Radiology Case Reports 19 (2024) 3508–3511

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case Report

# Exceptional case of a hemangioma-like rabdomyosarcoma in the hand's palm ☆

*Hassnae Tkak, MD[a,*], Ayad Ghanam, PhD[a], Mohamed Belahcen, PhD[c], Imane Kamaoui, PhD[d], Amal Bennani, PhD[e], Nadir Myri, MD[e], Aziza Elouali, PhD[a], Maria Rkain, PhD[a], Abdeladim Babakhouya, PhD[a], Noufissa Benajiba, PhD[b]*

[a] *Department of Pediatrics, University Hospital Mohamed VI, Faculty of Medicine and Pharmacy, University Mohamed first Oujda, Oujda, Morocco*

[b] *Department of Pediatrics, Ibn Rochd Hospital University, Casablanca, Morocco*

[c] *Pediatrics Surgery Department, Mohammed VI University Hospital, Oujda Faculty of Medicine and Pharmacy, Oujda, Morocco*

[d] *Radiology Department, Mohammed VI University Hospital, Oujda Faculty of Medicine and Pharmacy, Oujda, Morocco*

[e] *Anatomopathology Department, Mohammed VI University Hospital, Oujda Faculty of Medicine and Pharmacy, Oujda, Morocco*

ARTICLE INFO

*Article history:*
Received 11 February 2024
Revised 2 May 2024
Accepted 6 May 2024

*Keywords:*
Rhabdomyosarcoma
Mass
Vascular
Hand
Child

ABSTRACT

Rhabdomyosarcoma is a malignant tumor in children that might mimic a benign tumor, such as infantile hemangioma, particularly when detected early. Although rhabdomyosarcoma rarely occurs in the hand, its prognosis is generally poor, and successful treatment relies on a complete and radical surgical excision. We present a case of rhabdomyosarcoma located in the palm of an infant's hand, initially presenting clinical and radiological features suggestive of a vascular tumor. The resection of this mass was radical, and histological analysis and immunohistochemistry returned in favor of embryonic rhabdomyosarcoma. In similar cases recorded in the literature, the diagnosis may be first mistaken for that of a hemangioma, then confirmed by histology. This underlines the importance of a systematic anatomopathological examination of all tissues removed surgically.



☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

* Corresponding author.
E-mail address: hassnae.tkak@gmail.com (H. Tkak).

https://doi.org/10.1016/j.radcr.2024.05.023

## Introduction

Rhabdomyosarcoma is a highly malignant soft tissue tumor that develops from striated muscle cells [1]. It is rare in the hand, and cases involving the extremities are associated with a poor prognosis [1–3]. When superficial, a histological study is required for confirmation since it may be mistaken for an abscess or vascular tumor. Our observation highlights a rare mode of revelation of rhabdomyosarcoma, with particularities in terms of age of occurrence and unusual location.

## Case report

We present the case of an 11-month-old male infant with no significant pathological history who presents with a swelling affecting the entire hypothenar region of the right hand, gradually increasing in volume, initially painless, then painful, evolving 4 months before admission in the context of apyrexia and preservation of the general state. On clinical examination, the mass extended over the hypothenar region of the palmar side of the hand, measuring about 40 × 30 mm. It was soft, non-pulsatile, and not transilluminating, with no neurological deficit or distal vascular abnormality (Fig. 1).

Radiologically, the initial ultrasound revealed a hypoechoic, Doppler hypervascularized mass located on the palmar surface, suggesting a vascular tumor evoking a hemangioma. MRI showed a tissue mass that was enhanced after contrast injection (Fig. 2).

The diagnosis of hemangioma has been established through a bundle of clinical and radiological arguments. Surgical exploration found a hypothenar mass extending into the carpal tunnel.

The resection of this mass was radical, with no artery branches or ulnar nerve preserved. Histological examination and immunohistochemistry revealed a striated muscle tumor compatible with embryonic rhabdomyosarcoma. This proliferation came into touch with the deep and lateral edges (Fig. 3).

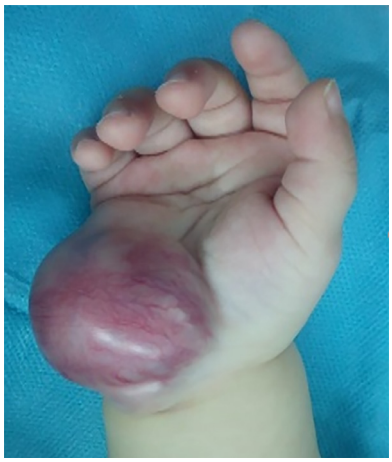

**Fig. 1 – The clinical aspect of rhabdomyosarcoma resembling a vascular mass.**

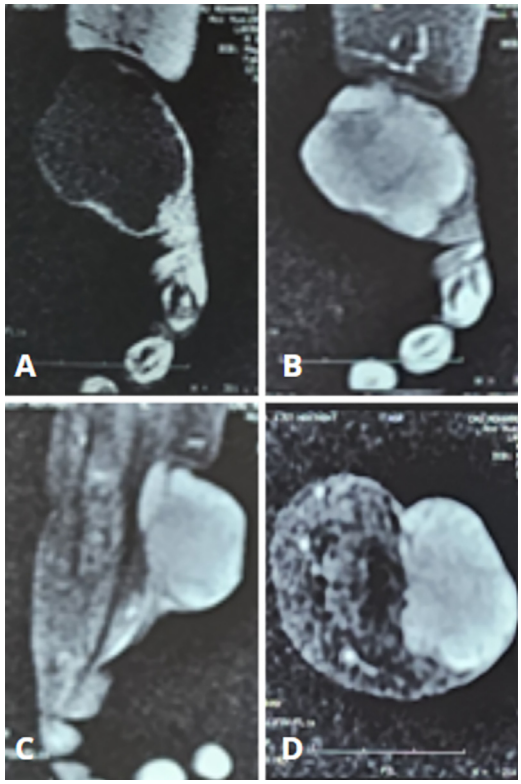


**Fig. 2 – Magnetic resonance imaging of the right hand in non-fat-saturated T1-weighted coronal section without contrast injection (A), fat-saturated T1-weighted sequence in coronal section with contrast injection (B), fat-saturated T2-weighted sequence in sagittal section (C), and fat-saturated T2-weighted sequence in axial section (D) showing a well-defined, lobulated soft tissue mass with regular contours, located in the subcutaneous tissues of the thenar and hypothenar compartments of the right hand measuring 42 x 35 mm. The mass demonstrates exophytic growth with isointense T1 signal, hyperintense T2 signal, and intense homogeneous enhancement after contrast injection.**

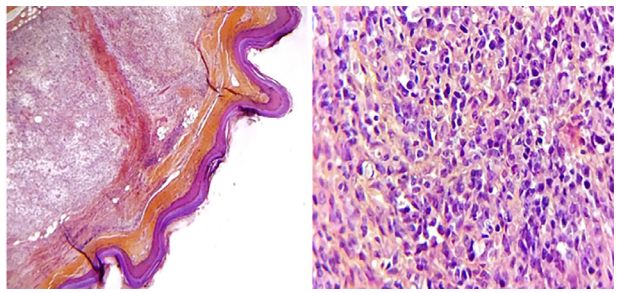

**Fig. 3 – Histological examination of the lesion reveals a dermal proliferation arranged mostly in sheets and large nodules (L: HES, x40). At higher magnification, tumor cells show a high nuclear to cytoplasmic ratio, with round and enlarged hyperchromatic nuclei, and scant cytoplasm (R: HES, x400).**

The extended radiological assessment revealed homolateral axillary lymphadenopathy, and the tumor was classified as stage II, group F, high risk.

The patient received chemotherapy following the 2005 MMT protocol. Given our patient's age of less than 3 years and perfect response to surgery and adjuvant chemotherapy, radiotherapy was not recommended. The clinical and radiological evolution was favorable, with no recurrence reported after 4 years.

## Discussion

Rhabdomyosarcoma is a highly malignant tumor with striated muscle differentiation. It is relatively common and accounts for about 5% of pediatric cancers [4]. In a retrospective study, rhabdomyosarcoma ranks third after leukemia and neuroblastoma in children [5]. Rhabdomyosarcoma is most commonly found in children under the age of 6. Indeed, it is rare in babies, but with a high risk of complications [6]. Rhabdomyosarcoma can occur anywhere in the body, and staging criteria depend on the tumor's size and location [7].

In most cases, it appears as benign tumors at the beginning of its evolution and goes misdiagnosed for months before quickly spreading to distant and locoregional metastases, usually in the lungs and bone [5].

It may show up as a well-defined nodule in soft tissues or as a benign-looking polypoid structure in an orifice of the body [5], as it can frequently be confused with primitive neuroectodermal tumors or Ewing sarcoma; furthermore, superficial tumors can simulate an abscess or a vascular tumor, such as hemangioma, with features such as redness, sensitivity, and increased local angiogenesis [8].

The disease mostly affects the head and neck, accounting for 27% of all cases. It can arise from the middle ear, sinus, palate, nasopharynx, orbit (10%), face, or neck. Other locations are rare and concern the urogenital area, which accounts for 21% of cases and might originate from the vagina, bladder, prostate, or anus; 20% of occurrences concern the extremities and trunk, with arms being the most commonly affected [1,3].

Primary hand malignancies are extremely rare and occur in less than 1% of cases in a large series reported in the literature (Cohen et al. 1987).

In a retrospective study of 1843 patients with non-metastatic rhabdomyosarcoma, Malempati et al. [9] found that 76 cases were under the age of 1 year, and 16% of them had extremity involvement. When rhabdomyosarcoma occurs in the extremities, its prognosis is poor due to the high risk of locoregional lymph node metastases [1–3. Generally, upper limb lesions have a better prognosis than lower limb lesions, and the most distal lesions have a better prognosis than the most proximal lesions [1,3,10]. Therefore, the survival of this tumor is most often related to its location [7].

The Indian literature reports a case of a 3-month-old baby who presented a mass extending over the dorsal and palmar sides of the hand since birth. The ultrasound revealed a massive echogenic mass on the palmar side of the left hand, at the subcutaneous and intramuscular level, encompassing the tendons, with some arteries and veins most likely suggesting a hemangioma. The child underwent a biopsy that turned out to be congenital rhabdomyosarcoma with metastases in the lymph nodes [8]. Cohen et al. also reported a similar case of an 18-month-old infant with Apert syndrome and congenital embryonic rhabdomyosarcoma. Since birth, he has had a 1 cm blue soft mass on the palm of his right hand, which was initially considered a hemangioma. At the age of 14 months, this mass gradually increased in size despite the administration of intra-lesional steroids. At the same time, another mass appeared on the forearm, and axillary lymphadenopathy developed. The biopsy confirmed the diagnosis of rhabdomyosarcoma.

All of the masses were substantially removed, and the child received chemotherapy and radiotherapy with favorable outcomes [2].

Rhabdomyosarcoma is classified into three histological types, each with its own set of characteristics: embryonic, alveolar, and pleomorphic rhabdomyosarcoma, while 15%-20% of cases do not fit into any of the categories, remaining unclassifiable tumors [1]. There is a clear association between the histological subtype and the anatomical site of the presentation. The embryonic type occurs frequently in genitourinary tumors (90% of cases) and in the head and neck; however, it is rarely described in the extremities. Alveolar and undifferentiated tumors are most commonly seen in the trunk and retroperitoneum [1]. There are no particular serum markers for the diagnosis of rhabdomyosarcoma. Indeed, his definite diagnosis is based on a biopsy of the mass, an anatomopathological examination, and immunohistochemical analysis.

The management of children with rhabdomyosarcoma is primarily based on the search for a cure without recurrence, which is why the initial surgery should only be considered if it is complete and radical. Limited excision is therefore not recommended for rhabdomyosarcomas, given the risk of recurrences [10]. Radical excision of the hand may require amputation, leading to potential loss of limb function or aesthetic problems, but it remains the treatment of choice [11].

In literature-reported studies by Linscheid et al. [10], Siegel et al. [12], and Xarchas et al. [1], a relatively high rate of recurrence was found in patients who had undergone limited excision of rhabdomyosarcomas in the extremities.

## Conclusion

Rhabdomyosarcomas are rare malignant tumors of the hand. They can take the form of misleading radioclinical masses, mimicking benign tumors such as hemangiomas. This observation illustrates the need for systematic histological examination of any pathological tissue collected during surgery.

## Patient consent

Informed consent has been obtained from the patient involved in the study, and appropriate ethical review board approvals have been obtained.

## REFERENCES


[1] K. Xarchas, N. Papavassiliou. Rhabdomyosarcoma of the hand two case reports and a review of the literature. J Hand Surg. 10.1016/s0266-7681(05)80194-9

[2] Cohen M, Ghosh L, Schafer ME. Congenital embryonal rhabdomyosarcoma of the hand and Apert's syndrome. J Hand Surg Am 1987;12:614–17. doi:10.1016/S0363-5023(87)80220-4.

[3] Hays DM, Soule EH, Lawrence W, et al. Extremity lesions in the Intergroup Rhabdomyosarcoma Study (IRS-I): a preliminary report. Cancer 1982;49:1–8. doi:10.1002/1097-0142(19820101)49:1<1::aid-cncr2820490102>3.0.co;2.

[4] Bianchi L, Orlandi A. Solid alveolar rhabdomyosarcoma of the hand in adolescence: a clinical, histologic, immunologic, and ultrastructural study. Pediatr Dermatol 1995;12(4):6. doi:10.1111/j.1525-1470.1995.tb00198.x.

[5] Pinkel D, John Pickren J. Rhabdomyosarcoma in children. JAMA 1961;175(4):293–8. doi:10.1001/jama.1961.03040040025005.

[6] Crist WM, Anderson JR, Meza JL, et al. Intergroup Rhabdomyosarcoma Study-IV: results for patients with non metastatic disease. J Clin Oncol. 10.1200/JCO.2001.19.12.3091.

[7] Hayes-Jordana A, Andrassy R. Rhabdomyosarcoma in children. Curr Opin Pediatr 2009;21:373–8. doi:10.1097/MOP.0b013e32832b4171.

[8] Narayana Kurup JK, Kamble VC, Acharya AM, Bhat AK. Massive embryonal rhabdomyosarcoma of the hand in an infant with metastasis at birth: management dilemma. Hand (N Y) 2017;12(5):NP109–12. doi:10.1177/1558944716685827.

[9] Malempati S, Rodeberg DA, Donaldson SS, et al. Rhabdomyosarcoma in infants younger than 1 year: a report from the Children's Oncology Group. Cancer 2011;117(15):3493–501. doi:10.1002/cncr.25887.

[10] Lynched RL, Soule EH, Henderson ED. Pleomorphic rhabdomyosarcomata of the extremities and limb girdles: a clinic pathological study. J Bone Joint Surg Am 1965;47:715–26. https://pubmed.ncbi.nlm.nih.gov/14299664/.

[11] Sabapathy SR, Venkatramani H, Shankar SU, Ramkumar S. Rhabdomyosarcoma of thumb: a case report with review of the literature. Indian J Plast Surg 2007;40:189–93. doi:10.4103/0970-0358.37766.

[12] Siegel HJ, Connor GS, Lee D, Lopez-Ben R, Kelly DR. Synchronous bifocal alveolar rhabdomyosarcoma: a case report. J Bone Joint Surg Br 2006;88:955–8. doi:10.1302/0301-620X.88B7.17508.