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Distal ulna leiomyosarcoma treated with custom polymethylmethacrylate prosthesis with a 4-year follow-up

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Abstract
Background Intraosseous leiomyosarcoma is a rare neoplasm having an aggressive biologic behavior. The distal end of the ulna is a very uncommon site for this type of primary bone tumor frequently mistaken for benign lesions. En bloc resection of the distal ulna with reconstruction is a valid option as a limb salvage procedure for the treatment of this difficult problem, minimizing local recurrence while preserving hand function.
Case description We present an unusual case of a 63-year-old woman with a primary leiomyosarcoma arising from the distal end of the ulna treated successfully with a wide excision and custom distal ulna, with 4-year follow-up and no recurrence.
Literature review Tumors to develop at the distal end of the ulna have been reported as part of large series such as Dahlin and few case reports. According to Cooney, Exner, and Mankin, reconstruction for distal ulnar neoplasms is not necessary to maintain function. However, Noble and Laurentin-Perez disagree because stabilization of the distal ulna following large resection, as in our case, can be a significant problem with associated pain and weakness due to a decreased interosseous space with ulnar stump impingement on the radial metaphysis and ulnar translation of the carpus.
Clinical relevance Custom methacrylate in situ radioulnar joint prosthesis for reconstruction of a large segment of the distal ulna can be a valid option to reestablish the mechanical continuity of the forearm, reducing pain and improving strength and function.

Statement of location This work was done at The University of Arizona, Department of Orthopaedic Surgery.

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Introduction

Primary intraosseous leiomyosarcoma is a rare malignant spindle cell tumor accounting for <1 % of all primary bone tumors and 0.7 % of all adult cancers [1, 4, 11, 21–24, 32]. In fact, it was not even reported in Dahlin's [7] classic article of malignant tumors involving bone of nonosseous origin. It is characterized by the expression of desmin, smooth muscle actin, cytokeratin, calponin, and other markers indicating smooth muscle cell origin [3, 4, 11, 15, 23, 32]. Usually, these tumors involve the femur, tibia, ilium, and pubis [4, 23, 32]. Radiographically, they present as osteolytic lesions that make them nonspecific, requiring biopsy and immunohistochemistry for the final diagnosis [4, 19, 24, 32].

Wide resection with clear margins of the distal end of the ulna, with or without reconstruction, has been described for the treatment of giant cell tumor, the most common distal ulnar neoplasm. To our knowledge, there is no case of leiomyosarcoma arising from the distal end of the ulna, making its reconstruction after ulnar excision a more challenging treatment. Although a custom fabricated oncological prosthesis may be available, stabilization of the ulnar stump has been described for painful distal radioulnar joint instability following inflammatory arthritis and posttraumatic events with tendon reconstruction, allograft replacement, and arthroplasty with mixed outcomes. [5–10, 13, 26, 30, 33]

The purpose of this report is to present a unique intraosseous leiomyosarcoma that was treated successfully with wide resection and a custom made intramedullary Rush rod with a methacrylate ulna.

Case report

A 63-year-old woman presented with a history of a painful mass involving her left wrist increasing in size for 4 months. Past medical history included cryosurgery of the cervix, hypothyroidism, insomnia, liver hemangiomata, bilateral adrenal adenomas, and seasonal affective disorder. Physical examination of her forearm revealed a diffuse fusiform swelling along the ulnar border of her left wrist, as well as soft tissue swelling on the volar aspect. Forearm rotation was without restrictions. Neurological examination and hand function were normal.

Plain films of the left forearm and wrist revealed an expansile, permeative lytic lesion involving the distal 5 cm of the ulna (Fig. 1). Chest X-rays were unremarkable. A bone scan showed a solitary focus of increased activity at the distal ulna corresponding to the lesion seen on plain films. A positron emission tomography (PET) scan of her left wrist lesion showed avidness for fluorodeoxyglucose, whereas lesions in the abdomen showed no evidence of PET-fluorodeoxyglucose avidness. Contrast enhanced T2-weighted magnetic resonance imaging of the left wrist revealed an intraosseous lesion in the distal ulna centered in the medullary cavity with a small area of cortical breakthrough distally and ulnarly, periosteal reaction, and a soft tissue component (Figs. 2 and 3). A CT-guided biopsy of her forearm lesion was suggestive of a spindle cell sarcoma with focal areas of osteoid production consistent with probable leiomyosarcoma. Immunohistochemistry was positive for smooth muscle actin and caldesmon. Preoperative CT scan suggested that approximately 7 cm of ulna was involved, with potential involvement of the extensor carpi ulnaris musculature.

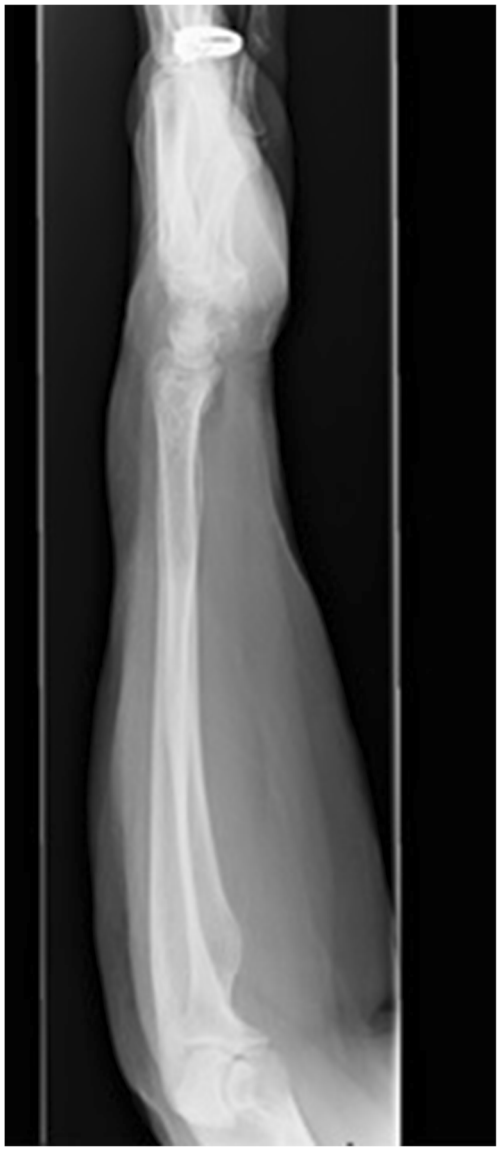

Fig. 1 Lateral radiograph of the left forearm and wrist revealed an expansile, permeated lytic lesion involving the distal 5 cm of the ulna

Operative technique

After placed in the supine position her arm was draped and later elevated for 5 min and the tourniquet inflated to 250 mmHg. The previous biopsy needle site was elliptically excised with a longitudinal ulnar incision. The skin flaps were elevated anteriorly and posteriorly. C-arm image intensification delineated the proximal extent of the anticipated osteotomy site proximal to the lesion. The ulna was osteotomized and endosteal curretings proximal and distal were sent for frozen section, which were negative. This suggested that the osteotomy level was sufficiently proximal to avoid tumor. The sixth dorsal compartment was entered, and the extensor carpi ulnaris tendon was isolated. The muscle was removed from the tendon and left with a deep portion of the sixth dorsal compartment. Volarly, the ulnar nerve was identified and protected.

The musculature of the flexor carpi ulnaris was left with the ulna and removed from the tendon up to the level of the osteotomy. There did not appear to be tumor within the radioulnar joint. The ulnar collateral ligament (UCL) and triangular fibrocartilage complex (TFCC) were transected at the level of the fovea of the ulnar styloid. This allowed for complete mobilization and resection of the distal ulna and lesion with a clear margin of tissue (Fig. 3). Various margins volar, dorsal, proximal, and distal were delineated on the pathology report, the specimen was then submitted. Frozen section margins were clear.

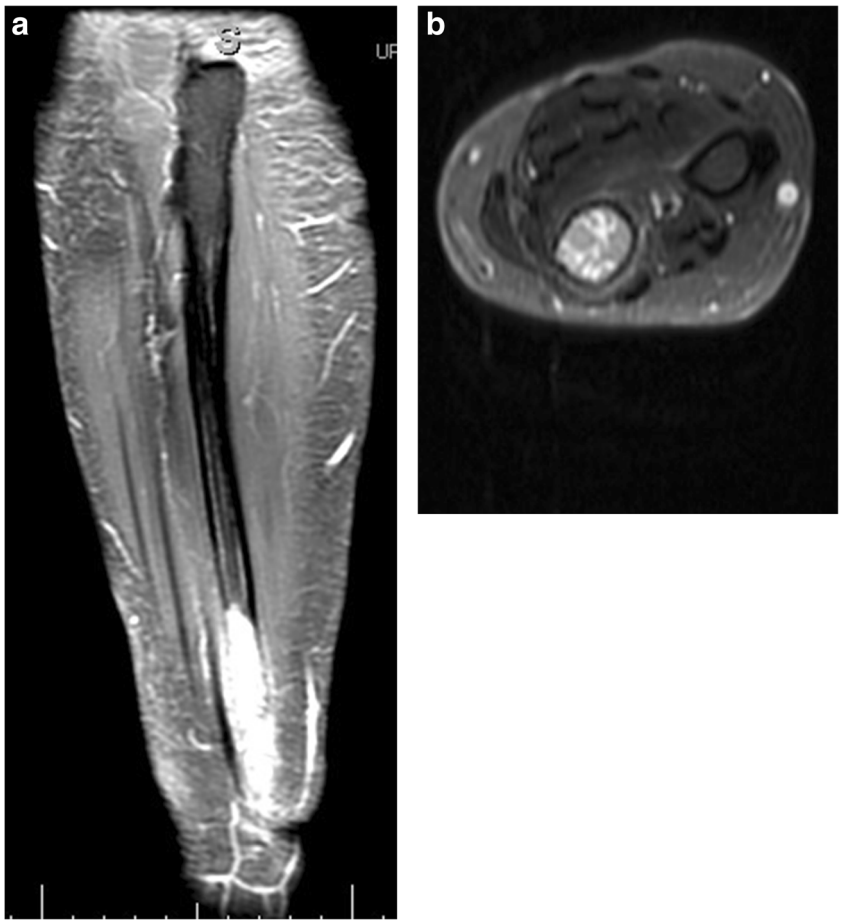

Fig. 2 **a**, **b** Contrast enhanced axial T2 weighted (**a**) and Cor T1 MRI (**b**) of the left forearm revealed an intraosseous intramedullary lesion in the distal ulna, with distal and ulnar areas of cortical breakthrough, periosteal reaction, and minimal extension of the mass into the surrounding soft tissue component, suggesting a potential for clean margins with wide excision

On the back table, the fabrication of a distal ulna reconstruction consisted of a small Rush rod with a chest tube measuring 8 cm, the approximate length of the ulna was made. A finger from a glove was sutured over the Rush rod and chest tube to contain the methylmethacrylate until it hardened. Number 2 Ethibond was placed through the glove–polymethylmethacrylate (PMMA) construct prior to hardening for subsequent TFCC repair. The chest tube and glove were then removed. A simulated distal ulna was therefore created. The implant was inserted with a trial reduction, and additional length had to be removed from the proximal portion to accommodate the defect. Once this was satisfactorily contoured and fashioned, the Rush rod was inserted until hardened, into the distal ulna (Fig. 4). Prior to hardening of the ulnar implant, no. 2 Ethibond suture was placed through the head of the ulna for repair of the ulnar collateral ligament and TFCC. Once the cement had hardened, the TFCC and ulnar capsule were then repaired with the no. 2 Ethibond. The remaining capsule was sutured and reinforced with the radial half of the ECU tendon, longitudinally split proximal of the new distal end of the ulna. The tourniquet was released and removed, and hemostasis was ensured. The subcutaneous tissue was then closed and sterile staples were used in the skin. Forearm rotation was observed through an arc of approximately 180 ° of pronosupination. Following surgery, the patient completed a 6-week course of adjuvant radiotherapy and forearm immobilization while using a removable Münster splint in supination.

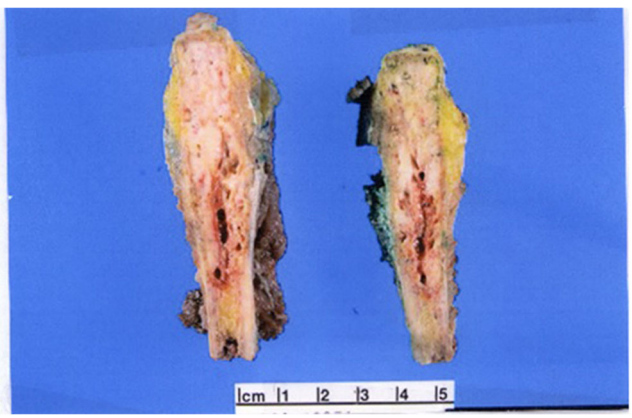

Fig. 3 Resection of the distal ulna and lesion within a clear margin of tissue. Pathology report indicated a 6.5×1.5×1.5 cm lesion with low nuclear pleomorphism and low mitotic activity, with no tumor necrosis present

At 4-year follow-up (Figs. 5 and 6), the patient, was pain free with no evidence of infection or distal radioulnar joint (DRUJ) instability, or nerve dysfunction, and has a well-

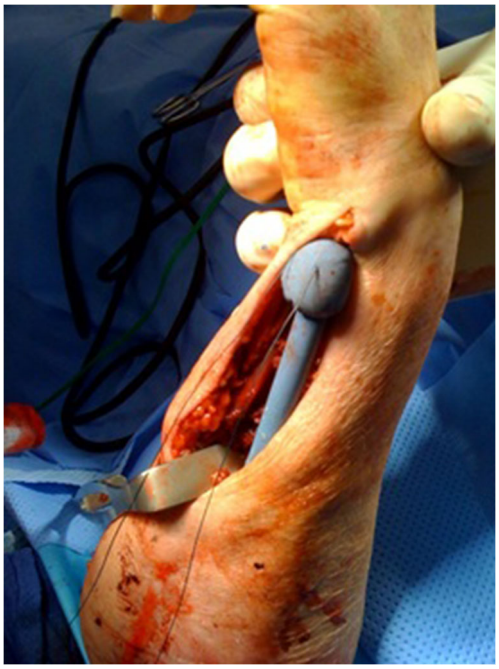

Fig. 4 Insertion of the methacrylate custom PMMA distal ulna

functioning left forearm with full range of motion and strength. There is no evidence of recurrence or metastasis on plain films, follow-up MRI, and PET scans.

Discussion

Leiomyosarcoma is an aggressive sarcoma thought to arise from vascular smooth muscle cells. It may either occur in soft tissue (uterus or gastrointestinal) or in bone [11]. Less than 120 cases of extra-facial leiomyosarcoma have been reported presenting in the fourth through eighth decades of life [4, 26, 32]. Most occur in the metaphysis of long bones, presenting usually in the femur, tibia, humerus, and ilium [32].

When a leiomyosarcoma presents in bone, one must first consider metastatic bone disease [11, 21]. In this case, the neoplasm was diagnosed and localized to the left ulna confirmed on the basis of preoperative staging studies including X-rays and MRI. Leiomyosarcoma of bone does not have a typical radiographic appearance and can mimic any other primary tumor [10, 24, 30]. Interestingly, this patient's lesions in the abdomen were not related to the lesion of the wrist due to the fluorodeoxyglucose PET avidness. MRI images of the liver were compared to an MRI scan obtained 3 years prior and appeared to have been present at that time and were essentially stable, suggesting that they were not metastatic disease.

The distal end of the ulna is an uncommon site for a sarcoma [6, 7, 10, 17, 19, 30]. Reported tumors to develop in this area include intraosseous ganglia and osteochondromas with fewer cases of fibromatosis of bone, desmoplastic fibromas, giant cell tumor, and traumatic epidermoid inclusion cysts [1, 5, 8, 10, 12, 13, 30, 33–35]. Functionally, the distal end of the ulna helps in both rotation of the forearm and grip strength, as well as in maintaining the relationship between the carpus and the distal end of the radius [5, 6, 17, 25]. In the maintenance of this anatomical relationship, the UCL and the TFCC play an important role [2, 10, 12–14, 19, 33]. Thus, resecting the distal part of the ulna may result in loss of ulnocarpal stability with dynamic radioulnar convergence and unstable rotation of the carpus around the ulnar axis causing painful snapping, carpal subluxation, or tendon rupture due to the loss of osseous support for the TFCC, as well as the volar and dorsal radioulnar ligaments [6, 25, 33].

Surgical excision with wide margins remains the gold standard for definitive management of leiomyosarcomas [14, 35]. Amputation is usually reserved for tumors with extensive soft tissue involving the neurovasculature. According to Cooney et al. [13, 19], Exner et al. [2, 16–18, 35], Wolfe et al. and Harness [2, 19], and Mankin [14], reconstruction for distal ulnar neoplasms is usually not necessary to maintain function. However, Noble [13, 25, 35], Laurentin-Perez [13, 16], and Bieber [16] disagree because stabilization of the distal ulna following large

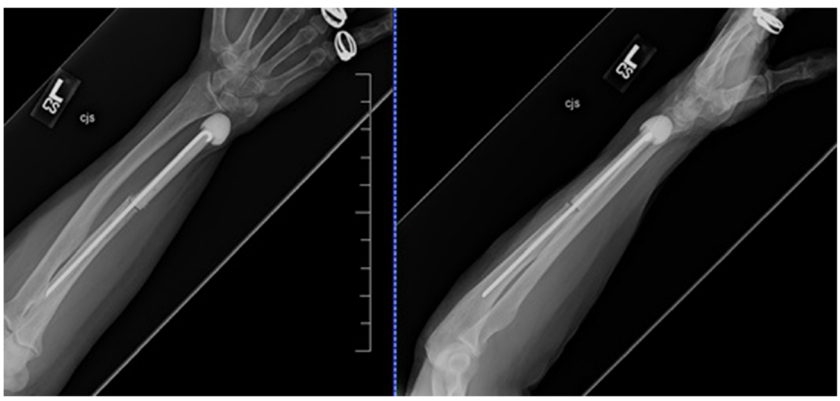

Fig. 5 Radiographs of the left forearm at 4-year follow-up showing no evidence of distal radioulnar joint instability, recurrence or metastasis

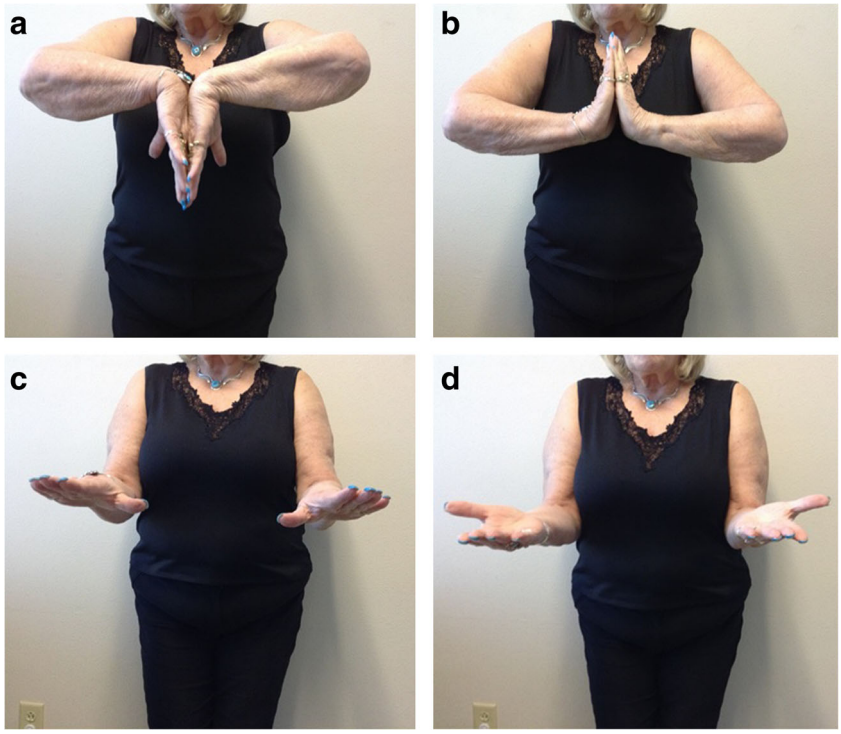

Fig. 6 **a–d** Clinical range of motion at 4-year follow-up

resection, as in our case, can be a significant problem with associated pain and weakness due to a decreased interosseous space with ulnar stump impingement on the radial metaphysis and ulnar translation of the carpus.

Several DRUJ prostheses have been developed to replace the ulnar head in patients with pain related to distal ulnar resection and post-traumatic arthritis. However, most of them are designed to be used in patients with intact soft tissue and stabilizing ligaments at the DRUJ. These devices, therefore, are not appropriate for use in patients who have undergone resection of the DRUJ because anteroposterior stability is not fully restored and suturing soft tissue limits mobility [20, 28, 29, 31]; reinsertion of the soft tissues at the metallic head may be difficult in patients who have undergone several surgical interventions, and to improve the adaptation of the implant, it may be necessary to remodel and deepen the sigmoid notch of the radius [27].

Moreover, when all of the soft tissue support is removed from the distal ulna (TFCC and the interosseous membrane), a constrained device should be used to substitute for this deficiency and to address the gross instability of the remaining ulna caused by a large segmental resection [10, 17].

An oncological prosthesis is currently not available to reconstruct the distal ulna following resection of a tumor. The Scheker DRUJ prosthesis is an option when the distal ulna is removed. Gracia et al. and Pirela-Cruz et al. [17, 22] treated a giant cell tumor of the distal ulna using an APTIS MEDICAL and a Scheker implant to treat a defect of 10 and 7.5 cm, respectively, both in whom rehabilitation played a key role. Without rehabilitation, Shipley et al. [36] have found that the implant fails primarily when it becomes unstable dorsally. In our case, the patient did not have access to a physical therapist, leaving us with the option to create a custom implant.

There have been some concerns that have been expressed regarding the use of this prosthesis, which are similar concerns with all prosthesis about the wrist. All questions are valid concerns, but currently, the reconstructive options for the DRUJ after a tumor in particular are limited; all have their own intrinsic clinical problems. Thus, the search continues to find a reconstructive procedure for the DRUJ that is consistent and acceptable for both the patient and physician [22].

This case demonstrates that a custom-made prosthesis can be used successfully to reconstruct and stabilize the distal radioulnar joint following the resection of a large tumor. This case report is the only documented case of a distal ulna primary leiomyosarcoma with 4-year follow-up, no recurrence, and normal hand function. Although long-term studies are required, custom methacrylate in situ radioulnar joint prosthesis for reconstruction of a large segment of the distal ulna can be a valid option to reestablish the mechanical continuity of the forearm, reducing pain and improving strength and function.

Conflict of interest Jose Arturo Pacheco-Nuñez, Joseph E. Sheppard and Andrew P. Mahoney declare that they have no conflict of interest.

Statement of human and animal rights This article does not contain any studies with human or animal subjects.

Statement of informed consent Informed consent was not obtained as there are no patients included in the study.

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