Adamantinoma of the Forearm: A Chronologic Presentation of a Slow-Growing Malignancy

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Abstract

The purpose of this report is to present a unique case of an uncommon tumor. Adamantinomas are inherently rare, low-grade malignant primary bone tumors that almost always involve the tibia. Although cases have been reported of these tumors involving the long bones of the upper extremity, we present an instance in which both bones of the forearm were affected with documented radiographic changes over a 7-year span. Surgical management is the mainstay of treatment for these tumors and usually requires limb reconstruction following resection. We also present an interesting reconstruction option for upper extremity limb salvage following surgical resection of these tumors.

Adamantinomas are slow-growing, low-grade malignant tumors usually found in the long bones. These tumors were first described in 1900. In 1913, Fischer coined the term “adamantinoma.” These tumors are uncommon, representing less than 1% of all malignant bone tumors. They are characterized as affecting the tibia in 80%–90% of cases and usually occur in patients in the second and third decades of life.

Although adamantinomas show a predilection for involvement of the tibia, other skeletal sites have been reported, including the femur, radius, and ulna. Because of their malignant characteristics, adamantinomas of the long bones are usually treated with wide resection and reconstruction. Surveillance for distant recurrence when treating these tumors is mandatory as metastases are observed in 10%–30% of cases.

Reports of involvement of the radius or ulna are limited to fewer than 10 cases, and we are unaware of any cases with simultaneous involvement of the radius and ulna. We report a unique case of an upper extremity adamantinoma ultimately affecting both the radius and ulna after a long period of preoperative observation. Treatment, therefore, included a wide resection of both the proximal radius and ulna and a subsequent reconstruction of the proximal ulna and ulnohumeral joint using an allograft-prosthesis composite. The aspects of this case—including its unique location, histopathology, and reconstruction using a total elbow arthroplasty-allograft composite—are discussed in the following sections.

Case Description

A 60-year-old, right-handed woman was referred to our institution for evaluation of a right forearm mass. Her medical history was significant for thyroid cancer in the remote past and a longstanding radiographic diagnosis of fibrous dysplasia of the right ulna. At the time of presentation, her main complaint was an increase in right forearm pain and of swelling and progressive decrease in her functional range of motion. She had received no other treatment beyond X-ray imaging before this presentation, and the progressive nature of the changes prompted referral (Figs. 1 and 2).

Physical exam revealed asymmetry of her right proximal forearm as compared to the contralateral side. A palpable mass was observed that appeared to be nonmobile. The flex-extension arc for elbow range of motion was observed to be 25 degrees of extension to 100 degrees in flexion, 0 degrees of pronation, and 20 degrees of supination. Neurovascular examination of the right upper extremity was unremarkable.
Earliest available radiographs of the right forearm were from an outside visit 6 years before her presentation (Fig. 1). When compared to current radiographs, the stark changes were consistent with a very narrow differential diagnosis of adamantinoma, osteofibrous dysplasia, metastatic disease from her thyroid cancer history, or other primary malignancy of bone.

The severity of these diagnoses prompted magnetic resonance imaging (MRI) of the right forearm, a computer tomography (CT) scan of the thorax, and a whole-body nuclear medicine bone scan under the belief that the findings were suggestive of a malignancy. The CT scan of the thorax and bone scan were negative. The MRI of the right forearm, as seen in Figure 3, showed involvement of the ulna and radius with an extensive soft tissue mass. A CT-guided needle biopsy was then performed, and pathology was consistent with an adamantinoma of the radius and ulna with osteofibrous dysplasia-like areas.
The patient was offered continued observation vs. limb ablation vs. surgical intervention to include wide resection and reconstruction of the right forearm. The patient elected to pursue limb salvage with wide resection of both the radius and the ulna along with allograft prosthetic reconstruction of the ulnohumeral joint.

**Surgical Technique**

The surgical technique required a posterior incision that coursed from the distal third of the posterior humerus to the distal ulnar border of the forearm. Given the extensive soft tissue mass, a volar forearm incision was also used to access the radius and help provide visualization of the

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**Figure 4**

Immediate postoperative anteroposterior (A) and lateral (B) forearm radiographs showing the total elbow arthroplasty-allograft composite. Sixteen-month follow-up lateral forearm (C), anteroposterior and oblique (D), and lateral elbow (E) radiographs show stable appearance of the allograft-prosthesis composite and a well-healed distal allograft-host bone junction.
involved interosseous membrane and neurovascular structures. The mass was removed en bloc following soft-tissue dissection of it, osteotomies of both ulna and radius, and transection of the triceps insertion on the proximal ulna.

Planned reconstruction involved an allograft-prosthetic reconstruction. An allograft with dimensions similar to the patient’s native bone was obtained from the Musculoskeletal Transplant Foundation. The allograft was an entire ulna with its soft tissue attachments.

A total elbow arthroplasty was cemented into the patient’s native humerus and the ulna allograft. The triceps mechanism of the elbow was reconstructed by suturing the remaining native triceps to the allograft triceps remnant. The distal tendon of the biceps was sutured into the remaining anterior soft tissues of the ulna allograft. Distally, the ulnar allograft-host bone junction was secured via compression plating with a plate and screw construct (Fig. 4). The wound was irrigated and closed, and the patient was placed in a long arm splint for 2 weeks.

**Postoperative Course**

Clear surgical margins were obtained. Both gross and histologic examinations were performed by a musculoskeletal pathologist. Histologic section showed large multilobulated nests of adamantinoma composed of cords of epithelial-like cells in a myxoid stroma. In addition, osteofibrous dysplasia-like areas were present. Focal high-grade areas exhibiting more epithelioid features in cellularity were also identified (Fig. 5). Simple areas of fibrous dysplasia were not reported.

Postoperatively, the patient had pronation to 90 degrees, supination to neutral, and a 105-degree flexion-extension arc. The patient had allograft-host bone junction healing at 10 months. Follow-up at 16 months has shown no evidence of local recurrence or metastasis.

**Discussion**

This report outlines a unique case regarding an adamantinoma of the upper extremity involving both the ulna and radius. Although adamantinomas affecting the radius or ulna have been reported, we are unaware of any with simultaneous involvement of both bones of the forearm after such a long period of observation. Further, the total elbow allograft prosthetic composite secured with compression plating techniques presents an interesting method of reconstruction.
As mentioned above, these tumors are usually found in the anterior cortex of the tibia. The presentation of these tumors is analogous to the presentation in this patient with pain, swelling, and/or deformity. Secondary to the tumor’s inherent malignant characteristics and insensitivity to chemotherapy or radiation, treatment entails surgical management. En bloc resection followed by the appropriate reconstruction is the usual pathway for surgical treatment.

Studies reporting patient survival are limited to small patient numbers because of the rarity of the tumor. Published mortality rates related to adamantinomas, however, have been reported to range from 13%–18%; the rate of metastases ranges from 15%–30%. Additionally, local recurrence of these tumors can occur 5–15 years after diagnoses, mandating that these patients be followed for an indefinite time.

Adamantinomas isolated to either the radius or ulna have been reported previously in the literature. Bourne et al reported on an adamantinoma involving the radius treated with en bloc excision and reconstruction using a vascularized fibula graft. Keeney et al studied 85 cases of adamantinomas located in various anatomic locations treated at their institution—only 2 involved the ulna and 1 the radius. Gianoutsos et al and Sherman et al published reports of adamantinomas of the ulna, while Soucacos reported on a case of an adamantinoma of the olecranon; this latter case was unique for both its location and its subsequent bony metastases. However, none of the above reports illustrated the chronological progression of disease or the simultaneous involvement of both bone of the forearm that our case presents.

Conclusion

Our case report illustrates a unique presentation of an adamantinoma of the upper extremity. This case is not only unique for its areas of involvement, but also its progression before and after referral to our institution. These tumors are traditionally slow-growing; however, our report was able to chronologically follow the progression of this particular patient’s malignancy, ultimately treated with en bloc resection and a unique reconstruction method.

References