Desmoplastic Fibroma Of Proximal Femur - Surgical Reconstruction. A Case Report And A Brief Review Of Management

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Abstract

Desmoplastic fibroma is a very rare bone tumor which histopathologically and biologically very much similar to extra-abdominal desmoid tumor of the soft tissue. En bloc resection is the treatment of choice in view of the high incidence of recurrence after curettage, but when the lesion is located at sites like proximal femur, resection of the tumor needs reconstruction of the defect either with endoprosthesis, autograft or allograft. Reconstruction of desmoplastic fibroma which is extending from neck of femur up to middle third of remur femur is not reported in literature. We report a case of an eighteen year old male presented with massive tumour of the proximal femur extending from neck up to middle of shaft that was widely resected and reconstructed by using structural fibula and cortico-cancellous graft supported by dynamic condylar screw and side plate. At five years follow up, there was excellent remodelling of the bone with no tumor recurrence.

Introduction

Desmoplastic fibroma of bone is a rare benign bone tumor. The incidence of this tumor is approximately 0.1%–0.3% and most of them are histologically benign. A locally aggressive distinct clinico-pathological entity was described by Jaffe in 1958. Tumor is made up of wavy fibroblasts and abundant collagenous tissue that histologically resemblance to extra-abdominal desmoid tumor arising from soft tissue. There is lot of literature regarding the management of tumour most of which suggest wide excision but reconstruction of the affected bone segment by autograft is described in very few reports. Clinical decision becomes difficult when the lesion is located at sites like proximal femur. Commonly described treatment in this scenario is excision and endo-prosthetic reconstruction. Reconstruction following excision of femur from neck up to middle third of shaft is not reported in literature. Here we are presenting this rare case with our management and a brief review of literature.

Case Report(s)

An eighteen years old male presented with history of pain and progressive swelling over proximal thigh right side since one year. Patient gave history of fall from bullock cart one year back. He developed pain and swelling over right thigh but he was able to walk. Patient took no treatment at that time except for some analgesics. Pain decreased but swelling persisted and gradually increased over time. At the time of presentation patient had pain and swelling over right thigh and limp while walking.

Examination revealed swelling of about 20x15 cm on antero-lateral aspect of right thigh. The swelling was firm and tender on palpation. The overlying skin was normal without any adhesions to the swelling. The margin of the swelling was not well defined. There was 4 cm shortening of the affected site. The range of motion at right hip joint was normal except for slight limitation of adduction and abduction movements. The draining lymph nodes were not enlarged.

On radiograph, the affected bone showed well defined expansile lesion with thin cortex and fine intraslesional trabeculae giving rise to multi-lobulated soap bubble appearance, with loss of corticomedullary differentiation. There was no periosteal reaction. The lesion was extending from base of neck up to the mid third of femur (Fig. 1). The whole body skeletal survey didn’t show similar lesion elsewhere in the body.

His routine blood investigations were within normal limits.

Open biopsy was done which revealed fleshy red and rubbery lesion with irregular margin on gross inspection. Histopathology of biopsy material showed multiple fragments of mature lamellar bone along with fibrous tissue which comprise of small spindled cells arranged in haphazard array with intervening collagenous tissue. These cells show minimal pleomorphism and no mitotic activity. There were few foci of new bone formation at interface between...
fibrous tissue and lamellar bone (Fig. 2). There were no giant cells, no aneurysmal blood spaces or any features of fibrous dysplasia. Conclusion drawn was desmoplastic fibroma.

Surgical treatment was done by wide resection of the whole lesion extending from neck down to midshaft of femur, and massive bone graft in form of structural fibula graft and corticocancellous graft chips from the both side of iliac crests. Construct was supported with 12 hole long side plate and dynamic condylar screw fixation in femoral head. The medial calcar was constructed by fibula which was held by cerclage wire to the side plate that formed the lateral pillar. Gap in between these two was filled by corticocancellous chips (Fig. 3). Post operatively hip spica was applied up to six weeks for additional support. Spica was removed at six week post operatively and bed side mobilization was started. Partial weight bearing was allowed after 3 month when adequate callus was visible in follow up radiographs. Full weight bearing was allowed after 6 months. The patient started walking without stick and doing some physical activity after 8 months. After 10 months, the patient had started all routine activities of day today life. At latest follow up of 5 years, the graft showed incorporation and good consolidation and no recurrence of the lesion without any restriction of the hip movement(Fig. 4&5). The patient was informed that data from the case would be submitted for publication, and gave his consent.

Discussion

Desmoplastic fibroma is a rare bone tumor, accounting for 0.3% of the benign bone tumors2. Fewer than 200 cases had been reported worldwide in the literature. Most of the patients reported are age less than 30, and equals in both sex. The most common site is the mandible, followed by pelvis. In the long bones, the lesion is usually metaphyseal location and centrally placed, but it may be located anywhere in the bone5, 12, 18. Radio-graphically the lesion gives lytic and honeycombed appearance3. Common differential diagnosis are unicameral bone cyst, fibrous dysplasia, chondromyxoid fibroma, non-ossifying fibroma, gaint cell tumor of bone, and fibrosarcoma of bone5, 9, 19, 20. Histologically the tumor has interlacing bundles of dense collagen and low cellularity. The fusiform cells which are present have no atypia and the nuclei are ovoid or elongated5, 19, 20. Distant metastasis is not common, but local recurrence is common.

According to Jaffe, trauma was believed responsible in some of the case histories, but there was no patient in whom he could find a causal relation to the lesion3. Incidentally our case was also associated with history of trauma.

Treatment modalities have been variable in the literature but are primarily surgical. Few authors have recommended intralesional procedures for small lesion and marginal or wide resection for lesion in expendable bones3, 9, 15, 16, 19-21. While some author's advocated wide local resection for all such lesion2, 4, 5, 23. Among all the reported cases in the literatures, the incidence of local recurrence was high after intralesional procedures, due to inadequate removal of the affected tissue. Review of the literature of the patients who were initially treated with resection (intralesional, marginal, or wide excision), 25% had a recurrence in most series4, 5, 9, 18-21.

In the literature, the routinely treatment described for the lesion in proximal femur is endoprosthetic reconstruction as it provides immediate stability and quicker rehabilitation. The problem with this treatment is the longevity of prosthesis. Considering young age of the patient, less longevity of endoprostheses and need of complicated revisions in future; we decided to reconstruct the defect with fibular strut autograft. We completely excised the bone lesion and reconstructed the defect with the help of structural fibular graft on the medial calcar side of femur and the lateral pillar was constructed with long dynamic compression screw and side plate and the intervening region was filled with autologous corticocancellous bone chips taken from both iliac crests. Final follow up radiograph at five years showed excellent union and remodelling. The fibula was converted into calcar and whole graft was incorporated. There was no restriction of the limb movements or limb length discrepancy and the patient returned to his routine physical activities.

Only few studies in literature had been reported regarding the bony reconstruction of such a big lesion in femur with such a good outcome24. One case of desmoplastic fibroma of middle third of femur reconstructed with fibula had been reported with good outcome25. Another report had shown a case of 17 year old girl with desmoplastic fibroma of the distal femur which was treated by en bloc proximal resection and distal intralesional curettage and anatomic specific allograft femoral replacement. There was no recurrence of the tumor three years after surgery, and function was excellent8. There are only few data to support the use of radiotherapy in treatment of desmoplastic fibroma21.
Conclusion

We concluded from our study that wide marginal resection with good bony reconstruction from allograft and internal fixation may be a good therapeutic option with excellent outcome, even when the lesion is located in major long bones and too proximally located or too large which is otherwise is treated by endoprosthetic reconstruction.

Authors contribution(s)

All the authors have significant contribution in the preparation of this manuscript.

References

Illustrations

Illustration 1

Fig. 1 Preoperative radiograph showing lesion in proximal femur

Illustration 2

Fig. 2 Photomicrograph showing small spindled cells arranged in haphazard array with intervening collagenous tissue (H&E x100)
Illustration 3

Fig. 3 Postoperative radiograph

Illustration 4

Fig. 4 Radiograph at 5 years follow up
Illustration 5

Fig. 5 Clinical photograph at 5 year follow up
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