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Chondrosarcoma of Right Upper Limb: Largest of Its Kind Operated in Our Institute

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ABSTRACT

Chondrosarcoma of extremities is the second most common site for this particular bone tumor. Neoadjuvant chemoradiotherapy is frequently used to down stage tumor for limb-sparing surgery in locally advanced tumor, but chondrosarcoma is relatively resistant to chemotherapy and radiotherapy, and hence sometimes mutilating surgery like forequarter amputation has to be performed. In this case also, patient presented with locally advanced chondrosarcoma of right upper extremity and we had to perform forequarter amputation to achieve adequate clearance.

Keywords: Chondrosarcoma, forequarter amputation

CASE REPORT

A 56-year-old man, presented to our Surgical Oncology OPD with complaints of massive swelling of right upper limb for past 2 years. To begin with, patient had noticed the swelling just below the right shoulder joint that gradually progressed to attain the present size. Patient had visited many oncology centers and he was offered multiple treatments consisting of combination of chemotherapy and radiotherapy, but all were fruitless. He then approached our institute. On examination, the whole of the upper extremity and the right shoulder was engulfed by the tumor (Fig. 1). Confirmation of the type of tumor was done by core cut biopsy, which revealed chondrosarcoma intermediate grade. Computed tomography (CT) scan of chest with upper abdominal cuts was done that revealed large tumoral mass involving the right humerus and scapula with involvement of shoulder girdle muscles. Pectoral muscles were also involved by the tumor. There was no evidence of metastasis in chest as well as in abdomen. According to the Enneking staging, it was Stage IIB. As the patient had already received neoadjuvant chemotherapy and radiotherapy but the tumor size was still increasing, he was planned for forequarter amputation. Patient tolerated the procedure well and the final histopathology report revealed evenly placed anaplastic chondrocytes within lacuna in a chondroid background suggestive of intermediate grade chondrosarcoma with all resection margins free of tumor (Fig. 2). On follow-up after 14 days, stitches



Figure 1. Preoperative figure of the tumor involving whole of the right upper limb and the right shoulder.

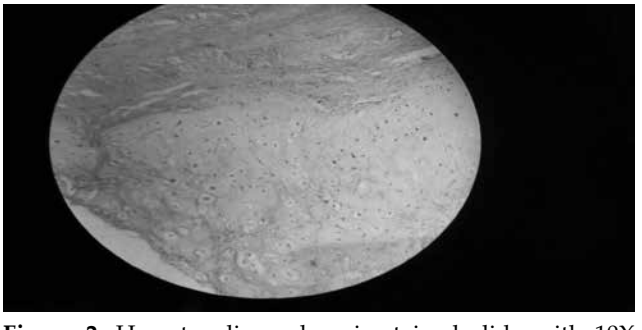


Figure 2. Hematoxylin and eosin-stained slide with 10X power showing evenly placed anaplastic chondrocytes in lacuna within chondroid background.

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Figure 3. Postoperative photograph of the patient after stitch removal.

were removed (Fig. 3), and he was kept on close follow-up to look for recurrence or metastasis. After 1½ years, the patient is still on follow-up and he is disease-free locally as well as in terms of distant metastasis.

DISCUSSION

Chondrosarcoma is a malignant cartilaginous matrix-producing tumor with diverse morphologic features. The peak age of occurrence is between 40 and 70 years. The most common primary sites include the pelvis followed by femur and humerus. Most common presenting symptoms are pain and swelling at the site of tumor. About 90% of chondrosarcomas are primary bone tumors, but around 10% develop from pre-existing osteochondromas, enchondromas or fibrous dysplasia. Chondrosarcomas are graded on a scale from 1 to 3, based upon nuclear size, staining pattern (hyperchromasia), mitotic activity and degree of cellularity. Histological grade is one of the most important indicators of clinical behavior and prognosis. Metastasis in chondrosarcoma depends upon the grade of the tumor. Patients with Grade II and III have higher chances of metastasis compared to Grade I and that can range from 60% to 70%.

Chondrosarcomas are relatively resistant to chemotherapy and radiotherapy and therefore treatment depends upon the completeness of surgical resection. En-bloc resection of the tumor has shown to improve the survival of the patient. Surgical resection margin and the grade of the tumor have been found to be independent prognostic marker for survival of the patient.

This patient who presented to our institute had already received neoadjuvant chemotherapy and radiotherapy to downstage the disease but there was no change. Complete right upper limb along with scapula and shoulder girdle muscle was involved. CT scan revealed no evidence of distant metastasis. Complete en-bloc removal of the tumor, i.e., forequarter amputation was required. Pros and cons of the surgery were discussed and with due consent patient underwent surgery. Now the patient has been kept on follow-up, as the final histopathology revealed intermediate grade chondrosarcoma. Patient has been explained about the risk of distant metastasis. Patient is being followed according to the National Comprehensive Cancer Network (NCCN) guidelines with physical examination, complete blood count and chest as well as local imaging every 3 months for the first 2 years, every 4 months during year 3, every 6 months for years 4 and 5, then annually. It has also been explained that routine post-treatment surveillance has to be extended up to 10 years, as late recurrences can occur.

CONCLUSION

Chondrosarcomas are usually low-grade tumors affecting mainly the pelvic region and the extremities. Complete surgical excision with negative margins is the best treatment to be offered to improve survival. As these tumors are relatively resistant to radiotherapy and chemotherapy, downstaging of the tumor for limb preservation surgery is not possible and patients in locally advanced stage disease have to undergo mutilating surgeries with functional compromise.

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