the hip joint (Ebeid et al, 2018).

4. Conclusions

We followed the patient for 24 months and have good outcome. The monostotic fibrous dysplasia arising on the hip can be successfully reconstructed with fibular graft-giving good functional outcome, stopping the progression of the disease, and preventing impending fracture. However, the other option is using internal

fixation with debatable outcome. Our experiences with non-vascularized fibular graft for reconstruction of monostotic fibrous dysplasia could be the great option with great result and limiting cost using implant for internal fixation, and also shorten operating time compared to autologous vascularized fibular graft.

Acknowledgements

None to acknowledge.

References

- Majoor BC, Peeters-Boef MJ, van de Sande MA, Appelman-Dijkstra NM, Hamdy NA, Dijkstra PD. What is the role of allogeneic cortical strut grafts in the treatment of fibrous dysplasia of the proximal femur?. *Clinical Orthopaedics and Related Research*®. 2017 Mar;475(3):786-95.
- Czerniak B. *Bone Tumors* 2nd edition. Elsevier saunders. Texas. 2016
- Ghosh S, Chaudhuri A, Datta S, Sirdar BK. Fibrous dysplasia of femur treated with fibular bone graft. *Saudi journal of sports medicine*. West Bengal. 2015
- Masquelet AC, Fitoussi F, Begue T, Muller GP. Reconstruction of the long bones by the induced membrane and spongy autograft. *Ann Chir Plast Esthet*. 2000
- Ozsen M, Yalcinkaya U, Bilgen MS, Yazici Z. Fibrous Dysplasia Clinicopathologic presentation of 36 cases. *Turkish Journal of Pathology*. Turkey. 2016
- Fang X, Liu H, Lang Y, Xiong Y, Duan H. Fibrous dysplasia of bone: Surgical management options and outcomes of 22 cases. *Molecular and Clinical Oncology*. 2018 Jul 1;9(1):98-103.
- Nishida Y, Tsukushi S, Hosono K, Nakashima H, Yamada Y, Urakawa H, Ishiguro N. Surgical treatment for fibrous dysplasia of femoral neck with mild but prolonged symptoms: a case series. *Journal of orthopaedic surgery and research*. 2015 Dec;10(1):1-7.
- Ebeid WA, Hasan BZ, Mesregah MK. Management of fibrous dysplasia of proximal femur by internal fixation without grafting: a retrospective study of 19 patients. *Journal of the american academy of orthopaedic surgeons Wolters Kluwer health*. 2018
- Costa RM, Cardinot TM, Mathias LN, Leporace G, Oliveira LP. Validation of the Brazilian version of the hip outcome score (HOS) questionnaire. *Advances in Rheumatology*. 2019 Jul 29;58.
- Srivastava KC, Aruldoss N, Sathick M, Shrivastava D, Austin RD. Monostotic and polyostotic forms of fibrous dysplasia of the jaws – case report. *International journal of advanced research*. 2016
- Yung D, Kikuta K, Sekita T, Asano N, Nakayama R, Nakamura M, Matsumoto M. Conservative surgical management of simple monostotic fibrous dysplasia of the proximal femur in a 19-year-old basketballer: a case report. *Journal of Medical Case Reports*. 2018 Dec;12(1):1-6.