

Primary hand rhabdomyosarcoma: A rare case report

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Abstract

Rhabdomyosarcomas are malignant, soft tissue tumors with striated muscle origin. Malignant round cell tumors are the common malignancy of childhood. The primary localizations are head and neck and genitourinary system. Only 7 % of lesion is seen in the upper extremity. Primer hand involvement is rare. In this article, we presented a 16-year-old girl who admitted to our clinic with a complaint of a painful mass that grew rapidly in the left hand thenar area. Imaging findings revealed a heterogeneous solid mass lesion between the 1st and 2nd fingers, which showed dense contrast enhancement, approximately 6x5x7 cm in size, leading to destruction in the bony structures. A histopathological examination of the mass indicated alveolar rhabdomyosarcoma. We decided to present this rare entity with the radiological imaging findings.

Keywords: Rhabdomyosarcoma; Hand; Ultrasonography; Magnetic Resonance Imaging.

INTRODUCTION

Rhabdomyosarcoma can occur in any part of the body. The primary localizations are head and neck and genitourinary system. Only 7% of lesion is seen in the upper extremity. Primer hand involvement is rare (1). Ultrasonography (US), multislice computed tomography (MSCT), and magnetic resonance imaging (MRI) are used in the radiological diagnosis of soft tissue masses. In this article; in a 16-year-old female patient, with alveolar rhabdomyosarcoma in the left hand was presented in the presence of US and MRI findings.

CASE REPORT

A 16-years-old female was admitted to our clinic with complaint of a rapidly growing painful mass in the left hand thenar area. Patient did not reveal any significant features about in their resume and history. Routine biochemistry and tumor markers were normal. On left hand physical examination, hard, fixed, solid mass with pain was detected. Examined by US; a heterogeneous solid mass lesion was observed with a lobulated contour between the 1st and 2nd finger, approximately 6x5x7 cm in size, with no test planes with metacarpal bone structures. Doppler US examination showed that lesion has intense vascular flow (Figure 1).

In contrast enhanced MRI examination revealed a solid mass lesion with a lobular contour was observed

between the 1st and 2nd fingers of the left hand, leading to destruction in the metacarpal bone structures. The identified mass was heterogeneous in the T1 weighted sequence, and showed dense contrast enhancement after I.V. contrast agent (Figure 2a-c). A histopathological examination of the mass was reported as an alveolar rhabdomyosarcoma (Figure 3). The patient who had no metastasis in the body was included in the chemotherapy and radiotherapy program. No recurrence or metastasis was observed in the follow-ups during the 1-year period.

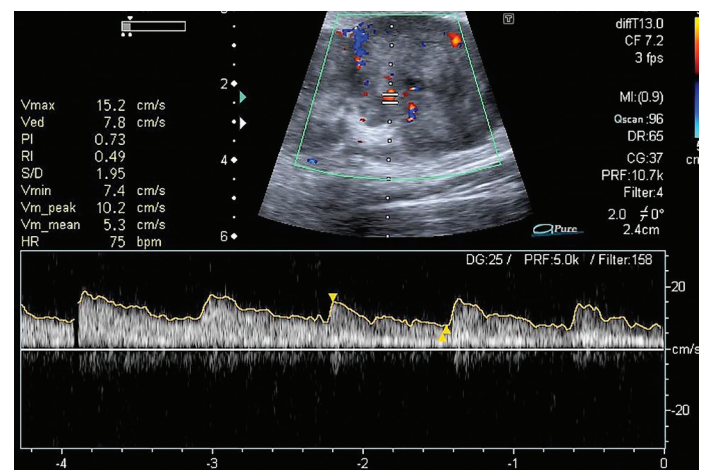


Figure 1. In the hand US; a solid mass lesion is observed with a heterogeneous echo, lobulated contour, between the 1st and 2nd finger. Doppler US examination showed that lesion has intense vascular flow

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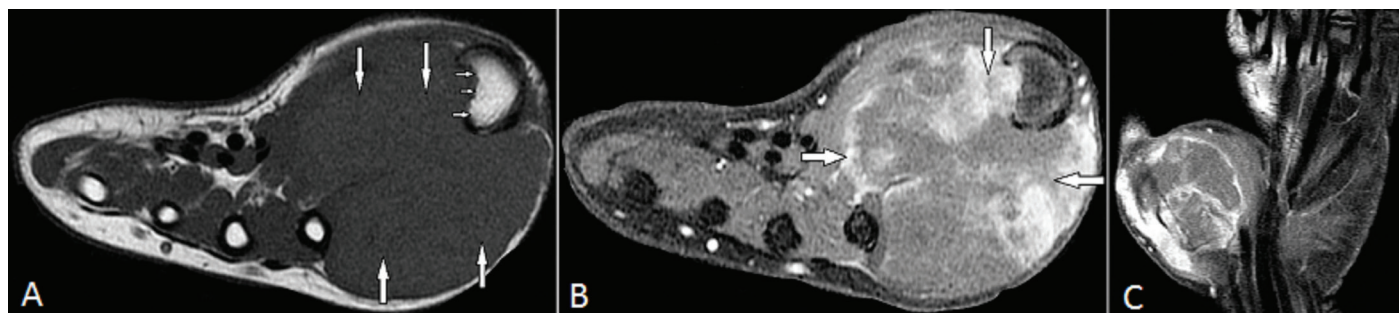


Figure 2. In the T1 weighted axial MRI; a solid hypointense mass lesion was observed (arrows) leading to destruction in the bone structures (small arrows) (a). Contrast enhanced axial (b) and coronal (c) hand MRI showed dense contrast enhancement (arrows)

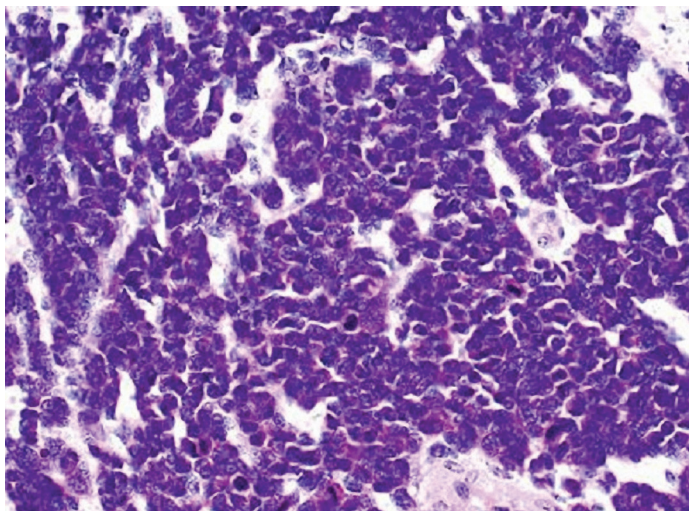


Figure 3. In histopathological examination; the solid variants are composed of sheets of darkly staining small round cell (arrows) (HEEx200)

DISCUSSIONS

Rhabdomyosarcomas are malignant neoplasms with striated muscle differentiation. It most commonly made metastases to the lung and especially to the regional lymph node. In histopathologically; rhabdomyosarcoma can be classified into spindle cell, embryonal, botryoid, alveolar and undifferentiated types. Each of subtypes is common in particular age groups. The alveolar subtypes are more common in the adolescent; by the way the embryonal types are common in children under 8 years (2). However, classification has a significant correlation with prognosis. According to the International Classification of Rhabdomyosarcoma (ICR), alveolar rhabdomyosarcomas, account for 32% of all rhabdomyosarcoma, display a more aggressive clinical course and have poor prognosis (3). These tumors typically appear in the deep soft tissue of the extremity and broaden normally muscle fiber.

Microscopically, the tumor appearance may be confused with other small round blue cell tumors including Ewing's sarcoma and lymphoma (2). Immunohistochemically stains, myogenins and desmin. Cytogenetic analysis can be performed to distinguish subtypes (4).

Radiologically, plain radiography and MSCT show local

bone invasion that seen in 25% of all cases. Bone metastasis is usually seen as lytic and less as a mixt type. US findings are non-specific. It is usually seen as a heterogeneous hypoechoic solid mass in US. MRI imaging features is also non-specific in characteristics. In T1-weighted images, lesions are isointense with muscle and heterogeneous hyper intense in T2-weighted images. Often necrotic foci can be observed. Intensive heterogeneous enhancements are seen after contrast media injection (5,6).

Oncologic and non-oncological reasons should be considered in the differential diagnosis of rhabdomyosarcoma. Non-oncological causes include trauma. There is usually a crash story. Hematoma is seen usually soft. In sarcomas, there are usually no trauma histories, hard mass, without color change on the skin and able to grow over time. It may be also confused with benign lesions such as lipoma, rhabdomyoma and neurofibroma. Rarely, it is confused with other benign lesions such as myositis ossificans, pyogenic myositis. Beside this non-Hodgkin lymphoma, neuroblastoma, Ewing sarcoma familial tumors, acute myeloid leukemia, Langerhans cell histiocytosis should be considered in the differential diagnosis of rhabdomyosarcoma (2,4,7). The gold standard diagnostic method in soft tissue mass is histopathological examination.

In the literature, few cases of upper extremity and primer hand involvement have been reported with childhood rhabdomyosarcoma. Milanovic et al; described an alveolar type rhabdomyosarcoma in a 2 years old child (8). Mahana et al; reported spindle-cell rhabdomyosarcoma of the thumb (9). More rarely, congenital rhabdomyosarcoma in the shoulder and adult hand rhabdomyosarcoma have been reported (1,10,11). Local treatment resulting from surgery and/or radiotherapy, survival increased from 20% to 70% with the addition of systemic chemotherapy. Surgery, chemotherapy and radiotherapy are performed considering the location of the tumor, the stage, the histopathologic subtype, and the age of the patient (12).

In conclusion; extremity rhabdomyosarcoma has poor prognosis. Alveolar rhabdomyosarcoma is the most common subtype is seen. Early metastasis and high recurrence rates are available. In addition to extensive surgical resection adjuvant chemotherapy and radiotherapy may be useful in treatment. In the radiological

diagnosis of sarcomas; US, Doppler US, MSCT, MRI, and conventional radiography may be helpful. MRI has been reported as a more sensitive modality in the diagnosis of soft tissue pathologies.

REFERENCES

1. Valone F, Liu J, Genrich G, Lattanza LL. Alveolar rhabdomyosarcoma causing acute compartment syndrome of the forearm: a case report and review of the literature. *J Hand Microsurg* 2014;6(2):92-5.
2. Weiss SW, Goldblum JR. Enzinger and Weiss's soft tissue tumors. Fifth edition, Chapter 21. Rhabdomyosarcoma, Mosby, 2008.
3. Parham DM, Barr FG. Alveolar rhabdomyosarcoma. World Health Organization. Lyon, 2002;150-2.
4. Agarwala S. Pediatric rhabdomyosarcoma and nonrhabdomyosarcoma soft tissue sarcoma. *J Indian Assoc Pediatr Surg* 2006;11(1):15-23.
5. Allen SD, Moskovic EC, Fisher C, Thomas JM. Adult rhabdomyosarcoma: cross-sectional imaging findings including histopathologic correlation. *AJR Am J Roentgenol* 2007;189(2):371-7.
6. Davies CE, Davies AM, Kindblom LG, James SL. Soft tissue tumors with muscle differentiation. *Semin Musculoskelet Radiol* 2010;14(2):245-56.
7. Chen E, Ricciotti R, Futran N, Oda D. Head and Neck Rhabdomyosarcoma: Clinical and Pathologic Characterization of Seven Cases. *Head Neck Pathol* 2016;28.
8. Milanovic R, Vlajcic Z, Zic R, Stanec S, Rudman F, Stanec Z. Alveolar rhabdomyosarcoma of the hand in a 2 year-old child. *J Hand Surg Eur Vol* 2007;32(1):109-10.
9. Mahana S, Tomar R, Agarwal A, Sharma D, Khurana N, Gupta R. Spindle-cell rhabdomyosarcoma of the thumb: Rare site, rare tumor in a child. *Diagn Cytopathol* 2016;44(12):1094-7.
10. Khaleghnejad-Tabari A, Mirshemirani A, Rouzrokh M, Nariman S, Hassas-Yeganeh S, Gharib A, et al. Congenital rhabdomyosarcoma of shoulder. *Iran J Cancer Prev* 2012;5(3):164-6.
11. Seyhan N, Keskin M, Tosun Z, Savaci N. Rhabdomyosarcoma in an adult hand. *J Plast Surg Hand Surg* 2013;47(5):422-4.
12. Gupta AA, Chi YY, Anderson JR, Lyden E, Weigel B, Arndt C, et al. Patterns of chemotherapy-induced toxicities and outcome in children and adolescents with metastatic rhabdomyosarcoma: A report from the Children's Oncology Group. *Pediatr Blood Cancer* 2017;23.