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Successful treatment of a dedifferentiated chondrosarcoma of the proximal humerus with a hemicortical articular surface sparing allograft: A case report

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ABSTRACT

INTRODUCTION: Dedifferentiated chondrosarcomas are rare and highly malignant tumors that require wide surgical resection. Survival is extremely poor without adequate surgical margins. We present a case of articular sparing surgery of the shoulder for dedifferentiated chondrosarcoma with excellent functional outcomes and no evidence of disease after fifty-six months.

PRESENTATION OF CASE: A 29-year-old male was found to have a non-metastatic right proximal humerus dedifferentiated chondrosarcoma. He underwent resection with clear one centimeter margins sparing the medial cortex and the articular surface. Reconstruction of the bone was accomplished using a hemicortical allograft. A dermal allograft was used to help reconstruct the rotator cuff to the allograft bone. At fifty-six months after surgery he has excellent functional range of motion. His current MSTs, Quick Dash, and Constant Shoulder scores are 29, 2.3, and 80, respectively. He has remained free of disease, is back to work without restrictions and is active in outdoor activities.

DISCUSSION: Dedifferentiated chondrosarcoma has high recurrence and poor survival rates. Adequate surgical resection is vital for its treatment. Previously described reconstructive techniques have consisted of articular replacement with a prosthesis, allograft, or allograft-prosthetic composites. To our knowledge, this is the first report of an articular sparing reconstruction for dedifferentiated chondrosarcoma with fifty-six month survival and functional outcomes. When possible, sparing the articular surface can provide good functional outcomes that improve over time.

CONCLUSION: If adequate surgical margins can be obtained, an articular surface sparing reconstruction of the shoulder can provide effective functional outcomes and an alternative to joint replacement.

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1. Introduction

Chondrosarcomas are rare tumors that can be divided into two categories, primary and secondary. The majority of chondrosarcomas are well-differentiated, however, up to 11% can show transformation and high-grade anaplastic change known as dedifferentiation [1]. Dedifferentiated chondrosarcomas are highly malignant tumors and have reported 5-year survival rates of approximately 13% [2]. As with any tumor, work-up, staging and proper diagnosis are of paramount concern. In dedifferentiated chondrosarcomas, the rate of growth and metastasis is determined

by the non-cartilaginous component. Therefore, it is essential that the tumor be evaluated for dedifferentiation as this has been found to be the most important factor affecting survival [3]. Chondrosarcomas are most commonly located in the proximal femur, scapula, and pelvis with an occurrence rate of 10–15% in the proximal humerus [1,4].

Limb reconstruction in primary malignant bone tumors is a challenging endeavor. Wide surgical margins are necessary to achieve favorable outcomes in dedifferentiated chondrosarcomas [5,6]. The residual tissue deficit can leave patients with significant morbidity and functional deficits, making reconstruction a complex task. There are multiple reconstruction options for the proximal humerus, including prosthetic implants, osteoarticular allografts and allograft-prosthetic composites [7]. The purpose of this case report is to demonstrate the efficacy of a joint preserving reconstruction after resection of a dedifferentiated chondrosarcoma in a 29-year-old male with fifty-six month follow-up. To our knowledge, this is the first case reported of a joint preserving resection of a dedifferentiated chondrosarcoma with both functional outcomes

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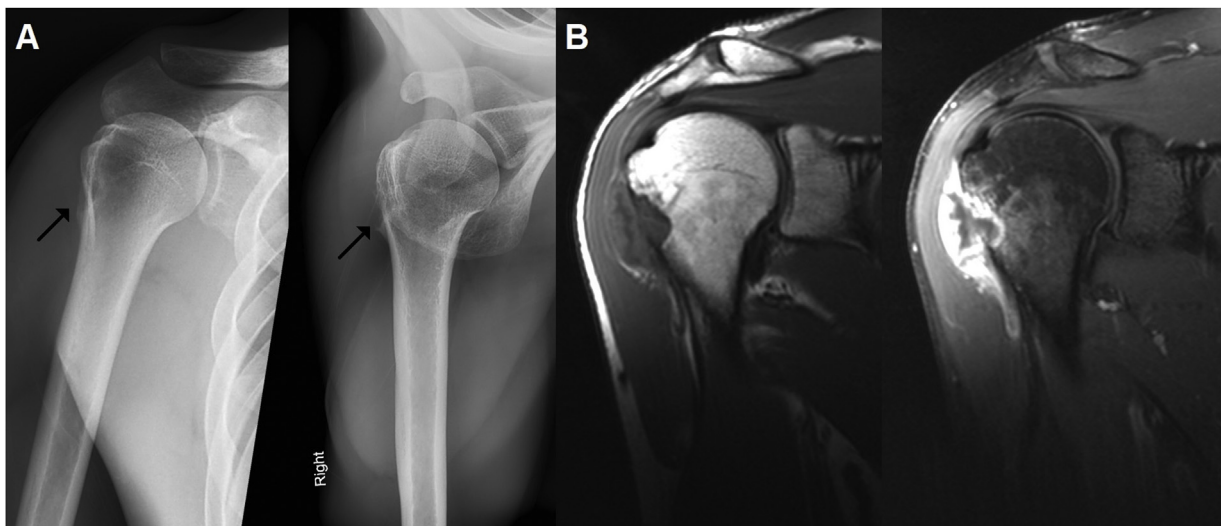


Fig. 1. (A) Initial AP and axillary radiographs of the right shoulder. The arrows demonstrate a lytic area involving the lateral aspect of the proximal humerus with a periosteal reaction. (B) Initial coronal T1- and T1-contrast enhanced MRI's of the right shoulder demonstrating a tumor involving the proximal humerus with cortical breakthrough and an associated soft tissue mass.

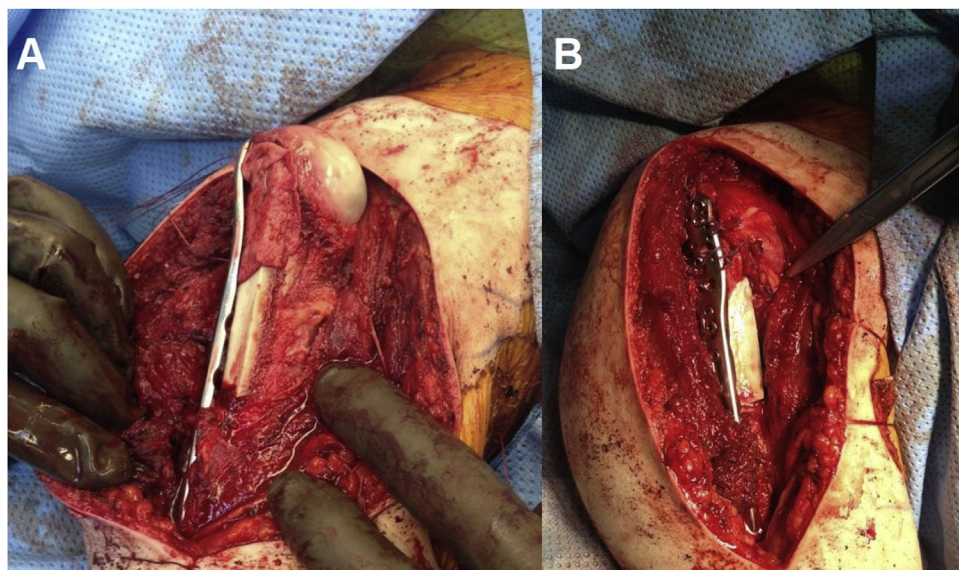


Fig. 2. (A) Intraoperative photograph of the proximal humerus after fixation of the allograft to the host bone. AlloDerm was placed under the plate proximally to provide a surface for repair of the rotator cuff. The articular surface was able to be preserved. (B) Intraoperative photograph of the proximal humerus after repair of the rotator cuff to the AlloDerm. Sutures were also placed through the plate for added fixation.

and longer-term survival. This work has been reported in line with the SCARE criteria [8].

2. Presentation of case

A right-hand dominant 29-year-old male presented with worsening right shoulder pain over a nine-month period. Physical exam revealed a firm, non-mobile mass in his anterior right shoulder. He had no pertinent medical history and had never smoked. He was referred by a local physician to our academic center. X-rays of the shoulder were performed (Fig. 1A) and revealed a lytic lesion with a periosteal reaction over the lateral proximal humerus. An MRI of his humerus revealed an aggressive lesion of the lateral proximal humerus (Fig. 1B) with cortical destruction and an associated soft tissue mass. Laboratory studies were unremarkable. An incisional biopsy was performed with pathology consistent with a dedifferentiated chondrosarcoma. Staging studies with a chest CT

and whole body bone scan demonstrated no evidence of metastatic disease.

The patient underwent wide surgical resection of the tumor through an extended deltopectoral approach. This included resection of the pectoralis major insertion, cephalic vein, proximal long head of the biceps tendon, insertion of the entire rotator cuff, and the anterior one third of the deltoid muscle. The humerus was dislocated and then cut using a high speed burr based on preoperative imaging in order to resect the entire sarcoma with wide margins. The proximal humerus and 5 cm of the lateral humeral shaft were resected. Proximally, the cut was made through the anatomic neck of the humerus, only preserving the articular surface, and distally along the medial cortex.

The defect was then reconstructed with allograft tissue. Bulk tibia allograft was contoured to fit the humeral defect caused by the resection and augmented with cancellous chips mixed with demineralized bone matrix and Trinity (Orthofix/Lewisville) bone

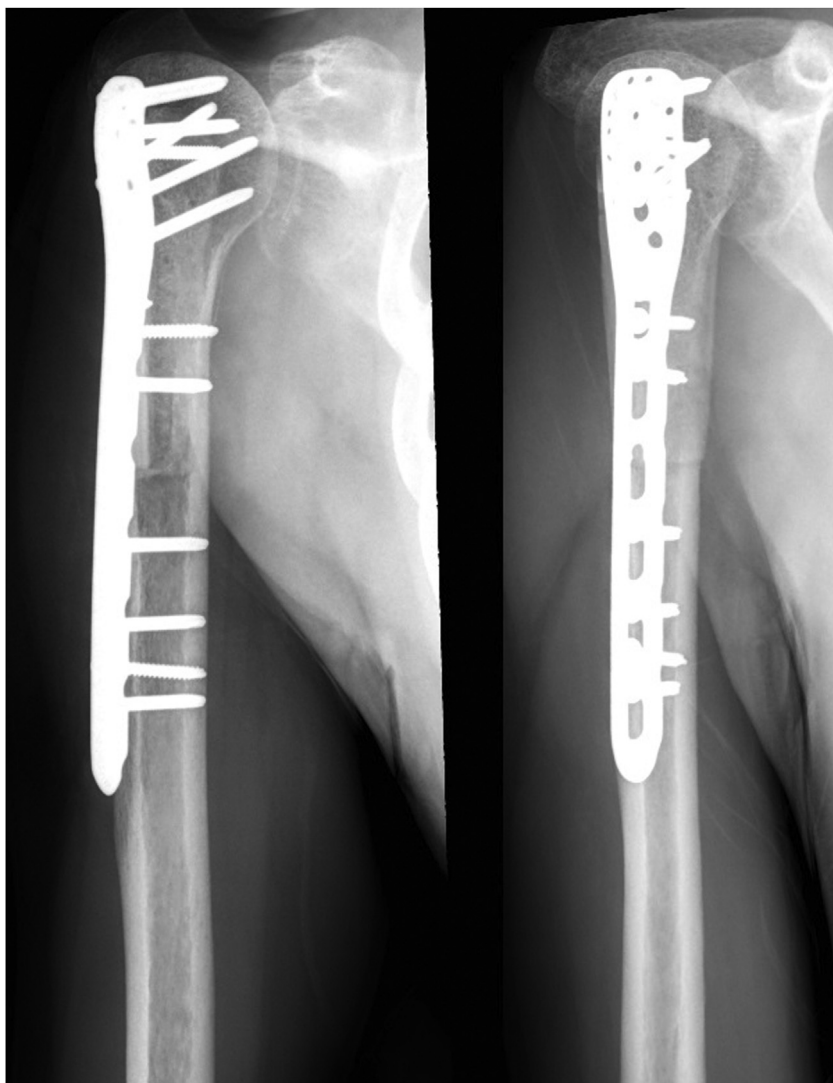


Fig. 3. Postoperative AP and lateral radiographs of the proximal humerus. The hemicortical allograft is demonstrated proximally with spanning plate fixation. The osteotomy site is still identified.

graft. Drill holes were made into the allograft tibia proximally and AlloDerm (Acelity/San Antonio) was attached to the allograft to enhance fixation of the rotator cuff repair to the allograft. A long proximal humerus plate and screws were used to secure and compress the allograft to the host bone (Fig. 2A). The humerus was then relocated. The rotator cuff was repaired to the AlloDerm tissue and the plate in order to reconstruct the deficient rotator cuff tendons. This repair was secured using FiberWire suture (Arthrex/Naples) (Fig. 2B).

The soft tissues were then mobilized to restore function and provide coverage of the plate and allograft. The long head of the biceps was repaired to the short head of the biceps tendon. The pectoralis major was rotated over the plate and repaired to the deltoid. A layered closure of the remaining wound was then performed. Postoperative radiographs are shown in Fig. 3.

Pathologic evaluation of the resection revealed a tumor measuring $3.5 \times 2.5 \times 2.0$ cm in size with a minimum of 1 cm clear margins. Histology demonstrated a low-grade chondrosarcoma juxtaposed with a high-grade spindle cell sarcoma confirming the diagnosis of a dedifferentiated chondrosarcoma (Fig. 4). The patient was subsequently treated with adjuvant chemotherapy with doxorubicin, cisplatin, and ifosfamide for 7 rounds over an 8-month period.

He was monitored for recurrence and metastatic disease with physical examination, humerus radiographs, chest imaging and whole body bone scans. Fifty-six months after surgery, there has been no evidence of metastasis or local recurrence. The allograft has radiographic evidence of incorporation without evidence of failure (Fig. 5).

Initial postoperative therapy consisted of 6 weeks in a shoulder immobilizer with active and passive elbow motion but avoidance of any shoulder motion. Active assist and gentle passive motion with a standard rotator cuff protocol was started at 6 weeks. Light weight bearing was allowed at 3 months and advanced as tolerated to full weight bearing at 6 months. At one year his active shoulder range of motion was 40 degrees of abduction, 20 degrees of forward flexion, and 45 degrees of external rotation. He had full passive range of motion in all planes. His Musculoskeletal Tumor Society (MSTS) score was 28, Quick DASH score was 20.5, and Constant and Murley score was 35. At fifty-six months after surgery, he is pain free and his active shoulder range of motion improved to 65 degrees of abduction, 45 degrees of forward flexion, and 60 degrees of external rotation. His current MSTS score is 29, with one point removed for hand positioning ability, Quick DASH score is 2.3, and Constant and Murley score is 80. He is now back to work as an Orthopaedic Surgeon and has been able to return to back country skiing, sport climbing, mountaineering, and search and rescue.

3. Discussion

Reconstructive techniques of primary malignant bone tumors of the proximal humerus are challenging and controversial. Surgical en-bloc resection is the major curative step for dedifferentiated chondrosarcomas [6,7]. In spite of this aggressive treatment, local recurrence remains up to 50%, and most patients are not alive two years after diagnosis [3]. Lex et al. reported an improved overall survival rate in patients with a dedifferentiated chondrosarcoma of the pelvis where surgical margins greater than 4 mm were achieved [9]. Patients with tumors larger than 8 cm in size, primary involvement of an extremity, inadequate resection, pathologic fracture, and metastatic disease have a worse prognosis [10,11]. The prognosis remains poor even with advanced surgical techniques with a median overall survival of 13.9 months [11]. Although controversial, chemotherapy has shown promise in some studies including an improvement in progression-free survival, but not overall survival when patients were treated with doxorubicin and cisplatin. In this case, the patient is alive and disease free fifty-six months after undergoing surgery with a minimum of a 1 cm surgical margin combined with adjuvant chemotherapy. The small size of the tumor, adequate margins, and completion of chemotherapy are all likely contributors to the patient’s survival.

If limb salvage is the goal, reconstruction after tumor resection is the next step to optimize limb function. Commonly used

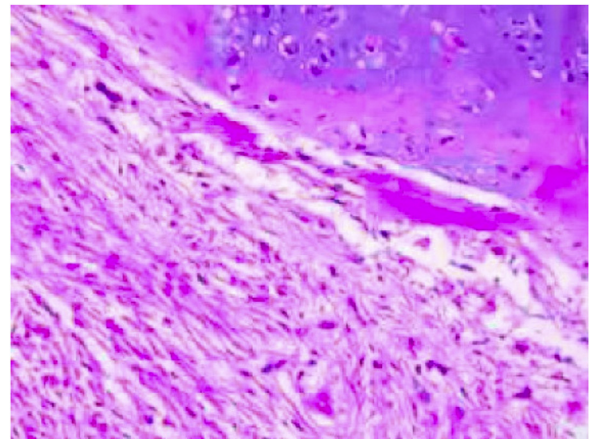


Fig. 4. Histopathology slide of the operative specimen. A low-grade chondrosarcoma in the upper right is juxtaposed with a high-grade spindle cell sarcoma in the lower left consistent with a dedifferentiated chondrosarcoma.

reconstruction options are arthroplasty of some form with or without an allograft component or allograft with surgical fixation [7]. Arthroplasty has been shown to have acceptable clinical outcomes in proximal shoulder reconstruction after tumor resection, but



Fig. 5. Radiographs at final follow-up. AP and lateral radiographs of the proximal humerus demonstrate incorporation of the allograft. The osteotomy site is no longer visualized and there is remodeling of the bone. The articular surface of the humerus is preserved.

implant survival rates, especially in young patients, limit this option [12,13].

Advantages of osteoarticular allograft reconstruction include preservation of joint kinematics, bone stock, and the articular surface [14,15]. There are disadvantages of allograft reconstruction, and the individual patient's prognosis and functional potential should be considered. Ideally host bone incorporates into the allograft by creeping substitution over time [15]. Nonunion between the host bone and allograft has been demonstrated to be a common cause of hardware failure and revision surgery in allograft and allograft-prosthetic composites [16]. Similarly, sections of allograft without ingrowth will weaken over time and possibly fracture [15,17]. In addition to fracture, degeneration of the articular surface is also a common issue [18]. For hemicortical resections and allograft reconstruction, such as the case presented, the most frequent complication cited has been fracture of the host bone with nearly all failures occurring in the first three postoperative years [14].

As shown in a systematic review of proximal humerus reconstructive options by Dubina et al., there is a paucity of literature with regard to preservation of the articular cartilage of the humeral head [19]. To our knowledge this is the first report of an articular sparing surgery of the proximal humerus for a dedifferentiated chondrosarcoma with functional outcomes. Our functional outcomes are on the high end compared to prosthetic reconstructions, and improved compared to osteoarticular reconstructions [12,13,18]. However, given the uniqueness of each tumor and resection, the significance of such comparisons is difficult to ascertain. As shown in our case report, the use of hemicortical allograft with preservation of the articular surface can provide comparable functional outcomes that improve over time.

4. Conclusion

Adequate resection is the most important step in the treatment of dedifferentiated chondrosarcoma. The authors believe that articular sparing surgery should only be attempted when adequate margins can be obtained. Every case is unique in regard to the degree of the resection required, the functional demands of the patient and ultimate prognosis. If the rotator cuff and articular surface can be spared, hemicortical allograft reconstruction allows for the preservation of bone stock and maintains native joint kinematics. This is with the understanding that the primary goal is to resect the tumor and the secondary goal is maximizing the functional status of the patient. This case demonstrates a proximal humerus dedifferentiated chondrosarcoma that was effectively treated with a joint preserving resection without evidence of tumor recurrence or metastasis at fifty-six month follow-up.

Declaration of Competing Interest

The authors state that they have no conflict of interest for this report.

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Ethical approval

This case report is exempt from ethical approval at our institution.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Charles Gomez: Conceptualization, Writing - Original draft preparation and Editing and final approval.

Mark Anderson: Conceptualization, Writing - Original draft, Editing and Review, and final approval.

Scott Epperly: Conceptualization, Writing - Review and Editing, Investigation, Visualization, and final approval.

Lee Zuckerman: Treating surgeon, Conceptualization, Writing - Review and Editing, Supervision, and final approval.

Registration of research studies

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Guarantor

Lee M. Zuckerman, M.D.

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