Case Report

# Desmoplastic Fibroma of Proximal Tibia Mimicking Malignant Tumor

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## ABSTRACT

**Introduction**: Desmoplastic Fibroma is one of the most uncommon bone tumors found in only approximately 0.1% of all primary bone tumors and 0.3% of benign bone tumors. This benign bone tumor composed of spindle cells with minimal cytological atypia and abundant collagen production, but locally aggressive. It occurs most often in the first three decades of life and in long tubular bones, mandible, and pelvis. Surgical resection with a wide margin has been advocated for the treatment.

**Methods:** We present a rare case of Desmoplastic Fibroma in an 18year old female with a progressive mass on her right popliteal for the last 1 year. The large and hard consistency of the mass and increased in lactate dehydrogenase (LDH) serum level along with infiltration into the soft tissue characteristics shown in the magnetic resonance imaging (MRI) increased the suspicion of it as a malignant bone tumor. Fine Needle Aspiration Biopsy (FNAB) results were inconclusive, so we performed histopathology examination with core biopsy. All data were discussed in the Clinico-pathological-conference and the conclusion was chondromyxoid fibroma. In accordance with this conclusion, we, therefore, planned to perform marginal excision, followed by curettage to preserve proximal tibia bone and inserted a bone graft completed with internal fixation with plate and screw.

**Results:** Intraoperatively, we found lobulated solid mass extended distally to the posterior compartment of cruris. The tumor was then resected completely and curettage was conducted to the intramedullary mass. We sent the tumor mass to the Pathology Anatomy to confirm the diagnosis. After the next clinico-pathological-conference, it was concluded that the diagnosis was Desmoplastic Fibroma of proximal tibia. The first pathological result of the patient from core biopsy was chondromyxoid fibroma, but at the second pathological result from intraoperative biopsy was Desmoplastic Fibroma. Surgical treatment is the most effective method for Desmoplastic Fibroma. Curettage has been associated with local recurrence rate varying between 37% and 72%. One study reported that marginal resection could result in recurrence compared to wide excision and reconstruction, which showed no recurrence after 1-year follow-up.

**Conclusion**: Desmoplastic Fibroma is not easy to diagnose because it is mimicking malignant bone tumor and should be established in CPC (Clinic-Pathologic Conference). Good surgical planning is very important to minimize complication and rates of recurrence.

## ABSTRAK

**P*endahuluan :*** *Desmoplastic Fibroma adalah salah satu tumor tulang yang paling jarang ditemukan hanya sekitar 0,1% dari semua tumor tulang primer dan 0,3% dari tumor tulang jinak. Tumor tulang jinak ini terdiri dari sel-sel spindle dengan atypia sitologi minimal dan produksi kolagen yang melimpah, tetapi agresif secara lokal. Paling sering terjadi pada tiga dekade pertama kehidupan dan pada tulang tubular yang panjang, rahang bawah, dan panggul. Reseksi bedah dengan margin lebar telah dianjurkan untuk perawatan.*

***Metode:*** *Kami menyajikan kasus Desmoplastic Fibroma yang langka pada wanita berusia 18 tahun dengan massa progresif pada popliteal kanannya selama 1 tahun terakhir. Konsistensi massa yang besar dan keras dan peningkatan kadar serum laktat dehidrogenase (LDH) bersama dengan infiltrasi ke dalam karakteristik jaringan lunak yang ditunjukkan pada pencitraan resonansi magnetik (MRI) meningkatkan kecurigaan sebagai tumor tulang yang ganas. Hasil Biopsi Aspirasi Halus (FNAB) tidak dapat disimpulkan, jadi kami melakukan pemeriksaan histopatologi dengan biopsi inti. Semua data dibahas dalam konferensi Clinico-patologis dan kesimpulannya adalah fibroma chondromyxoid. Sesuai dengan kesimpulan ini, kami, oleh karena itu, direncanakan untuk melakukan eksisi marginal, diikuti oleh kuretase untuk melestarikan tulang tibia proksimal dan memasukkan cangkok tulang yang dilengkapi dengan fiksasi internal dengan plat dan sekrup.*

***Hasil:*** *Secara intraoperatif, kami menemukan massa padat berlobus yang membentang ke kompartemen posterior cruris. Tumor kemudian direseksi sepenuhnya dan kuretase dilakukan ke massa intramedulla. Kami mengirim massa tumor ke Patologi Anatomi untuk mengkonfirmasi diagnosis. Setelah konferensi klinik-patologis berikutnya, disimpulkan bahwa diagnosis adalah Desmoplastic Fibroma dari tibia proksimal. Hasil patologis pertama pasien dari biopsi inti adalah fibroma chondromyxoid, tetapi pada hasil patologis kedua dari biopsi intraoperatif adalah Desmoplastic Fibroma. Perawatan bedah adalah metode yang paling efektif untuk Desmoplastic Fibroma. Kuretase telah dikaitkan dengan tingkat kekambuhan lokal bervariasi antara 37% dan 72%. Satu penelitian melaporkan bahwa reseksi marginal dapat mengakibatkan kekambuhan dibandingkan dengan eksisi luas dan rekonstruksi, yang menunjukkan tidak ada rekurensi setelah 1 tahun follow-up.*

***Kesimpulan :*** *Desmoplastic Fibroma tidak mudah didiagnosis karena meniru tumor tulang ganas dan harus ditetapkan dalam CPC (Clinic-Pathologic Conference). Perencanaan bedah yang baik sangat penting untuk meminimalkan komplikasi dan tingkat kekambuhan.*

**Keywords**: Desmoplastic Fibroma, mimicking malignant tumor, proximal tibia bone tumor

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## INTRODUCTION

Desmoplastic Fibroma (DF), one of the most uncommon bone tumors, is originally described by Jaffe in 1958.[1-3] It occurs in approximately 0.1% of all primary bone tumors and 0.3% of benign bone tumors.[4] This benign bone tumor composed of spindle cells with minimal cytological atypia and abundant collagen production, but locally aggressive.[1] It is also known as desmoid tumor of bone or aggressive fibromatosis of bone or intra-osseous counterpart of soft tissue fibromatosis.[1,2,5]

Desmoplastic Fibroma occurs most often in the first three decades of life and is found equally in men and women.[5-7] Any bone can be affected, and the most common sites are the long tubular bones (56 %), mandible (26 %), and pelvis (14 %).[2,8,9]

Patients present with a variety of symptoms. Some have pain, others present with deformity or loss of function.[1,2] The differential diagnosis includes a low-grade central osteosarcoma and fibrous dysplasia.[1,2,5]

Surgical resection with a wide margin has been advocated for the treatment.[10] Curettage has been associated with local recurrence rate varying between 37% and 72%.[11,12]

We present a case of Desmoplastic Fibroma (DF) found in the proximal metaphyseal region of tibia of an eighteen year old female which was first suspected as malignant tumor.

## CASE ILLUSTRATION

An eighteen year old female patient was referred to our institution with an enlarging mass on her lower limb since 1 year before admission. One year ago, the mass grew from the popliteal region and extended distally to cruris without any pain and wound, the size was approximately 1 cm in diameter. There was no history of trauma. Since then, the mass grew slowly and she began to feel pain on the popliteal. The pain was felt at midnight during her sleep and sometimes felt when she walked or stand. The patient then went to seek alternative medicine (massage) and was treated 3 times, but there was no improvement. Since there was no improvement by alternative medicine, the patient decided to go to the local general hospital and was then referred to our institution.

In physical examination, a hard lump was found on her posterolateral right knee, with a size of 8 x 7 x 6 cm, diameter approximately 30 cm, contralateral 28 cm [Fig. 1]. In palpation, it was found a lump with hard consistency and isolated, the skin felt warmer than the surrounding, but there was no tenderness on the lump or any disturbances on the neurovascular function distal to the lump. The movement of the knee joint was limited.

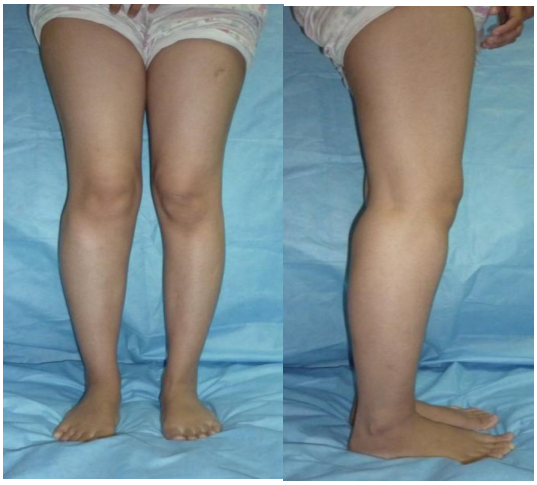


**Figure 1**. Clinical Presentation of the Mass on the Popliteal of an Eighteen Year Old Female Patient

From the laboratory findings, we found an increased level of LDH. From the radiological examination, in the plain radiograph of the right knee, there was shown a tumor in the metadiaphyseal proximal region of the right tibia, suggestive a malignant tumor [Fig 2]. Based on these findings, we suspected it as a malignant surface osteosarcoma.

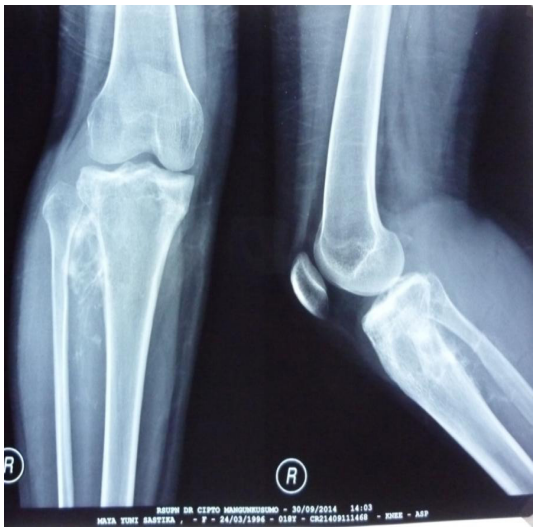


**Figure 2**. Plain X-ray Showed Sclerotic and Lytic Lesions on the Lateral Surface of the Proximal Tibia with Osteolytic Lesion at the Proximal

The MRI of the patient showed the lower limb with clearer image in the right tibia. It showed tumor in the epimetadyaphyseal proximal region of the right tibia with infiltration and soft tissue expansion, suggestive a malignant tumor [Fig 3].

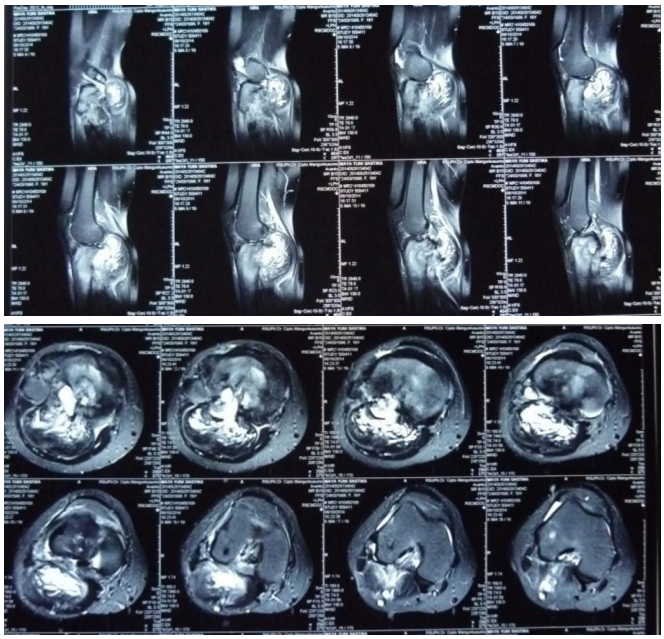
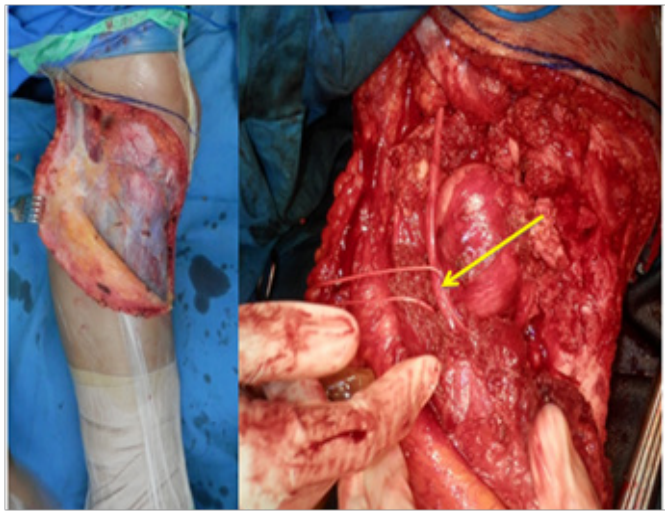


**Figure 3**. MRI of the Right Knee Suggestive a Malignant Tumor

FNAB was performed for the first time, but the result was inconclusive, so core biopsy was conducted followed by cytology examination. From all the findings from these tests, a CPC (Clinic-Pathologic Conference) was performed. From the meeting, it was concluded that the patient had chondromyxoid fibroma and then we suggested to do a marginal excision, followed by curettage to preserve the proximal tibia bone and inserted a bone graft completed with internal fixation with plate and screw.

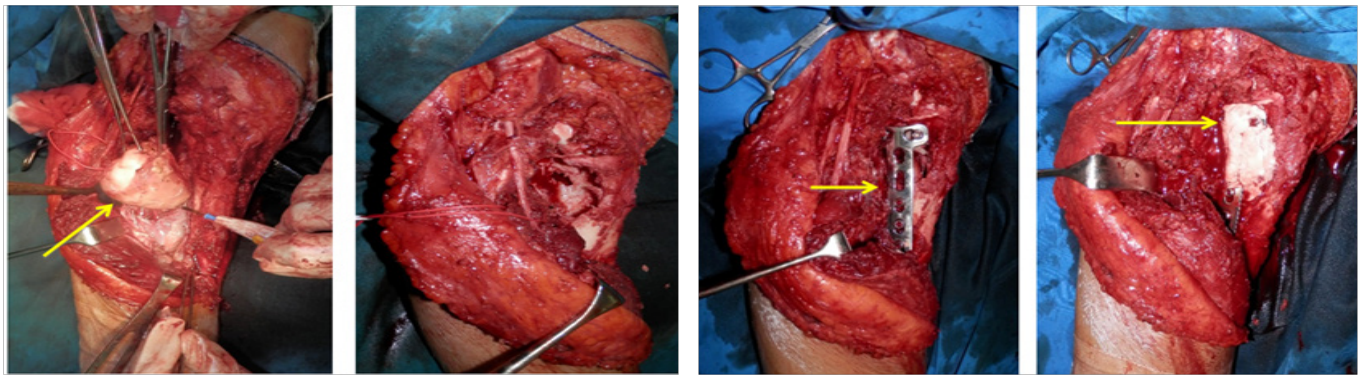
On the operating table, the patient was positioned on the left lateral decubitus in order to expose the right tibia. A curvilinear incision was performed starting from the posterior knee curving distally to one third of the proximal tibia [Fig 4]. Afterwards, we searched for peroneal communis and suroles nerves to be preserved. Blunt dissection was then carefully performed to expose the tumor mass [Fig 4].



(a) (b)

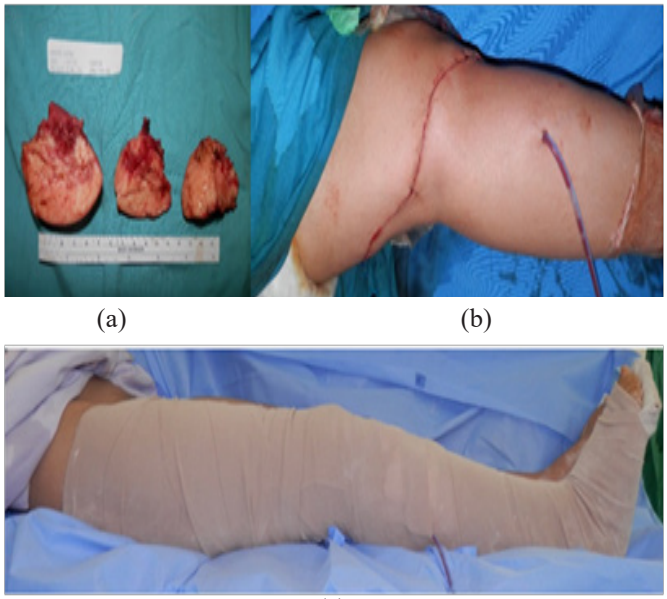
**Figure 4**. (a) Curvilinear Incision of The Skin; (b) Peroneal Communis Nerves to be Preserved.

After fully identified the tumor mass, we found lobulated solid mass extended distally to the posterior compartment of cruris. Proximal tibia osteotomy was then conducted to expose the lateral tibia and the tumor was resected completely [Fig 5]. Curettage of intramedullary mass was performed. Afterwards, we reconstructed bone with bone cement insertion and plate and screw using an 8-hole T-plate [Fig 5]. We sent the tumor mass to the Pathology Anatomy to confirm the diagnosis [Fig 6], and then we closed the wound and inserted a drain [Fig 6]. We put a blackslab from femur to distal tibia after the operation completed [Fig 6].



(a) (b) (c) (d)

**Figure 5.** (a) Identified Tumor Mass; (b) After Proximal Tibia Osteotomy; (c) Internal Fixation with T-plate 8 Holes; (d) Insertion of Bone Cement



(a) (b)

(c)

**Figure 6**. (a) Gross Pathology of The Tumor; (b) Closing the Wound with Drain; (c) Blackslab

After the next CPC, the result was different from our last CPC. The result stated that the diagnosis was a Desmoplastic Fibroma of proximal tibia.

## DISCUSSION

Desmoplastic Fibroma (DF) is one of the rarest tumors of the bone, and the incidence is reported to be between 0.1% and 0.2 % of bone tumors.[1-4,13] It can involve the whole bones such as mandible, femur, pelvic bone and radius.[14] Tibia is one of the commonest locations for this tumor. It occurs most often in the first three decades of life and is found equally in men and women.[5-7]

Most patients give signs and symptoms that are usually nonspecific. Pain and swelling are the predominant symptoms, but some patients may be asymptomatic. Pathologic fractures are reported in 9-15% of cases.[15] One study reported that the average size of a Desmoplastic Fibroma is 7 to 10 cm,[2] in which our patient had a tumor size of 8 cm. From the physical examination, we suspected the mass to be malignant due to its hard consistency and increased lactate dehydrogenase (LDH) level. From many studies, DF may be found incidentally but often presents with continuous mild pain.[1,2] Local swelling and lump is usually palpable. The symptoms can last months to years with progressive lump size. The lump usually does not cause any disabilities, limitation of the adjacent joint range of motion, and pathological fracture.[15]

Radiographs of DF show lytic lesions, often with sclerotic margins.[15] Radiographic differential diagnosis includes tuberculosis, chondromyxoid fibroma, low-grade intramedullary osteosarcoma, giant cell tumor, aneurysmal and solitary bone cyst, hemangioma, fibrous dysplasia, non-ossifying fibroma, and fibrosarcoma.[6,16] In our patient, from the plain radiography result, clinical judgement could not be accurately made because the result of this radiography was not specified to Desmoplastic Fibroma. That's why we needed more advanced diagnostic tools, such as MRI or CT Scan of the tumor, to visualize the tumor more clearly.

In MRI scan, the lesions showed heterogeneous mass with low and high signal intensity. The low signal may represent dense collagen, and the high signal represents the fibromyxoid matrix.[6,16] Although CT best illustrates the extent of bone destruction, MRI better visualizes the medullary as well as the soft tissue extension of the tumor.[6] Therefore, CT and MRI are complementary imaging techniques in suspected DF. In this patient, MRI was chosen to visualize the tumor, the result suggested that it was a malignant tumor because the characteristic was very aggressive and it had infiltrated and extended to the soft tissue nearby. However, to ascertain the diagnosis, pathological examination was performed to find out definitive diagnosis from this patient.

DF is classified as a benign tumor.[1,2] The typical pathological appearance consists of fibroblasts and various amounts of collagenous matrix.[1,2] Fibroblasts are spindle-shaped cells with elongated hyperchromatic nuclei.[1,2,13] No giant cells or hemosiderin deposits will be observed.[13] Some reported the absence of mitotic figures.[13] In some cases, there is reciprocal translocation on t(2;11)(q31;q12).[13] The most important differentiation should be made between malignant tumor, such as fibrosarcoma, and low-grade osteosarcoma, for which a delay in diagnosis could lead to death to the patient. Fibrosarcoma of bone is differentiated by the presence offibroblasts with atypia and mitotic figures.[13] Low-grade osteosarcoma will contain osteoids.[13] In our patient, the first pathological result from the core biopsy was a chondromyxoid fibroma, but the second pathological result from the intraoperative biopsy was a desmoplastic fibroma.

Reports on genomic alterations in DF are limited. There are conflicting reports of whether there is abnormality

in the karyotypes. Trombetta *et al*, reported recurrent chromosome aberration in 11q and 19p after short-term culturing of the tumor.[11]

Surgical treatment is the most effective method for DF.[1,2] Curettage has been associated with a local recurrence rate varying between 37% and 72%.[11,12] Wide excision and marginal resection can be selected for DF.[13-15] One study reported that marginal resection could result in a recurrence compared to wide excision and reconstruction, which gave no recurrence after 1-year follow-up.[13] Marginal excision with placement of bone graft and bone cement was performed to our patient. This procedure was chosen primarily for our first diagnosis, which is a chondromyxoid fibroma. However, according to our latest diagnosis, it was actually a Desmoplastic Fibroma of proximal tibia. Based on the literature, our treatment gives a high risk of recurrence as the appropriate surgical treatment for Desmoplastic Fibroma is wide excision.[13] Wide excision generates less number of recurrence rate.[13] That's why a good surgical planning is needed to give maximal results of treatments for the patient. However, in situations in which surgery is not feasible, radiotherapy may be considered as an alternative mode of definitive management.[16]

The tumor behaves in a locally progressive/aggressive manner. Malignant transformation is very rare. There are only occasional reports of malignant transformation.[17]

## CONCLUSION

Diagnostic of Desmoplastic Fibroma is not easy, because it is mimicking malignant bone tumor and should be established in CPC (Clinic-Pathologic Conference). Good surgical planning is very important to minimize complication and the rate of recurrence.

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