Embryonal Rhabdomyosarcoma of The Forearm: A Diagnostic Dilemma and Surgical Management

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Abstract

Establishing the diagnosis of primary soft-tissue sarcoma is a difficult task even for an experienced clinician. There is a plethora of clinical manifestation yet its radiological appearance can be very deceiving in every way. We report a case of embryonal rhabdomyosarcoma in an adult female with lump in her left distal forearm, with clinical manifestation and radiological appearance that immitate bone sarcoma. The diagnosis was not established until the biopsy result came up which also confirmed by immunohistochemistry stain. The treatment is a novelty limb salvage surgery with modified forearm segmental amputation and shortening procedure. Six months after the surgery the patient had good functional outcome without local recurrence and distant metastasis.
Keywords: *embryonal rhabdomyosarcoma, limb salvage surgery, forearm segmental amputation*

Rabdomiosarkoma Embrional Antebrakhii: suatu Masalah Diagnosis dan Penatalaksanaan Bedah

Abstrak

Menegakkan diagnosis sarkoma jaringan lunak primer merupakan pekerjaan sulit, bahkan di tangan klinisi yang berpengalaman sekalipun. Sejumlah kasus sarkoma jaringan lunak menunjukkan manifestasi klinis, tetapi gambaran radiologisnya tidak sesuai. Pada makalah ini dilaporkan satu kasus rabdomiosarkoma embrional pada perempuan dewasa dengan benjolan di antebrachii distal kiri dengan manifestasi klinis dan gambaran radiologi menyerupai sarkoma tulang. Diagnosis pasti ditegakkan setelah pemeriksaan pulasan imunohistokimia. Penatalaksanaan bedah yang dilakukan merupakan inovasi baru dengan cara limb salvage surgery amputasi segmental antebrachii yang dimodifikasi dan shorthening procedure. Pada pengamatan 6 bulan pascabedah, pasien memiliki luaran fungsional yang baik tanpa disertai rekurensi lokal dan metastasis jauh.
Kata kunci: *rabdomiosarkoma embrional, limb salvage surgery, amputasi antebrakhii segmental*

Introduction

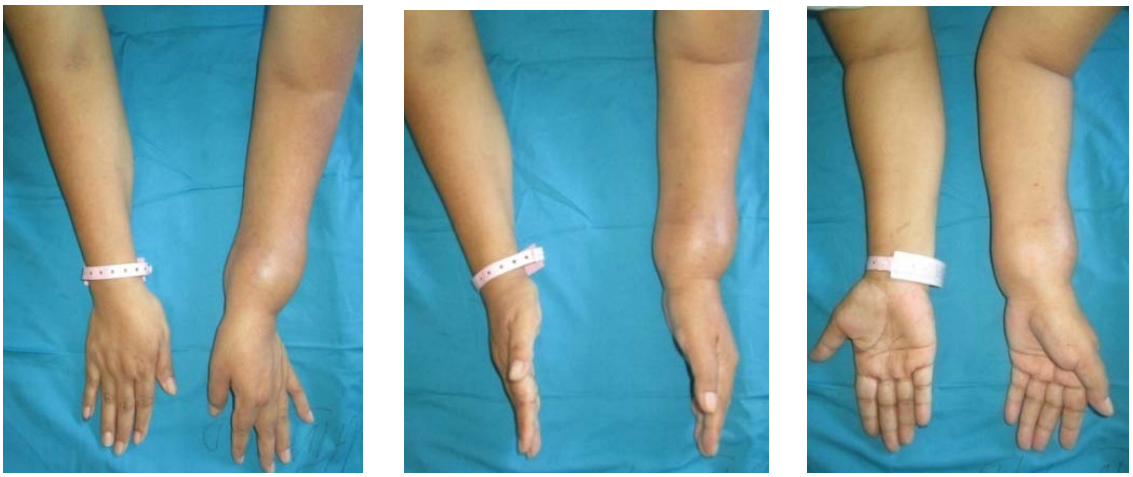
Embryonal rhabdomyosarcoma is a primitive, malignant soft-tissue sarcoma that recapitulates the phenotypic and biological features of embryonic skeletal muscle.[1] It is the most common pathologic subtype of rhabdomyosarcoma in children but recently have been found also quite common in adults.[2] Only 1% of adult malignancies are soft tissue sarcomas and rhabdo-myosarcoma constitutes approximately 3% of all soft-tissue sarcomas.[3] In children, these tumors can arise almost everywhere and most often found in the head and neck region followed by the genitourinary tract. It is not the same in adults where extremity is the most common site involved.[4] With the vast array of clinical findings, it is very difficult to diagnose rhabdo-myosarcoma especially if the presentation is atypical.

The recommended treatment for rhabdomyosarcomas (soft-tissue sarcomas) are incorporate surgery, radiation therapy and chemotherapy.[5] Here, we present a case slow-enlarging mass with radiological appearance resembles bone sarcoma. The treatment is a novelty with modified forearm segmental amputation and subsequent shortening procedure.

Case Report

A forty-seven-year-old female presented to our hospital with enlarging mass on her left wrist since 6 years prior to admission. At first, she only felt pulsating pain on her wrist without prior history of trauma. Six monthslater,she noticed a 2 cm mass on her wrist. The mass became bigger into 5 cm in five year period. She went to bonesetter several times but therewas no improvement. Subsequently, she went to an orthopaedic surgeon in general hospital for further evaluation, diagnosed as bone tumour and referred to our hospital.

On physical examination, there was a hard, well-defined circumferential mass with approximately 8 cm in diameter (Figure1). The mass was tender with visual analogue scale (VAS) 3-4. The wrist joint movement was limited due to pain and mass.



A. **B.** **C.**

Figure 1. Clinical presentation of the circumferential mass on the left wrist of a forty-seven years old female patient: (A) from dorsal site, (B) from radial site, and (C) from volar site.

Radiograph of the wrist joint showed permeative pattern of destruction on the radial distal epimetaphysis. Wide transitional zone, prominent softtissue involvement and aggresive periosteal reaction suggest this lesion as a bone sarcoma (Figure 2).

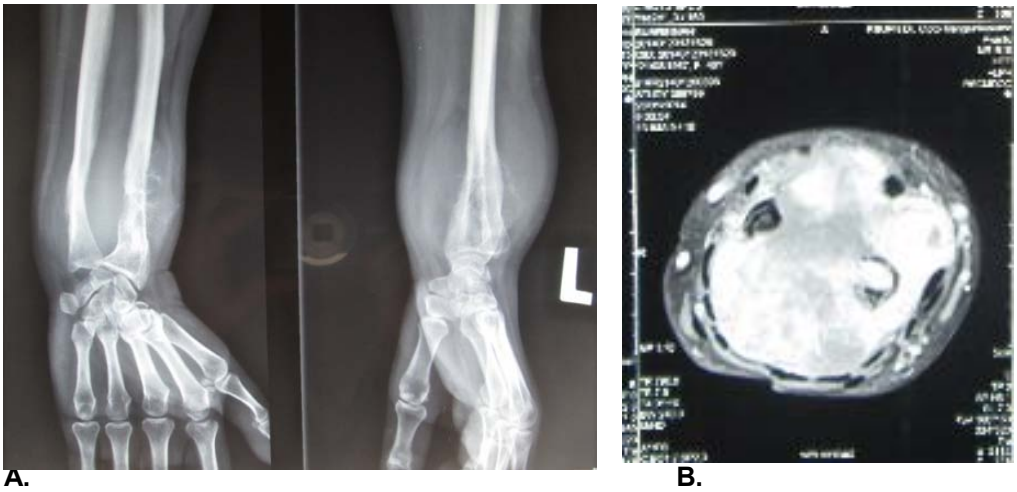


Figure 2. (A). Initial antero-posterior (AP) and lateral view radiographs showed permeative pattern of destruction on the radial distal epimetaphysis. Wide transitional zone, prominent softtissue involvement, and aggresive periosteal reaction suggest this lesion as a bone sarcoma. (B) MR axial T2-weighted image showed hyperintense signal intensity consistent with a solid soft tissue mass with necrosis.

Further evaluation with chest radiograph and bone scintigraphy showed no distant metastases. Magnetic resonance imaging (MRI)showed a solid mass in the region of distal radius with involvement of the radioulnar joint and its surrounding structures, including median nerve. Radial artery, vein and nerve were displaced peripherally along with ulnar artery, vein and nerve (Figure 3).

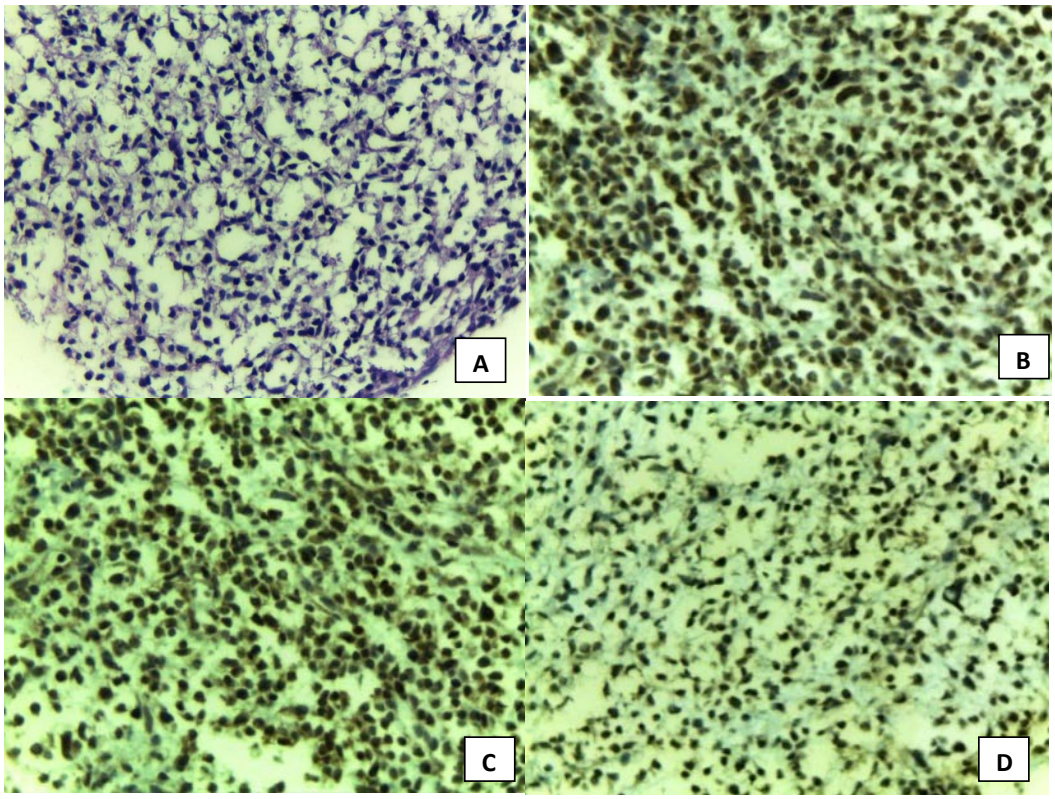


Figure 3. A. The biopsi specimen showed round cell tumor with elongated and eosinophilic cytoplasm that most appropriate for embryonal rhabdomyosarcoma (H&E 100x). Immunohistochemistry staining showed(B) Vimentin positive (100x), (C) Desmin positive (100x) and (D) MyoD1 positive (100x).

Histopathologic result from core biopsy was to our surpise, embryonal rhabdomyosarcoma (Figure 4). Further confirmation with immunohistochemistry revealed positive for vimentin, desmin, and myoD1 (Figure 4). We decided to do limb salvage surgery with modified forearm segmental amputation and shortening procedure of the left distal forearm followed by adjuvant chemotherapy.

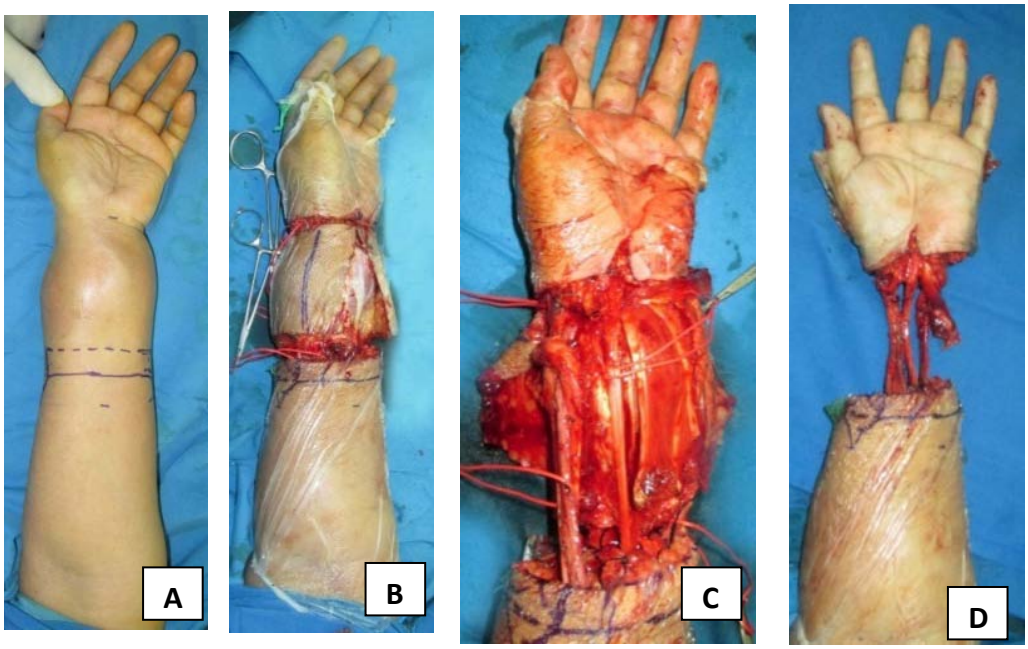
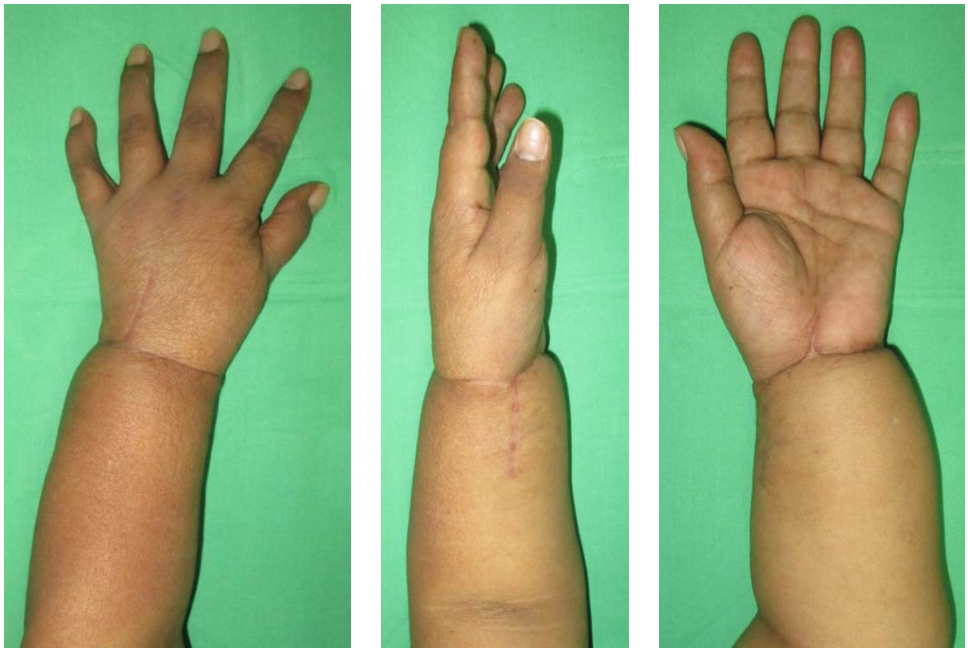


Figure 4. Intraoperative pictures. (A) The circumferencial design; (B)The skin incision that made accordingly; Identification of the structures of the foream. (C) Preservation of the neurovascular bundle of the distal forearm. (asterisk; superficial radial nerve and radial artery, plus sign; median nerve, pound sign; ulnar nerve and ulnar artery); (D). After tumoural resection (osteotomizedthe radius and ulna), cut the tendons and performed disarticulation of the left wrist joint.

A circumferential incision design was performed well above the margin of the tumour.Another similar incision was made at the level of the wrist joint. Those incisionswere connectedwith a longitudinal third incision whichwas perpendicular from the first two incisions. Neurovascular structureswere identifiedand preserved. The muscles at the level of first incision (mid-forearm) and the tendons at the level of the wrist joint were also identified and marked with sutures and loops. The tumour mass wasresected by osteotomizedthe radius and ulna), cut the tendons and performed disarticulation of the left wrist joint. (Figure 5). Subsequently, arthrodesis were performed of the wrist joint using reconstruction plate. All of the marked tendons were sutured back using the pulvertaft technique.



A B C

Figure 5. Clinical presentation of six months after surgery: (A) from dorsal site, (B) from radial site, and (C) from volar site.

Post-operatively, the patient underwent several sessions of chemotherapy and rehabilitation. Six months follow up after the surgery, she still had little difficulty in using her hand to grab object, but felt dramatic improvement in pain level. The outcome of the surgery was measured using Michigan Hand Outcomes Questionnaire and the score improved from 24 to 35.

Discussion

Rhabdomyosarcoma is a very rare neoplasm in adult population. Due to its rarity, its clinical characteristic is poorly understood and large multi-institutional clinical trials have not been performed.[2] Establishing the diagnosis of rhabdomyosarcoma in adults are as difficult as treating it. The clinical presentation is highly variable in patients with bone or soft-tissue sarcoma. A general consideration regarding diagnosis is that bone sarcomas are painful whereas soft-tissue sarcoma are not but there are exceptions to this.[6] In unfavorable sites such as the distal forearm, even soft-tissue sarcoma can produce pain mimicking bone sarcoma.[6]This patient came to us with painful mass which presented similar with bone tumour.Embryonal rhabdomyosarcoma is the most common histological findings in adults and extremity as well as genitourinary tract shares the same anatomic sites of origin.[2,3]

Anatomically, the distal radius is a common skeletal site for primary bone tumour and is the third most common site for giant cell tumour of the bone.[7] In this case, not only it has ill-defined permeative pattern with wide transitional zone, prominent soft-tissue involvement, calcification, but also interupted periosteal reaction that resembles bone sarcoma. The MRI findings further add up to the diagnosis with a highly suspicion of primary intramedullary bone tumour that extends into the soft-tissue. A surprising result of histopathology as a gold standard then followed by immunohistochemistry stain confirmed that our suspicion was wrong the whole time. The lesion was soft-tissue in origin.

There are only few case report that presents rhabdomyosarcoma of the forearm.[8-10] The distal forearm is a location not previously reported in current literature, therefore no standard guidelines for management available.[10] Limb salvage surgery is still the treatment of choice in treating sarcomas of the forearm.[11,12] Here, we decided to do limb salvage surgery with modified forearm segmental amputation and shortening procedure of the left distal forearm with preservation of the neurovascular bundle. This method may have a role in selected cases for partial limb salvage surgery regarding local control, long-term survival and useful limb function during long-term follow-up.[12] In cases which wide surgical margin can be achieved around the tumours, segmental resection of the tumour-bearing region in a cylindrical fashion and reattaching the distal part with or without

microsurgical technique can be an alternative procedure to maximize function rather than amputation.

The most important prognostic factor was in localized disease, 5-years survival rate were 82% for children and 47% for adults.[2]Close monitoring of the patient after surgery and chemotherapy is needed to maintain disease-free state. Furthermore, rigorous rehabilitation will be needed to achieve favorable remaining hand function. Decrease in muscle strength will be expected post operatively due to loss of tension during operation. This can be encountered also with decent rehabilitation.

In summary, although very rare, adults maybe affected with rhabdomyosarcoma and its most common site of involvement in adults is the extremity. Diagnosing rhabdomyosarcoma in this sitemay be difficult due to a variety of clinical symptoms and radiological findings mimicking bone sarcomas.Through this case, segmental forearm amputation and subsequent shortening can be considered as a decent option in selective case due to its preservation of remaining hand function.

Conflict of Interests

The authors declare that there is no conflict of interests regarding the publication of this paper.

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