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Case Report

Surgical Reconstruction Methods following Radical Excision of Distal Ulna Osteosarcoma in Both Skeletally Mature and Immature Patients

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Distal radioulnar joint · Osteosarcoma · Reconstruction · Hand surgery · Prosthesis

Abstract
The distal ulna has always been considered to be expendable and its removal has been advocated for a variety of post-traumatic degenerative and oncological conditions but recent studies showed that the distal radioulnar joint allows supination and protonation of the forearm and is important to one's grip strength and lifting ability. Several prosthesis models have already been made to replace the mechanical functionality of the distal radioulnar joint. We present two cases of females aged 22 and 12 years, respectively, who presented with wrist pain and swelling without any history of trauma and with terminal degree limitation in wrist movements due to tenderness and swelling. Both of them did not have any distant metastasis upon radiographic staging. The skeletally mature patient underwent radical excision of the distal ulnar osteosarcoma and received a distal radioulnar joint replacement prosthesis (Scheker prosthesis). The other skeletally immature patient underwent radical excision of the involved distal ulnar osteosarcoma with stabilization of the residual ulnar stump using the extensor carpi ulnaris sling in a modified version of the Goldner and Hayes technique. Both of our patients were treated according to the protocols of our multidisciplinary clinic sarcoma team by starting with neoadjuvant chemotherapy, followed by surgery and adjuvant chemotherapy. Both registered an almost complete restoration of the normal wrist and hand function and were in complete remission for 26 and 24 months, respectively. Based on our literature review, these are some of the extremely rare cases in which the osteosarcoma affected an unusual site (the distal ulna where they underwent a rare type of reconstruction status following radical excision of a malignant tumor).

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Introduction

Our report attracts attention to the fact that osteosarcoma of the distal ulna is an extremely rare entity; therefore, there are no clear guidelines about the preferred surgical option in managing and reconstructing the distal radio-ulnar joint (DRUJ) post-tumor excision, especially focusing on the DRUJ surgical reconstruction differences between adults and pediatric patients. We have expanded the usage of the non-oncology DRUJ prosthesis (Scheker prosthesis) to reconstruct our skeletally mature patient following her tumor resection (oncology case), to the fact that we know this form of reconstruction was considered the first to be performed in the Middle East, confirmed by the providing company APTIS®, whereas the other skeletally immature patient underwent radical excision of the involved distal ulna osteosarcoma with stabilization of the residual ulnar stump using the extensor carpi ulnaris sling. Both of our patients had an almost fully intact hand and wrist function and were able to resume their daily activities after physiotherapy.

Case No. 1

A 22-year-old female patient presented with an 8-month history of left wrist pain and swelling with a negative history of trauma. On physical examination, she had localized tenderness and swelling, which was evident on the dorsal aspect of the distal ulna, as well as terminal degree limitation in wrist movements due to tenderness and swelling. Radiological workup including X-ray and magnetic resonance imaging (MRI) showed a metaphyseal, ill-defined lesion originating from the distal ulna with a periosteal reaction, osteoid matrix and cortical disruption with extension to involve the distal epiphysis with a soft tissue component measuring 1.8 × 0.7 × 3 cm. Metastatic workup with a chest computed tomography (CT) scan showed no metastasis. Needle core biopsy from the mass came back with the diagnosis of osteosarcoma (Fig. 1 a, b). The patient and her family were counseled about the diagnosis and a sarcoma multidisciplinary team explained the treatment plan including 2 cycles of cisplatin and doxorubicin followed by local surgical excision. MRI following neoadjuvant treatment showed no significant interval changes regarding the destructive lesion and the patient was admitted for the planned resection. The preoperative templated prosthesis was done by APTIS Medical® (Glenview, KY, USA) using the X-ray markers for scale; the template demonstrated a size 10 radial plate with a 4-cm extended ulnar stem with a 4.0-mm diameter (Fig. 2).

Under a bloodless field without exsanguination, we performed the standard hockey-stick incision on the dorsal and ulnar aspect as we incorporated the biopsy tract with our incision, raised the retinacular flap and preserved the extensor carpi ulnaris (ECU). We were able to dissect around the tumor of the distal ulna from distal to proximal without violating the tumor as well as preserving the major neurovascular structure and the remainder of tendons. Resection of the ulna was 6 cm proximal to the ulnar styloid process, samples for a frozen section from the proximal medullary canal and radial periosteum were sent and came back negative for malignancy (Fig. 3a). Then, preparation of the radial plate was performed by removing the volar lip to adequately seat the radial plate precisely along the ulnar border of the radius without any dorsal angulation or prominence, the plate was secured by k-wires and checked with C-arm. After insuring a satisfactory position, we applied a radial plate size 10 according to the preoperative template with two locking screws after inserting one compression screw without violating the volar or dorsal cortex of the radius to avoid any future possible irritation similar to De Quervain's, shifting to ulna preparation the small medullary canal approached with extra care when predrilled. Copious amounts of irrigation were used to keep the drill bit and bone from overheating. Also, regular removal and cleaning

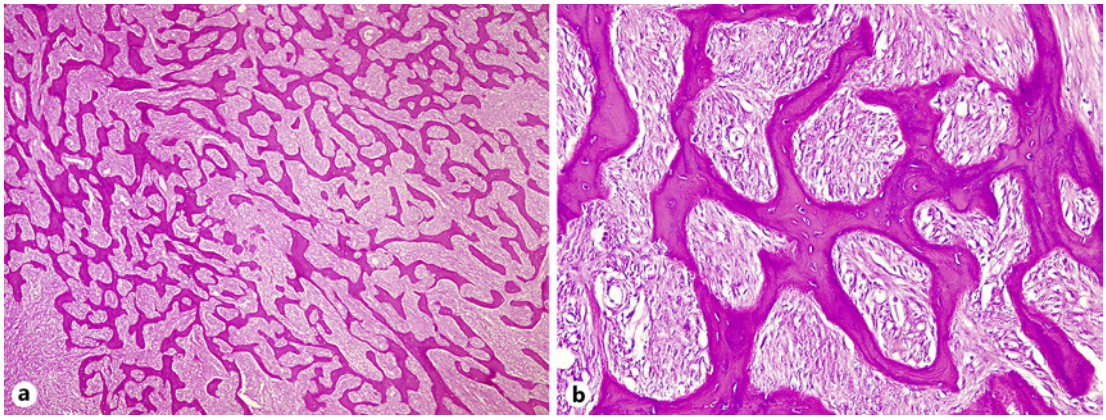

Fig. 1. Histological images showing the diagnosis of osteosarcoma. Microscopic examination reveals well-formed bone trabeculae with hypercellular fibrous intertrabecular areas containing large atypical cells with prominent nucleoli confirming the diagnosis of osteosarcoma. No necrosis was seen. **a** H&E. ×10. **b** H&E. ×40.

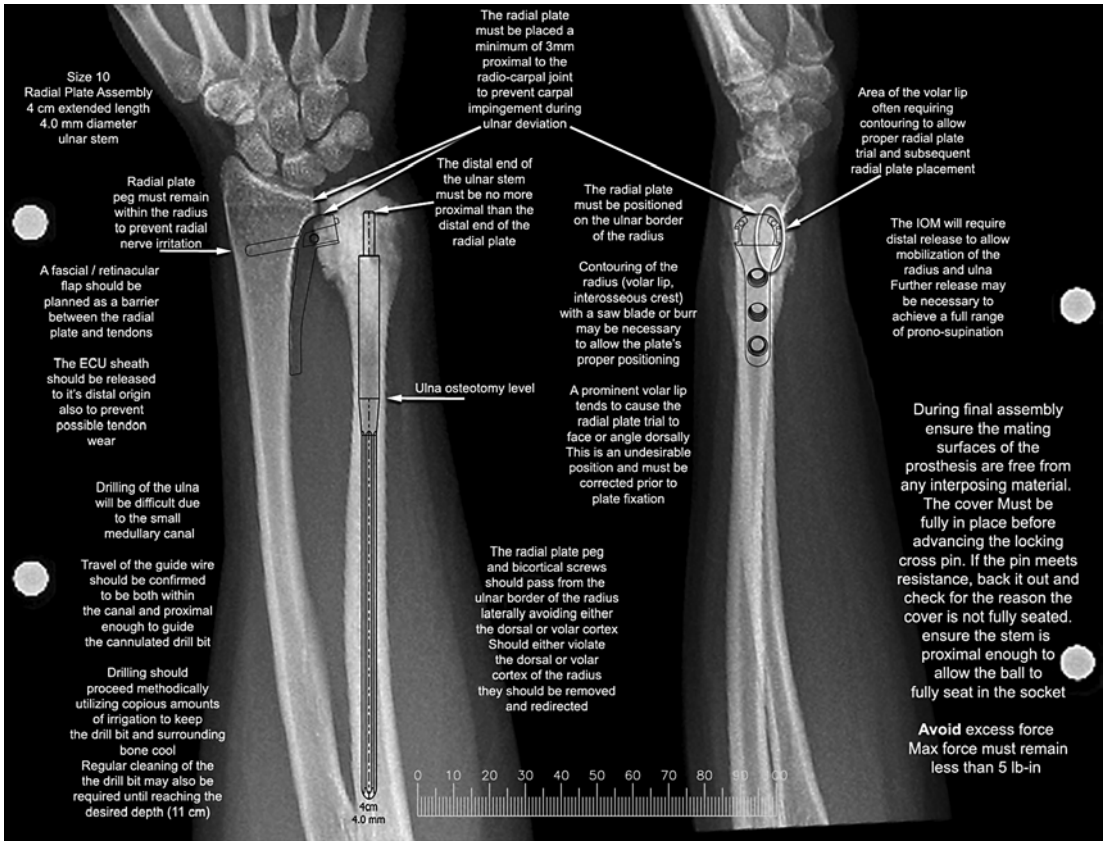

Fig. 2. X-ray of the left upper limb [anterior-posterior (left), lateral (right)], showing a lesion on the distal ulnar metaphyseal area and APTIS medical ® integrated a pre-operative prosthesis templating showing where the various parts of the Scheker prosthesis are to be implanted.

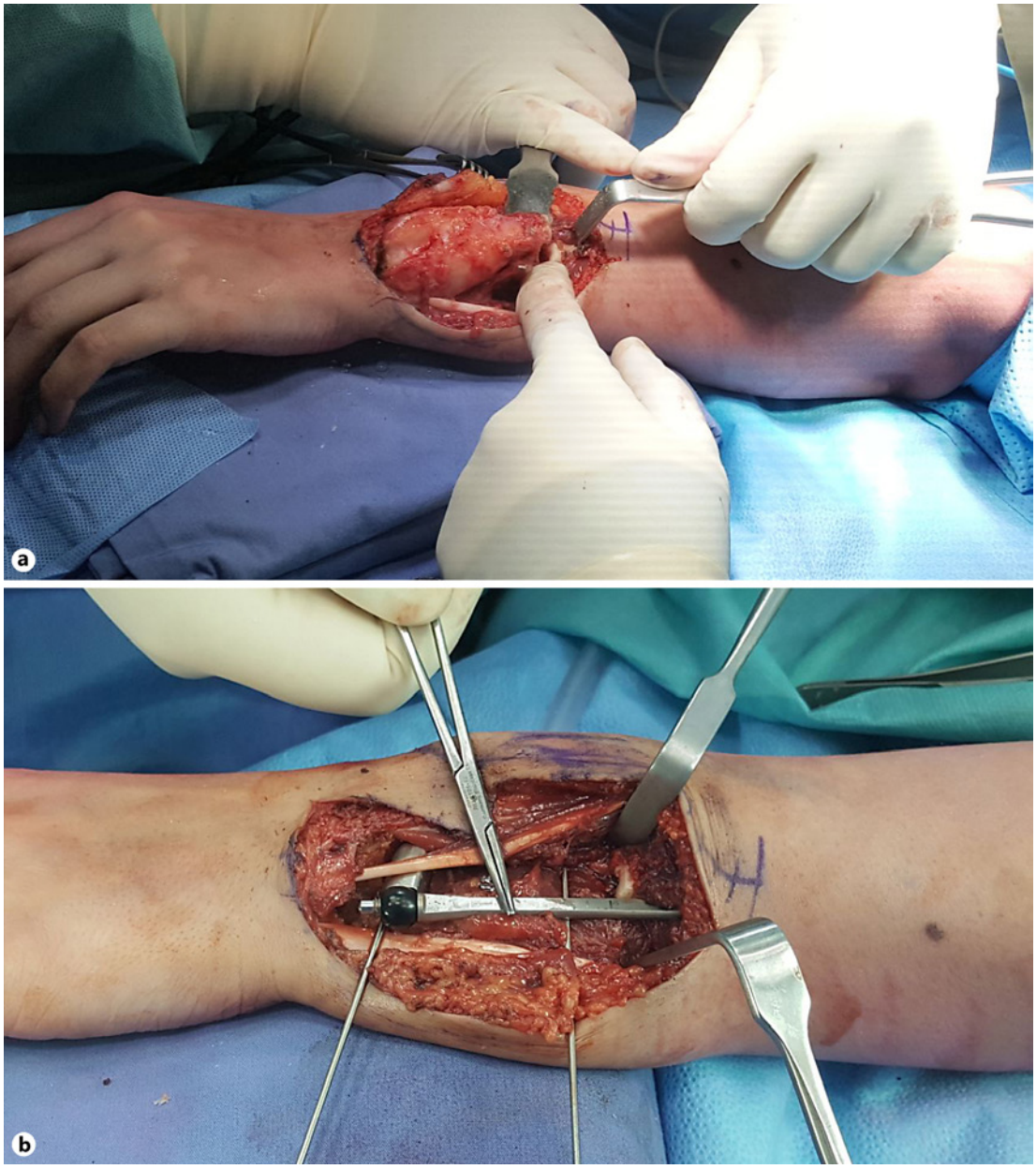

Fig. 3. Intra-operative images. **a** Radical excision of the distal ulna osteosarcoma with a safety margin. **b** Checking the ulnar stem trial was proximal enough to allow the trial ball to be fully seated within the socket of the radial plate before implanting.

of the drill bit were applied before we reached the required depth (11 cm), as we constantly made sure that our guidewire was kept in place and proximal enough before utilizing the cannulated drill bits to prevent them from passing through the side of the ulna. The ulnar stem was proximal enough to allow the trial ball to be fully seated within the socket of the radial plate.

A full trial of the ulnar stem along with the trial head was conducted and was in a good position (Fig. 3b), implantation of the cementless ulnar stem and application of radial head and final assembly was done by placing the ultra-high-molecular-weight polyethylene (UHMWP) ball over the distal peg of the ulnar stem and applying both the cover and pin. The

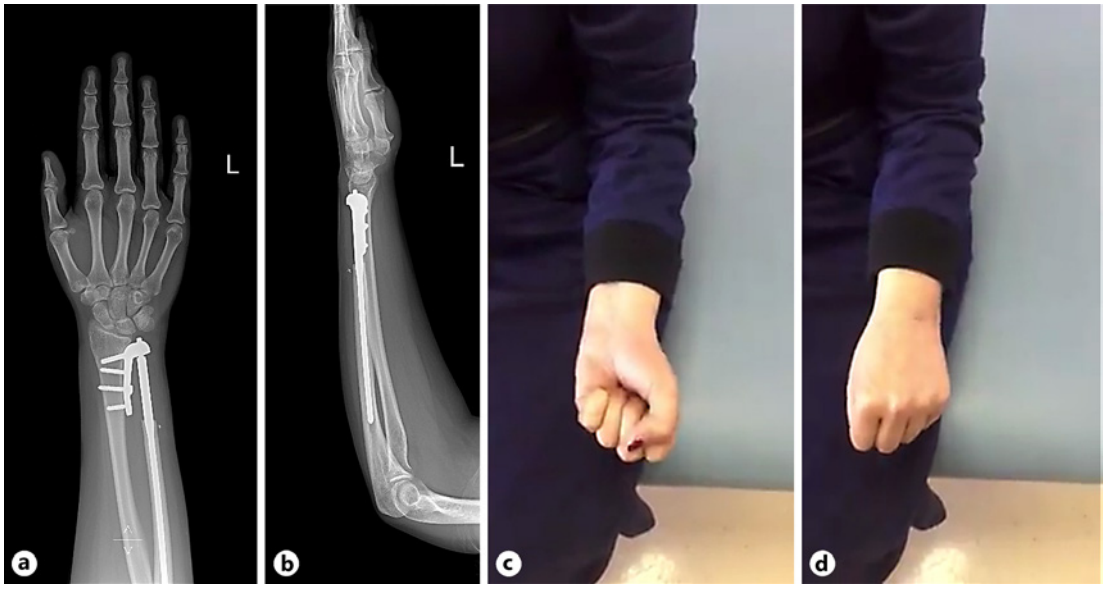

Fig. 4. Follow-up images showing the radiological position of the prosthesis and full range of mobility.

cover fully slid into place before inserting and advancing the single locking cross pin. Also, we ensured the pin was threading properly and the screwdriver in alignment with the pin while it was advanced.

Checking the final seating, position, and range of motion, all were satisfactory. Copious irrigation was done and then we turned our attention to soft tissue reconstruction which was done using allo-fascia lata graft to prevent friction between the prosthesis and the tendons. The closure was performed in layers. We utilized skin stapler and above elbow backslap as well as a sugar-tongue splint for 3 weeks, which was removed thereafter with the initiation of occupational therapy.

The status of all resection margins came back negative for tumor cells, and the patient was followed up on a biweekly basis. Then she was started on adjuvant chemotherapy 23 days after the procedure, at that time her incision was completely healed, during which she started occupational therapy protocol and achieved a very good range of motion 8 weeks after surgery. She continued to be followed up for 2 years postoperatively and she is still in remission (Fig. 4).

Case No. 2

A 12-year-old female patient presented with a 2-month history of progressive left wrist pain and swelling following a minor trauma. On physical examination, localized tenderness and swelling were noted on the dorsal aspect of the distal ulna. As for the range of motion, a 10-degree loss of ulnar deviation and a 20-degree loss of supination/protonation were noted in comparison to the unaffected side. Physical examination of the respiratory system was insignificant. Imaging studies including X-ray and MRI (Fig. 5) showed an aggressive lesion originating in the distal ulnar metaphysis-diaphysis that extends into the epiphysis and the articular surface, with periosteal reaction, cortical destruction, and a large soft tissue component. A three-phase bone scan showed early and delayed abnormally increased radiotracer uptake in the distal third of the left ulna and around the wrist joint. Metastatic workup

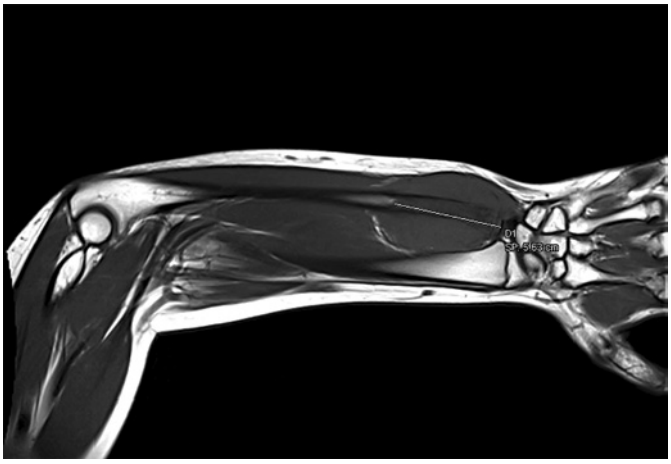

Fig. 5. MRI, T1-weighted image of the left forearm. Showing a lesion involving the distal diaphysis, metaphysis, and also the epiphysis with extension into the articular surface with the intramedullary component of the lesion measures 5.6 cm in craniocaudal dimension and evidence of surrounding mild oedematous changes.

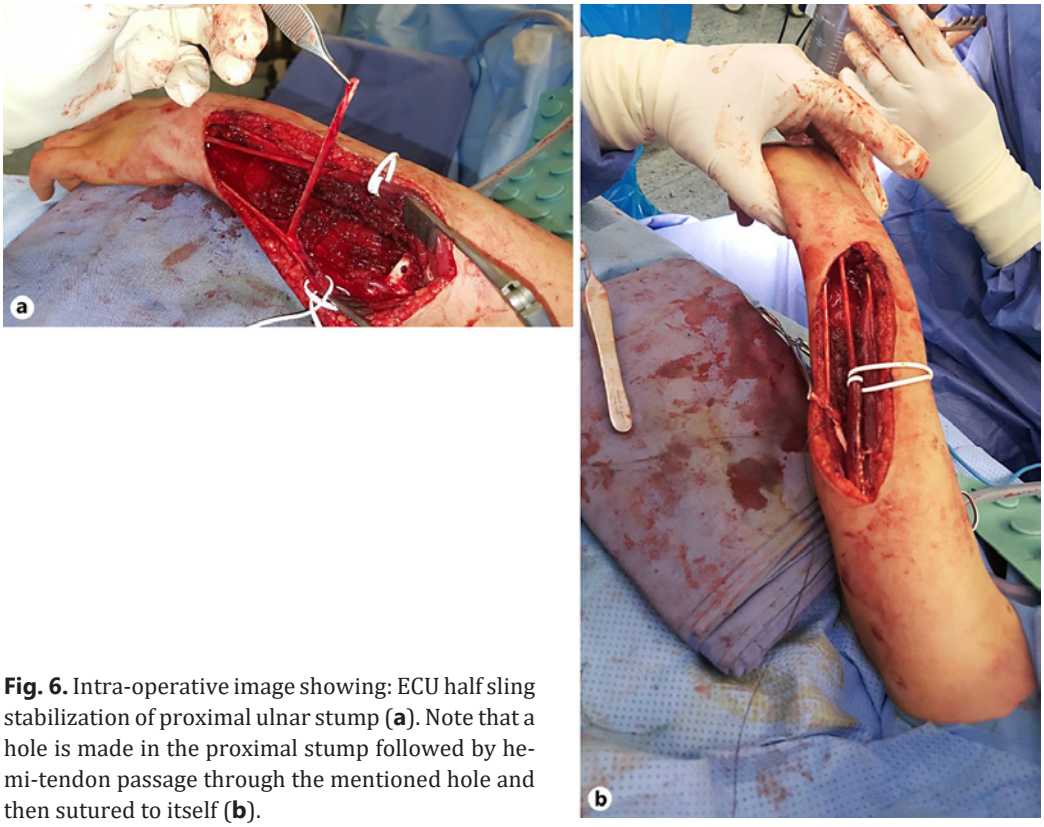

Fig. 6. Intra-operative image showing: ECU half sling stabilization of proximal ulnar stump (**a**). Note that a hole is made in the proximal stump followed by hemi-tendon passage through the mentioned hole and then sutured to itself (**b**).

with a chest CT scan showed no evidence of metastasis. Ultrasound-guided biopsy of the mass and histopathological examination revealed high-grade chondroblastic osteosarcoma.

After discussing the case at the sarcoma multidisciplinary clinic with the patient and her parents and explaining all the treatment options available, the first dose of methotrexate was given but resulted in acute kidney injury and high levels of methotrexate in the patient's blood, which required the administration of carboxypeptidase. Neoadjuvant chemotherapy was switched to only doxorubicin and cisplatin. MRI was done following 4 cycles of chemotherapy and showed a minimal regression (5–10%) of the soft tissue component associated with the bone lesion followed by a fifth cycle that was given before local control was carried out.

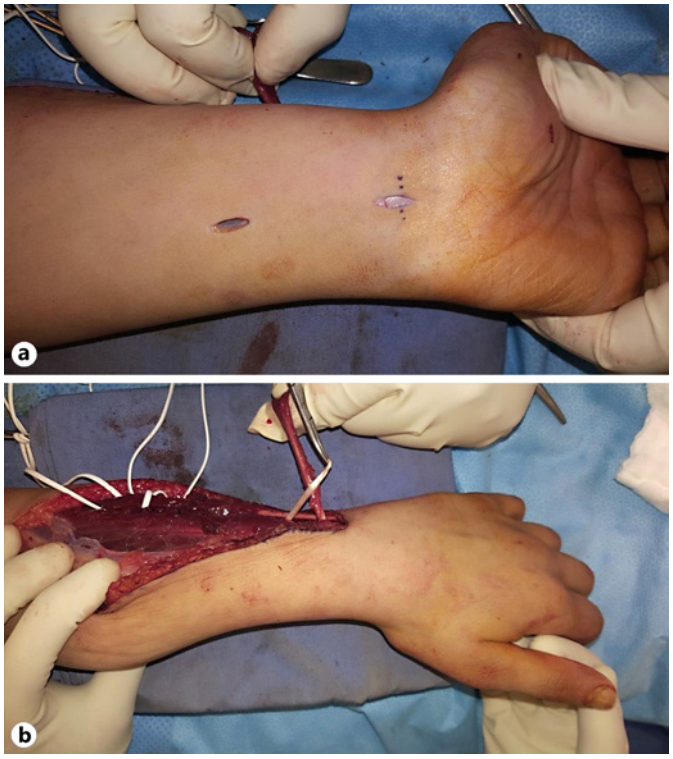

Fig. 7. Intra-operative images showing: **a** Harvest of the palmaris longus tendon using 2 small incisions. **b** Crossing of the palmaris longus and the extensor pollicis longus tendons for a pulvertaft weave.

Under a bloodless field using tourniquet without exsanguination, L-shaped incision was utilized on the dorsal/ulnar aspect of the forearm incorporating the previous core needle biopsy tract, and dissection was carried out around the distal ulna with care not to violate the tumor.

Proximal osteotomy of the affected ulna was done first, which was roughly about 10 cm in length measured from the most distal aspect of the ulna. A frozen section of the canal was sent from the ulnar stump and it came back negative for tumor cells. We continued dissection from proximal to distal manner. The sheath of the ECU was easily freed from the mass by blunt dissection. The sheath was opened longitudinally and then resected with the rest of the specimen, salvaging the ECU tendon. The radically excised specimen was tagged for orientation and sent for a permanent pathology. Tourniquet was deflated after tumor resection and we ensured adequate hemostasis.

After tumor resection, the radial half of the ECU tendon was longitudinally split into a distally based slip. A hole was drilled in a dorsal-volar direction close to the edge of the ulnar stump (Fig. 6a). The hemi-tendon was passed through the hole and then sutured to itself with a modified Golder and Hayes technique [1] (Fig. 6b). A significant portion of extensor pollicis longus muscle origin was removed with the tumor resection. We decided to do palmaris longus tendon harvest through two small incisions and transferred it to the extensor pollicis longus in the same session using a pulvertaft weave (Fig. 7).

Closure in layers over a drain that was brought out 1 cm proximal to the line of our incision.

All surgical margins were free of tumor cells and one cycle of adjuvant chemotherapy was given a month after the surgery. Sugar-tongue splint including a thumb extension splint was applied and it was removed 2 months after the surgery and an occupational therapy protocol was then started with a good range of motion achieved 4 months after surgery. A full range of motion was regained at 7 months and the patient resumed her daily life and is still in remission 2 years after the operation (Fig. 8).

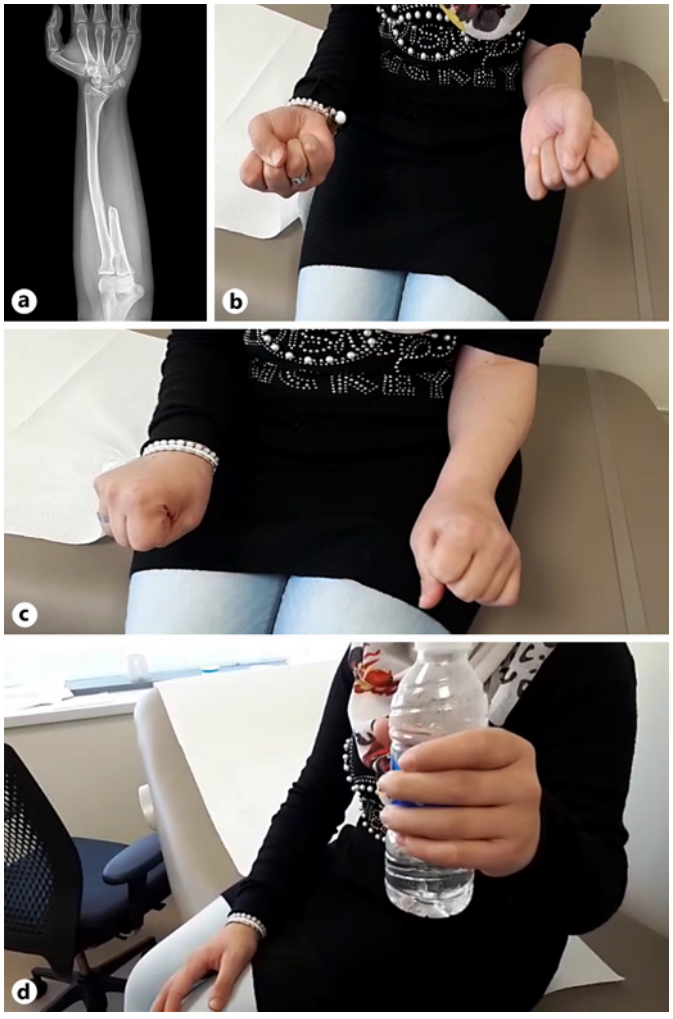

Fig. 8. Post-operative images showing: **a** X-ray of the left forearm showing post-en-block excision and proximal ulnar stump. **b–d** The patient is able to perform full supination, pronation, and holding a bottle respectively.

Discussion

Osteosarcoma, characterized histologically by osteoid-producing malignant cells, is the most common primary malignant bone tumor of childhood, and the second most common in adulthood following plasma cell myeloma [2, 3]. It is classified based on its location within the bone, composition, grade, and the condition of the underlying bone [3]. A few-month history of a painful mass, which is often attributed to trauma or strenuous exercise, is the usual presentation [4].

The presence of a lytic, sclerotic, or a mixed lesion on simple radiography raises suspicion of osteosarcoma. To confirm the diagnosis, a biopsy is required. Several other investigations are essential to evaluate the magnitude of the disease. First, to determine the local extent of the disease, an advanced imaging technique is utilized. Then, a radionuclide bone scan is carried out to assess the primary tumor and search for multiple lesions. A CT scan of the lung is needed to look for distant metastases, as it is the most common site of metastatic disease [4].

Surgery is the sole means of tumor local control, as osteosarcoma remains immune against radiation therapy. Systemic chemotherapy is necessary as microscopic metastases are present in almost all patients [5].

The metaphysis of long bones is the most common site of osteosarcoma occurrence in the young. Of these, the distal femur, proximal tibia, and proximal humerus are the most frequent sites, respectively [4]. While it is possible for osteosarcoma to occur in any bone, the ulna holds the lowest rate of involvement, accounting for only 2% of all long bone tumors [6]. In a review of 1,649 patients with osteosarcoma, 4 cases of ulnar involvement were reported, with only 2 being in the distal ulna [7].

The distal radioulnar joint (DRUJ) is of paramount importance in forearm supination and protonation, as well as grip and lifting power. The hand, wrist, and even elbow can be affected by DRUJ pathologies [8, 9].

The resection of the distal ulna as a treatment for several conditions have been recommended, as it has been consistently thought to be dispensable. Pain and limitation of the range of motion are the complications of such a procedure [10].

Although several prostheses have been created to replace the DRUJ mechanical function, most of them are made to be used in patients with intact ligaments and soft tissue surrounding the joint. Specifically invented for the alternate patient population, as it possesses intrinsic stability, is the Scheker prosthesis [9].

The prosthesis is made of cobalt-chromium, titanium, and UHMWP, which grants the constructed DRUJ normal grip strength and lifting abilities. The design is made up of a radial component and an ulnar one. The radial component is a plate which has a distal socket. The other component is a fluted stem that inserts into the ulna and has a polished peg distally onto which a UHMWP ball slides. The ball, which is seated in the radial socket, is covered by a metal cap that secures its housing. The ball moves freely within the socket, and the peg moves freely within the ball, permitting complete protonation and supination range of motion [9, 11].

Twenty-three total DRUJ reconstructions using the prosthesis were performed by Dr. Scheker on patients who had had at least one salvage procedure involving partial or total ulnar head excision. Before the insertion of the prosthesis, all 23 patients reported severe pain, disability, and ulnar instability. Following the insertion, not only did all patients report total pain relief, normal protonation and supination ranges of motion, and ipsilateral hand ability to lift weights, with an average of 14 lb, but they also reported no complications [11]. The Scheker/APTIS® DRUJ prosthesis gained the US Food and Drug Administration approval for marketing in January 2005 [9].

While the prosthesis is intended for patients who underwent ulnar head resection following arthritis, trauma, or previous surgery [9, 11], we expanded its usage to include reconstruction of DRUJ following radical resection of distal ulnar osteosarcoma using DRUJ prosthesis.

As for the skeletally immature population, tumors invading the epiphysis of the distal ulna in children can cause a serious ulnar deviation deformity of the wrist and affect forearm rotation function, the traditional methods for the treatment of these diseases include distal ulnar resection and/or ulnar lengthening with correction of the radius. The main aim whenever possible is to improve the appearance of the forearm, reduce the distal radius ulnar deviation, transmit the stress more evenly between the radius and ulna, and guide the ulna in longitudinal growth. However, these traditional operations will not prevent growing children without a distal ulnar epiphysis from developing gradual wrist deformities years later. Lacking the growth plate in the distal ulnar epiphysis, the secondary wrist ulnar deviation deformity caused by gradual ulnar shortening will eventually be unavoidable. Moreover, bone-lengthening procedures are associated with many complications, such as infection, vascular and nerve injuries, joint dysfunction, and non-union [12].

Though in our skeletally mature patient, the reconstructed DRUJ prosthesis showed good functional wrist outcome, still challenges might be encountered in skeletally immature patients where applying those DRUJ prosthesis might interrupt the growing wrist and due to

that other alternatives need to be considered. In our skeletally immature case, we decided not to reconstruct the DRUJ and instead, we stabilized the ulnar stump using the ECU slip, as it retains the ulna in its original direction and prevents unwanted mobility of the free edge of the ulna during supination, protonation, and ulnar or radial deviation. After 2 years of following the patient, the functional outcome is good without deformities.

The other way of reconstructing the DRUJ in the growing child was through the biological reconstruction; the feasibility of distal ulna reconstruction with the proximal fibula in children was mentioned. Transfer of the vascularized proximal fibula including the growth plate can provide sufficient bone for the diaphyseal reconstruction of the articular surface and can potentially lead to axial growth. Furthermore, the harvest of the proximal fibular graft is noninvasive for the knee. Also, there are sufficient tendons, ligaments, and capsules attached around the fibular head that can be used to reconstruct the distal radioulnar joint and stabilize it [12].

Osteosarcoma of the distal ulna is an extremely rare entity; therefore there are no clear-cut guidelines about the preferred reconstructive modality after en bloc excision. One of the limitations of this study is the number of patients where this might invite further patient recruitment for future research to conclude comparative studies on long-term functional outcomes as well as the kinematic point of view among these different reconstructive methods in both skeletally mature and immature patients.

Acknowledgment

Not applicable.

Statement of Ethics

This is a case report. We have obtained written informed consent from the patients to publish this report.

Disclosure Statement

The authors have nothing to disclose.

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Author Contributions

M.K.A.C. and N.A. performed the literature search and wrote the manuscript guided by S.A.A. H.H. obtained and reviewed the pathology samples and provided the images. H.A. obtained consent from both patients. W.A. reviewed the case and provided radiological images. S.A.A. and A.M.S. oversaw the manuscript's inception, guided the literature search, counseled the patient, performed the procedures, wrote portions of the manuscript, and provided major revisions. All authors read and approved the final manuscript.

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