



Case report

Cement arthrodesis of the knee using combined intramedullary nail, plate and screws after wide excision in huge chondromyxoid fibroma of proximal tibia, a case report

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ABSTRACT

Introduction and importance: Chondromyxoid fibroma is a rare benign cartilaginous neoplasm, a mixture of benign cartilage and fibrous and myxoid tissue. It usually affects young people, commonly in the second and third decades of life with male being predominant. Differential diagnoses in chondromyxoid fibroma, are giant cell tumor, bone cyst, enchondroma, osteoblastoma, and low-grade chondrosarcoma.

Case presentation: We present a case of an 18-years-old male, with chief complaint of pain on the right knee since 3 year ago with an increasing size reaching diameter of 55 cm within three years. The radiograph and MRI revealed lytic blastic expansile multiloculated lesion, with wide transitional zone, and periosteal reaction on the epimetadiaphyseal part of right proximal tibia. The surgical procedure was performed consist of wide excision, reconstruction by cement knee arthrodesis using Kuntscher-nail, dynamic compression plate, bone cement and gastrocnemius flap.

Clinical discussion: After 3 months of surgery follow-up, there was no sign of recurrence, the patient walked partially weight bearing with a crutch. In 1 years post operative follow up, the Musculoskeletal Tumor Society Scoring (MSTS) was 80%. There was no sign of recurrence.

Conclusion: In cases with large tumor size, wide excision with bone and soft tissue reconstructive surgery is required as mechanical and biological reconstruction. The cement arthrodesis provides a faster duration of surgery, lower risk of infection, and good functional outcome.

1. Introduction

Chondromyxoid fibroma is a rare benign cartilaginous neoplasm, composed a mixture of benign cartilage and fibrous and myxoid tissue, lobulated formed by spindle shaped cell with myofibroblastic features at the periphery, stellate and chondrocyte-like cells to the center. It generally develops in long bones of the lower extremity [1–4]. Chondromyxoid fibroma is a very rare tumor, represents of less than 0.5% of all primary bone tumor, widely age range from first to seventh decades of life, more commonly in the second and third decades of life with predominantly male [1,2,4,5]. It was described firstly by Jaffe and Lichtenstein in 1948, when presented 8 cases of this benign neoplasm as malignant lesion [2]. It usually occurred at almost any osseous site, frequently at the long bone. 25% of cases occur at flat bones, mainly in the ilium. The histogenesis of chondromyxoid fibroma is still uncertain.

It is also evident that the tumor is associated with chromosome 6 aberration heterogenous, include 6p23-25, 6q 12-15 and 6q13-17 [1,3].

Clinical features of chondromyxoid fibroma are pain, and swelling associated with a lump. The pain is usually intermittent and not distressing [1,3,4]. Radiological features showed a lytic radiolucent medullary lesion, eccentric, with a thin sclerotic rim, on metaphyseal, sharply marginated, oval zone, rarefaction, with expansion on the cortical area. Most lesion is lucent, approximately 10% may show focal calcified matrix. The cortical destruction is usually affected but within the periosteum [1,4,6]. The lesion rarely contiguous bone is affected. The differential diagnoses of chondromyxoid fibroma are benign tumor lesions such as giant cell tumor, simple bone cyst, aneurysmal bone cyst, enchondroma, eosinophilic granuloma, osteofibrous dysplasia, osteoblastoma, osteofibrous dysplasia, and nonossifying fibromas [1,3,7].

Gross pathology features of chondromyxoid fibroma include well

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defined margin, bluish grey or white tumor which lack necrosis area, cystic change and liquefaction, multilobulated and well demarcated from surrounding bone. Histopathological features showed sharply demarcated from surrounding bone, and the lobule's part may be separate from the main lesion. The lobular pattern consists of a spindle-shaped cell in myxoid background. Rare cartilage changes are present. In older patients and in flat bones, the lesion may show calcification usually coarse [1,3,8].

There is no specific recommendation in the management of chondromyxoid fibroma due to limited published articles with chondromyxoid fibroma patients that are already treated. The surgical options include curettage and excision, with or without filling of the cavitory defect, wide excision to avoid recurrence [1,3]. In advanced cases, amputation can be a choice due to the large size of the tumor, or as a result of recurrences. Adjuvants such as polymethyl methacrylate (PMMA) are recommended due to local aggressiveness of chondromyxoid fibroma. Curettage alone has resulted in a rate of high recurrence in most series. There is no indication for radiation therapy except for very rare surgically inaccessible tumors [2,6,13].

In addition, wide excision in the proximal tibia, is considered to be the site that is complicated, where the complication rate is high, and whose functional outcome is poorest [9]. The lack of muscle coverage of tibia, relatively small blood vessels around the leg, insertion of extensor mechanism are the reasons for the complicated surgery in the proximal tibia. In case of a large bone or where soft tissue defect after tumor resection creates another problem. The possible reconstruction includes primary arthrodesis, prosthetic replacement, and allograft replacement. The allograft has higher rates of nonunion and infections. The endoprosthesis has a good functional outcome but it is very expensive especially in developing countries [9,10]. The use of knee arthrodesis, provides shorter time of surgery, lower infection rate, and early bearing [11].

We present a case of huge chondromyxoid fibroma of proximal tibia, treated with the wide excision, reconstruction using knee arthrodesis, bone cement, and gastrocnemius flap in 18 years old male with good postoperative functional outcome. This study is managed and written in line with the Surgical Case Report Guidelines (SCARE) Criteria [12].

2. Case illustration

A male, 18 years old high school student, with chief complaint of pain on the right knee since 3 year ago. He had history of a fall while playing ball after that he went to a traditional bone setter. The pain didn't subside and there was a lump the size of a tennis ball which further grew 4 times bigger within three years. He had difficulty with mobilization due to huge mass and pain. He performed daily activities and ambulation with the assistance of his family. He had been taken to traditional bone setter several times. He also had a core biopsy at the other hospital with the result of chondromyxoid fibroma, but he didn't

continue medication. There was no history of previous allergy or history of the malignancy in his family. The physical examination, revealed that there was tumor mass on the proximal part of the right cruris, visible venectation, no distal oedema with the knee in a flexed position. The size of the mass was 55 cm circumferentially with a contralateral size of 37 cm circumferentially. The mass was immobile, with a well-defined border, warmer than surrounding, and with normal distal sensory and distal capillary refill time. The range of motion of the knee was limited and fixed at 80° (Fig. 1).

The laboratory examination revealed a normal tumor marker and an infection marker. The hemoglobin level was 14.1 g/dL, leucocyte was 10.050/ μ L, alkali phosphatase was 174 U/L; lactate dehydrogenase was 167 U/L; C-reactive protein was 9.2. The radiograph and MRI revealed lytic blastic expansile multiloculated lesion, with wide transitional zone, with a periosteal reaction around the proximal tibia. There was no soft tissue involvement, and the lesion didn't affect the distal part of the femur. There was neurovascular bundle involvement based on MRI (Fig. 2). The histopathology from a previous hospital revealed multiple lobule chondroid tumor, with stellate or spindle-shaped cells, consisting of hypercellular area, round/oval nucleus, slight pleomorphic and hyperchromatic cells. From the radiology and previous biopsy, we concluded the diagnosis was chondromyxoid fibroma.

The surgery was then performed consisting of wide excision, reconstruction by knee arthrodesis using Kuntscher-nail, dynamic compression plate 4.5 narrow, cerclage-wire, bone cement and gastrocnemius flap. Intraoperatively, the patient was set in a supine position. Sterile preparation and draping were done, the tourniquet was set in the proximal thigh. The skin was incised, the mass is dissected until the tumor was exposed. Fortunately, the neurovascular bundle in the popliteal is not affected by the tumor. The tumor was already affecting the joint; therefore, the distal part of the femur was cut, and the tumor was removed by extraarticular resection.

After the tumor was excised, the defect was 30 cm from the distal resected femur to the proximal resected tibia. The cement arthrodesis was then performed. First, the Kuntscher-nail was inserted from the distal femur into the proximal tibia and the length of the nail that is not inserted into the long bone is 30 cm to ensure the same length as the contralateral leg. Two 4.5 narrowed dynamic compression plate were added, with three cortices had purchased in distal femur and proximal tibia using 4.5 cortical screws. The 1.2 cerclage-wire was augmented to add strength and construction stability. The bone cement was augmented to fill the defect between the prosthesis (Fig. 3). The length was also evaluated and equal with the contralateral limb. The soft tissue reconstruction was performed by covering the prosthesis using gastrocnemius rotational flap. The gastrocnemius muscle was identified, then separated from soleus and Achilles attachment. The soleus muscle is pulled anteriorly to cover the middle segment of the prosthesis and the medial gastrocnemius is used to cover the proximal segment. The medial gastrocnemius muscle is detached from the musculotendon junction,



Fig. 1. Preoperative clinical condition.

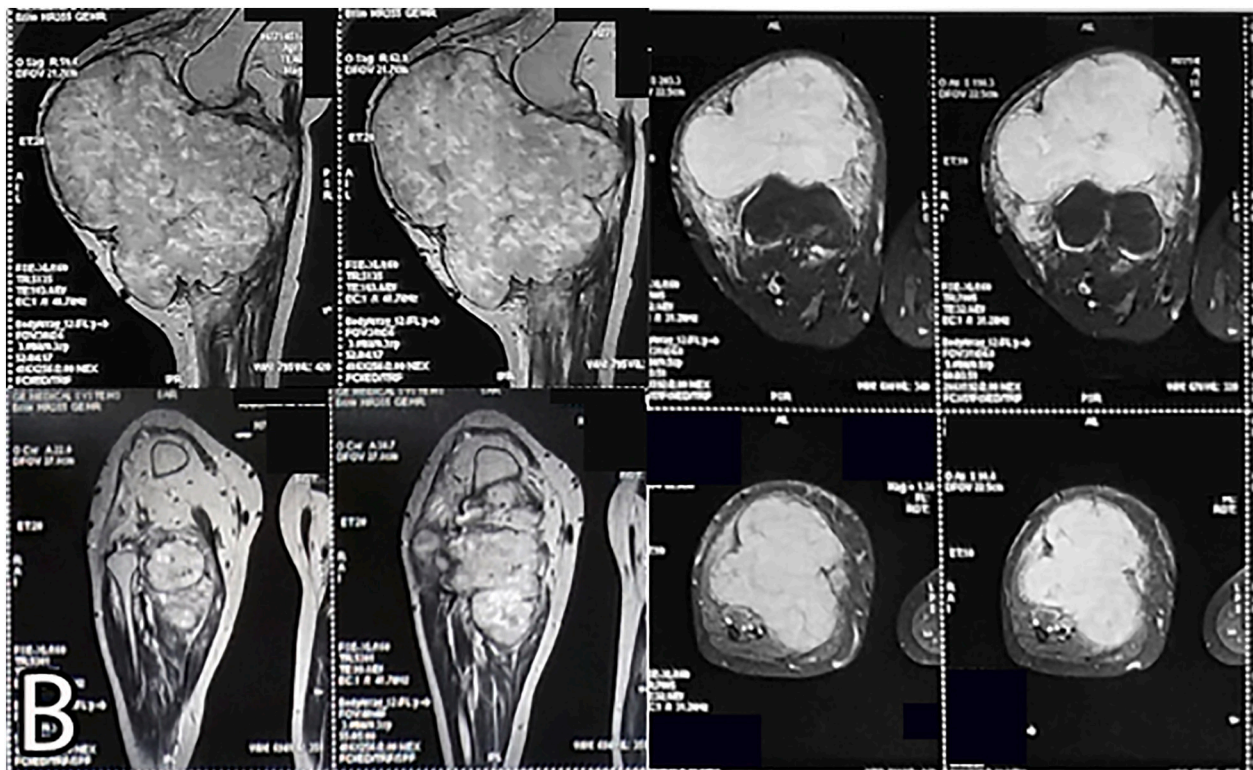


Fig. 2. A. Anteroposterior and lateral view of radiographic right knee, B. MRI with contrast showed expansile lesion on epiphysis, metaphysis and diaphysis of right proximal tibia, with narrow transitional zone, no periosteal reaction, associated with contrast enhancement. There was no neurovascular bundle involvement.

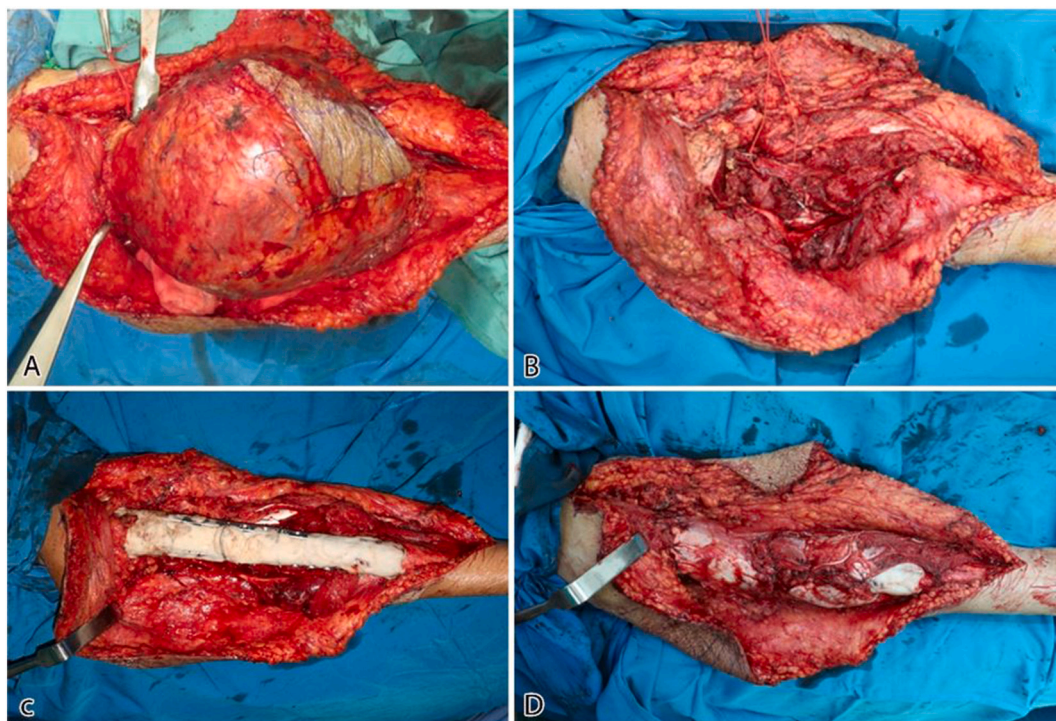


Fig. 3. Intraoperative findings, A, huge tumor mass already dissected from surrounding tissue, B, post wide excision with the length of bone defect 30 cm including distal part of the femur. C. Post reconstruction with Knee arthrodesis. D. Gastrocnemius flap.

rotated anteriorly and its upper pole suture to the lateral gastrocnemius parallel with the preservation of vascular pedicle (Fig. 4). The skin was then sutured and dressed in sterile gauze. We are not the pioneer of this method, but this study provides evidence of a successful technique.

The biopsy result showed hypercellular in the border region consists of oval nucleus, pleomorphic, hyperchromatin, eosinophilic cytoplasmic and osteoclast like giant cell. There are hypocellular areas consisting of stellated cells, with chondromyxoid matrix (Fig. 5).



Fig. 4. The gross pathology of tumor showed a destructive bone lesion infiltrating the surrounding soft tissue.

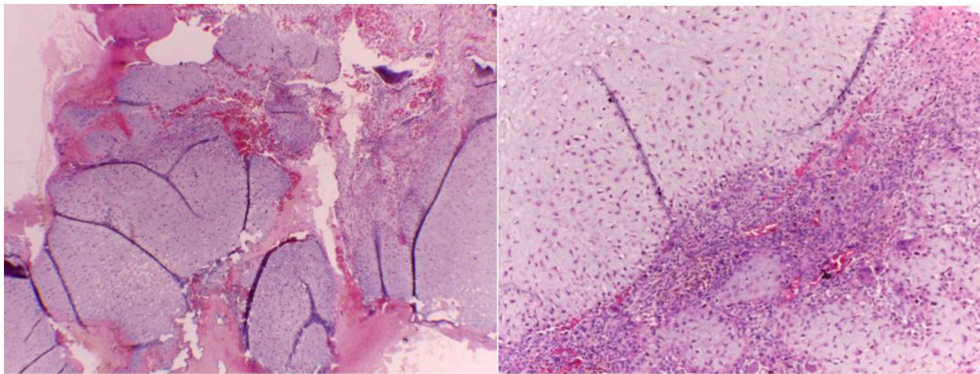


Fig. 5. The microscopic pictures showed tumor mass destructing the bone, and it was lobulated, with hypercellular in the border region consist of oval nucleus, pleomorphic, hyperchromatic, eosinophilic cytoplasmic and osteoclast like giant cell. There are hypocellular areas consisting of stellated cells, with chondromyxoid matrix.



Fig. 6. 3 months postoperative condition. A. Local state clinical condition. B. Postoperative anteroposterior and lateral radiographs.

Postoperatively, the wound is good, with no sign of infection nor dehiscence. After discharged, the patient was underwent rehabilitation and routinely went to an orthopaedic clinic. After 3 months postoperative follow up, the patient was able to walk and partially weigh bearing using crutches. After 1 year postoperative follow up, there was no sign of infection nor local recurrences. The patient is able to perform mobilization partially weight bearing with single crutch (Figs. 6, 7). The functional outcome is good; the score was 80% according to the musculoskeletal tumor society score. The patient also satisfied with current condition compared to the previous preoperative condition.

3. Discussion

Chondromyxoid fibroma is a benign, locally aggressive tumor arising from cartilage with approximately less than 0.5% accounting for all bone tumors. Chondromyxoid fibroma is commonly found in metaphysis of long bone. The most common site is the proximal tibia, estimated at 28%–52% of all lower extremity based on literature [3]. The tumor presents as the insidious onset of pain and swelling. The swelling is tender and usually didn't affect the joint unless the size is big enough to limit the joint motion. A pathologic fracture can occur. The chondromyxoid fibroma is found in the 2nd and 3rd decades of life, with ratio male: female is 1.5:1 [1,7,13].

The complaint of the patient in chondromyxoid fibroma is, pain and swelling at the site of the lesion. The pain gradually increases from mild to severe pain, intermittent, associated with activity. In developed

country it is relatively rare to find such a large lesion. However, in developing country where the background of the patient and access to health service remained the problem, the tumor can be neglected and becomes bigger. Because the tumor is not a malignant, there was no involvement of metastases in the other organ [6,14].

The radiographic appearance of chondromyxoid fibroma is the lytic expansile geographic bone lesion, lobulated, septation or scalloped border, sclerosis and calcification within tumor matrix in metaphysis region. Anthony et al. revealed that only 13% of chondromyxoid fibroma showed calcification which is rare. MRI is helpful in understanding the extension of the tumor spread, intraarticular involvement or adjacent tissue involvement. In T1 weighted image, it showed hypointense to intermediate signal throughout the lesion. The MRI also can reveal internal intracortical osteolytic lesion, hyperintense foci, periosteal reaction, abnormal bone marrow and soft tissue signals [8]. The internal myxoid area generally corresponded to the central hyperintense area on T2-weighted images and the central non-enhancing area on contrast-enhanced T1-weighted images [15,16]. The differential diagnosis of chondromyxoid fibroma based on radiological examination can be aneurysmal bone cyst, nonossifying fibroma, giant cell tumor and chondrosarcoma [15,8,16].

Based on pathological examination, as the key diagnostic, the tumor is typically lobulated pattern with stellate or spindle shaped cells in a myoid background [17]. The lobules showed hypocellular center with hypercellular peripheries, but its center show morphological features that are similar to hyaline cartilage (extracellular matrix and cell composition). The cystic or liquefactive change is uncommon. These features were found in our case [3,18].

There are no major recommendations of chondromyxoid fibroma management due to the tumor being extremely rare. The options of management are curettage and excision, with or without filling defect. In a bigger lesion, wide resection or en bloc resection is the treatment of choice to avoid recurrence [3]. In this case, preoperatively and intraoperatively, the lesion didn't involve the main neurovascular bundle. Mechanical reconstruction and biological reconstruction are very important during reconstructive method. The mechanical reconstruction was then performed using cement arthrodesis technique with Kuntscher-nail, dynamic compression plate 4.5, cortical screw 4.5, cerclage wire 1.2 and bone cement. The gastrocnemius flap was performed as the biological reconstruction. This reconstructive method was chosen due to the limitation of advanced prosthesis and cost. Other alternatives such as advanced endoprosthesis were not feasible in our country due to its expensive cost. In addition, the cheaper implant still able to provide good functional outcomes, therefore it is selected like this case.

Previous study reported similar results to our findings. Capanna et al. performed resection and cement arthrodesis in 76 patients. It showed that postoperatively, the patient was able to weight bearing a few days after the operation. Reported complications indicated that 14% had infection and 16% had implant failure. According to these findings, cement arthrodesis is preferable which allows immediate weight bearing with a lower infection rate. This technique is also preferable in patients currently at their growing age. Even fourteen patients have been walking for more than 3 years [11].

In our case, the cement arthrodesis as the treatment after proximal tibia chondromyxoid fibroma showed a satisfactory result. The functional and clinical outcome 1 year postoperative is good. There was also no sign of local recurrence. Unfortunately, we didn't find any other study that reported similarly huge chondromyxoid tumor to compare.

4. Conclusion

Chondromyxoid fibroma is a rare benign bone neoplasm. It can often be misdiagnosed as another benign even malignant tumor. In the case of huge tumor size, wide excision with bone and soft tissue reconstructive surgery is required as mechanical and biological reconstruction. In case



Fig. 7. 1 year postoperative condition.

of limitation of the provided advanced endoprosthesis, the cement arthrodesis provides a faster duration of surgery, lower risk of infection, and good functional outcome.

Consent

Written informed consent was obtained from the patient for publication and accompanying images of this report.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Ethical approval

Ethical approval clearance was not required in the treatment of the patient in this report.

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Achmad Fauzi Kamal
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Research registration number

We are not pioneer of this surgery.

CRediT authorship contribution statement

Achmad Fauzi Kamal: Concept, design, surgeon, data analysis or interpretation, clinical studies, manuscript editing and review.
Riko Satriyo Wibowo: Concept, design, surgeon, data collection, manuscript editing and review.

Declaration of competing interest

The authors declare that there is no potential conflict of interest.

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