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CASE REPORT

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Soft tissue epithelioid hemangioendothelioma on the palm of the hand. Case report

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ABSTRACT

Hemangioendothelioma is a malignant vascular tumor, according to ISSVA classification of vascular tumors. This patient presented an epithelioid hemangioendothelioma; this type of tumor can exhibit significant local destruction, sometimes requiring limb amputation. With deferred Mohs micrographic surgery and reconstructive surgery with multiple conventional and microsurgical techniques, partial or total amputation of the hand was avoided.

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Epithelioid hemangioendothelioma; hand tumor; vascular sarcoma; hand reconstruction

Introduction

Epithelioid hemangioendothelioma(EHE) was initially described by Weiss and Enzinger in 1982 as a soft tissue tumor in the lungs and the liver [1]. Hemangioendotheliomas are classified in papillary intralymphatic angioendothelioma(Dabska tumor), retiform hemangioendothelioma, kaposiform, epithelioid, pseudomyogenic(resembling epithelioid sarcoma) and compound.

EHE has an incidence of 0.038/100,000 per year with a peak of presentation between fourth and fifth decades, and a prevalence of $<$1/1,000,000 cases, showing a slight predominance in women. EHE is extremely unusual in children [2].

It can be found in different locations such as bone, oral cavity, lymph nodes, and mediastinum, among others. It is a rare neoplasm of the vascular endothelium, considered an intermediate between benign epithelioid hemangioma and highly aggressive epithelioid angiosarcoma [3].

Its clinical presentation is not quite predictable when it affects the skin and occurs more in the extremities, trunk, head and neck region [4].

EHE has been described as a solitary lesion, although it can present as multiple lesions, with a clinical spectrum from erythematous or hypopigmented papules or plaques, to nodules with exophytic growth. The tumor is usually painful, can infiltrate adjacent structures like bone.

Histological diagnosis is often difficult as epithelioid endothelial cells can be seen in a variety of vascular lesions, ranging from benign and reactive to neoplastic and completely malignant, nevertheless histology and confirmation with immunohistochemistry for endothelial cell marker is mandatory for an accurate diagnosis [5].

Treatment of choice is complete local excision with or without regional lymphadenectomy, and depending on the infiltration grade, limb amputation may be required [6].

Results

CASE REPORT

A 27-year-old male patient, with no significant medical history, presented to the office with a painful lesion of three and a half years of evolution in the hypothenar region of the right hand(dominant) of 4 $\times$ 4 cms diameter, increasing in size, ulcerated, and indurated, with occasional bleeding and limited wrist flexion and

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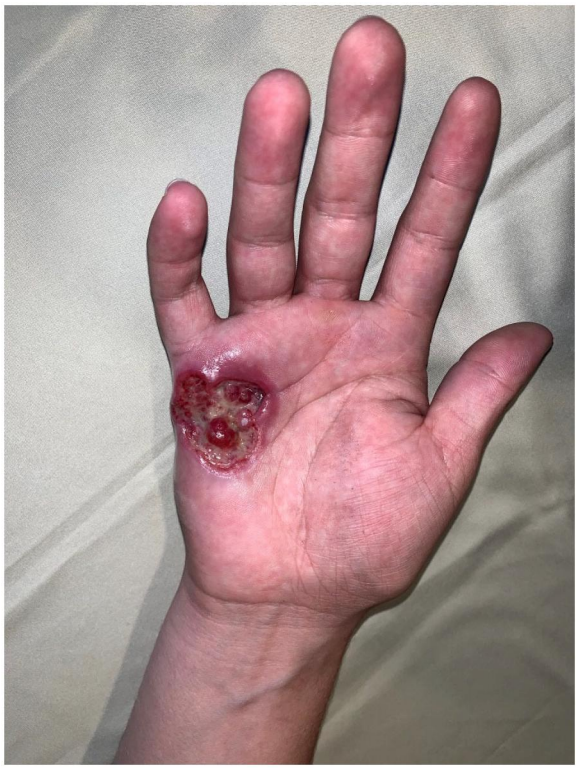

Figure 1. Soft tissue epithelioid hemangioendothelioma on the hand.

extension, without palpable regional adenopathies (Figure 1).

Laboratory results reported hemoglobin 18.5 g/dl, hematocrit 53.5%, WBC 9630, platelet count 279,000, ESR 2, Creatinine 0.96 mg/dl. Hand magnetic resonance reported lesions of the soft tissues of the ulnar region of the hand without bone infiltration.

A biopsy of the lesion was taken, which reported EHE with spindle cell proliferation, and immunohistochemistry reported CD31:focal, CD34:Positive, HHV-8:Negative, CD68:Focal, Ki67:30%, AML, Factor XIIIA, S100-CK5/6, Desmin, HMB-45 and P63:Negative, Giemsa, ZN, Metenam Silver: Negative for pathogens.

After the diagnosis of EHE, an en bloc resection of the tumor was carried out with delayed Mohs surgery (deferred histological evaluation performed on samples fixed in formalin and embedded in paraffin for ensure total resection of the lesion), including: muscles of the hypothenar region, joint capsule, volar plate, ulnar collateral ligament and accessory of the fifth metacarpophalangeal joint (MCP), ulnar vascular-nervous pedicle of the fifth finger, lumbrical and interosseous muscles of the fourth finger, flexor digitorum superficialis(FDS) of the fourth and fifth fingers (Figure 2).

Pathology reported the persistence of the tumor in the proximal region and the distal radial region on the

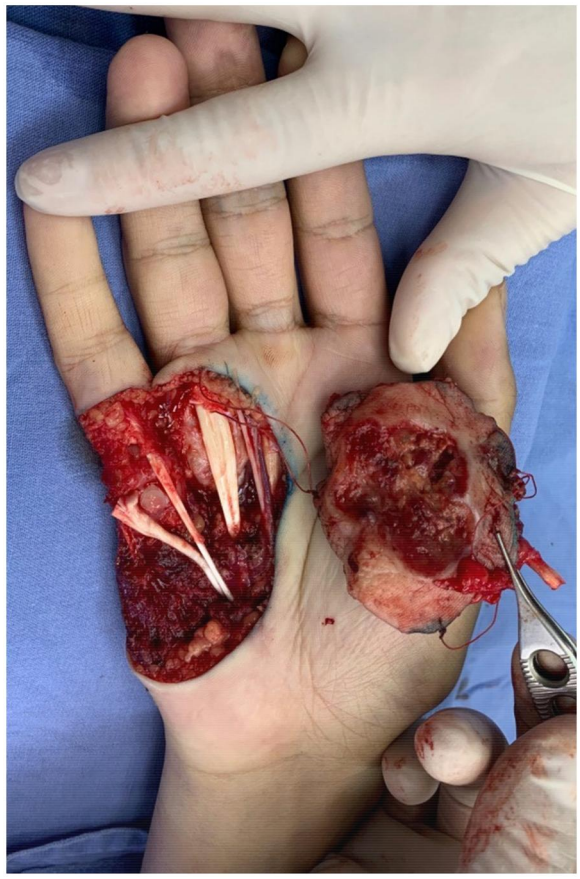

Figure 2. Resection of the tumor.

fourth MCP joint. Margin enlargement was performed with the achievement of complete resection.

To improve the sensitivity of the ulnar region and fifth finger of the hand, reconstructive microsurgery was performed using the ulnar nerve of the fourth finger, making an anastomosis between a branch of this nerve and the fifth proximal phalanx and proximal hypothenar region. FDS tendon graft of the 5th finger was performed to stabilize the fifth MCP joint, FDS tenodesis was performed to the fifth flexor digitorum profundus(FDP) and a proximal reverse fasciocutaneous flap from the radial artery from the forearm as a skin coverage (Figure 3).

The patient was evaluated by the Oncology and Radiotherapy group who did not consider complementary treatment as complete resection was achieved. At nine months of the follow-up a nodular lesion appeared on the palmar side of the third MCP. Extension studies with high-definition ultrasound and Magnetic Resonance evidenced a circumscribed nodular lesion in the subcutaneous tissue at the level of the third A1 pulley without compromising it (Figure 4).

Resection of the tumor up to the A1 pulley of the third finger and the fascia of the radial and ulnar interosseous muscles of the third finger was performed

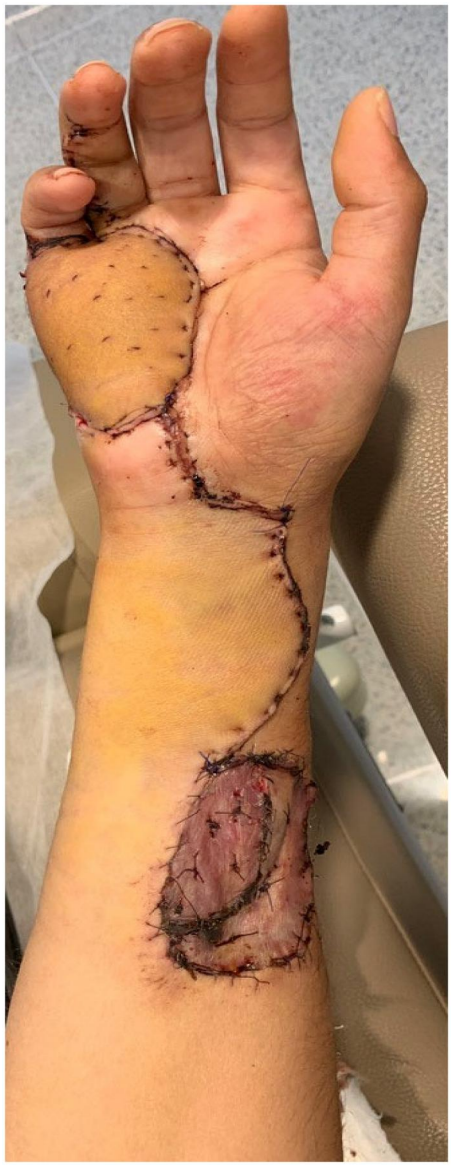

Figure 3. Reconstruction of the defect.

with Mohs surgery, also tenolysis of the flexors of the fourth and fifth fingers to improve flexion contracture; after the negative result of the pathology, the defect was covered with a neurotized island flap from the radial side of the third proximal phalanx to cover the defect. The donor area was reconstructed with a total skin graft (Figure 5).

During the follow-up for three and a half years(every 6 months), no local or distant recurrence has occurred.

To date the patient presents good protective sensitivity on the ulnar side of the fifth finger, the mobility of the fourth and fifth fingers is limited to full extension, however, the patient performs all kinds of activities and is currently working as an operator in the same job (Figure 6).

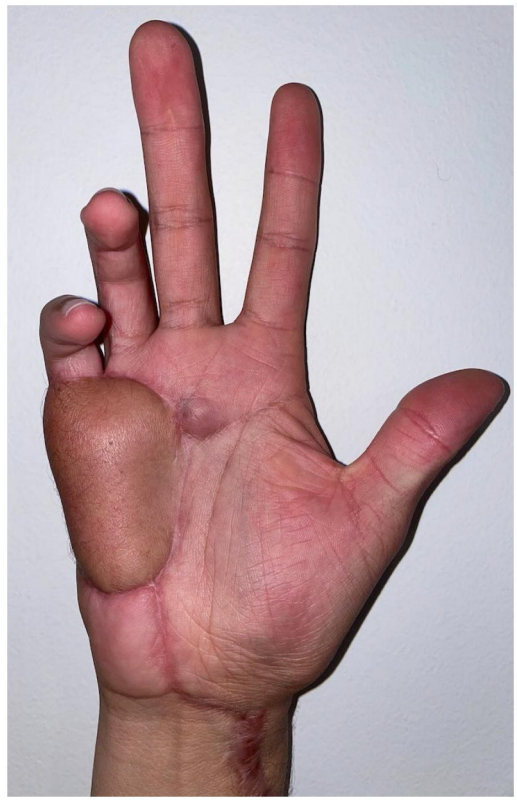

Figure 4. new lesion 9 months postoperatively.

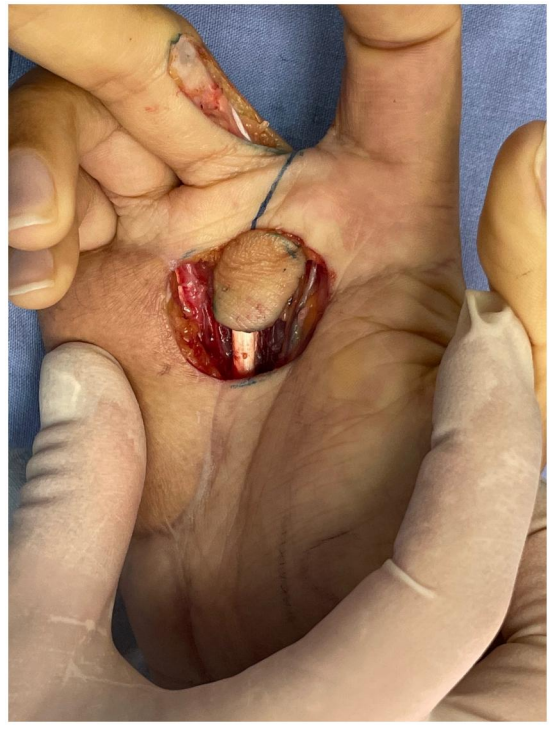

Figure 5. Reconstruction with a Neurovascular pedicled flap.

Discussion

EHE is a low-grade malignant or borderline vascular neoplasm developed from a vascular structure, usually

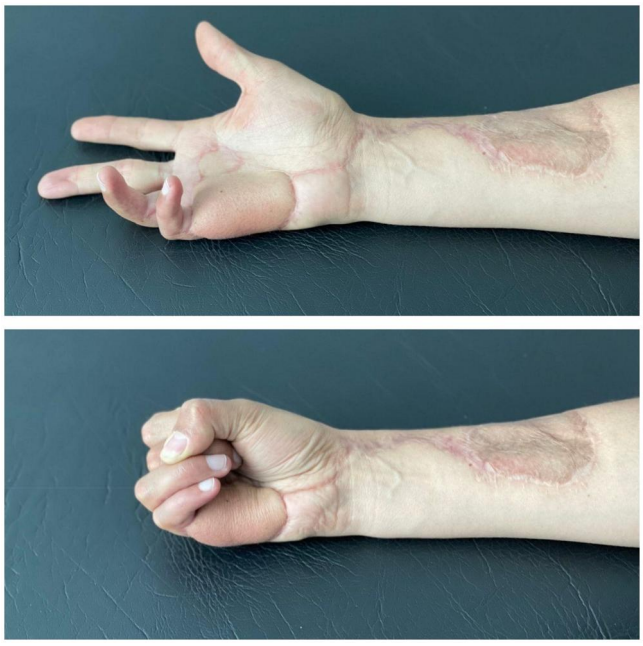

Figure 6. Current evaluation three and a half years postoperatively..

a vein, whose occlusion can cause obstructive symptoms such as edema and thrombophlebitis [1,7–12]. It represents 1% of all vascular neoplasms and is locally aggressive [3].

The initial diagnosis may be incidental in asymptomatic patients. In symptomatic cases, the most common presentation is pain (40%), palpable mass (6-24%) and weight loss(9%) [6,8,13].

Skin lesions often present an exophytic growth pattern, from asymptomatic single dermal nodules to hypopigmented or erythematous papules or excoriated plaques. The lesions commonly exhibit an infiltrating and more aggressive growth pattern [6,8,9,14]. Pain is usually severe, burning, hyperesthetic and accompanied by hyperhidrosis due to reflex sympathetic hyper-reactivity [2].

However, more than 50% of patients have metastatic disease, with lung, liver and bone involvement [8,9].

Microscopically, it is a poorly defined tumor, characterized by the presence of oval or polygonal endothelial cell aggregates with an epithelioid appearance, with eosinophilic cytoplasm and a rounded nucleus, within a myxoid stroma [2,13].

Initial staging consists whole-body imaging, including the brain and extremities, using computed tomography(CT), magnetic resonance imaging(MRI), or PET/CT [2].

Differential diagnoses include epithelioid sarcoma (ES), epithelioid angiosarcoma and pseudomyogenic hemangioendothelioma (PE).

Treatment of choice is wide local excision with or without regional lymphadenectomy [8]. Limb amputation is limited to cases where the tumor has a high grade of infiltration [15].

In the evaluation of this case, we did not find any other published article related to EHE in the palm of the hand. Some cases of the radial artery and the superficial palmar arch in the hand have been reported [14,15], involving the nail and the bone in the hand [10–12,16,17].

Mohs surgery ensures complete resection, although it is difficult to perform due to the different levels of the structures, making the diagnosis at their edges a challenge.

Recurrence nine months later, in the same place where the biopsy was positive, makes us think how difficult the evaluation is due to the complex anatomy of the hand. This is the second article published that involves Mohs surgery for tumor resection [18].

Hand salvage reconstructive surgery in oncological cases is a challenge to provide functionality before amputation.

Because of the low incidence of bone involvement (less than 1% of vascular tumors) and the variability of malignancy, a therapeutic protocol is difficult to adopt [11]. No active systemic agents are currently approved for EHE, which is typically refractory to the antitumor drugs used in sarcomas. The grade of uncertainty in selecting the most appropriate therapy for EHE patients and the lack of guidelines on the clinical management of the disease make the

adoption of new treatments inconsistent across the world, resulting in suboptimal outcomes for many EHE patients [2,9].

The role of radiotherapy is controversial and there are very few series of patients reported with favorable results; this therapy is reserved for cases in which complete resection was not possible [2].

Regarding the prognosis, there is no clear association between the size of the tumor, histological features, and clinical progress. In some studies, a high mitotic rate (>6 mitoses/10 HPF) was correlated with a poor prognosis.

Follow-up is important because local recurrences appear in up to 13% of cases, distant metastasis in 31% (50% to regional lymph nodes) and mortality is 13%.

Ethical approval

Not required

Disclosure statement

No potential conflict of interest was reported by the author(s).

References

[1] Weiss SW, Ishak KG, Dail DH, et al. Epithelioid hemangioendothelioma and related lesions. Semin Diagn Pathol. 1986; 3(4):259–287.

[2] Stacchiotti S, Miah AB, Frezza AM, et al. Epithelioid hemangioendothelioma, an ultra-rare cancer: a consensus paper from the community of experts. ESMO Open. 2021;6(3):100170. doi: 10.1016/j.esmoop.2021.100170.

[3] Mentzel T, Beham A, Calonje E, et al. Epithelioid hemangioendothelioma of skin and soft tissues: clinicopathologic and immunohistochemical study of 30 cases. Am J Surg Pathol. 1997;21(4):363–374. doi: 10.1097/00000478-199704000-00001.

[4] Schonauer F, Cavaliere A, Pezone G, et al. Epithelioid hemangioma of the nose: a challenging diagnosis. Healthcare. 2022;10(4):633. doi: 10.3390/healthcare10040633.

[5] Accardo G, Nele G, Turrà F, et al. A rare case of "histiocytoid haemangioma" of the hand. J Plast Reconstr Aesthet Surg. 2013;66(11):e318–20–e320. doi: 10.1016/j.bjps.2013.04.027.

[6] Gherman CD, Fodor D. Epithelioid hemangioendothelioma of the forearm with radius involvement. Case report. Diagn Pathol. 2011;6(1):120. doi: 10.1186/1746-1596-6-120.

[7] García-Arpa M, Rodríguez-Vázquez M, Cortina P, et al. Cutaneous epithelioid hemangioendothelioma. Actas Dermosifiliogr. 2005; 96(6):386–391. doi: 10.1016/s0001-7310(05)73097-0.

[8] Flucke U, Vogels RJC, de Saint Aubain Somerhausen N, et al. Epithelioid hemangioendothelioma: clinicopathologic, immunhistochemical, and molecular genetic analysis of 39 cases. Diagn Pathol. 2014; 9(1):131. doi: 10.1186/1746-1596-9-131.

[9] Rosenbaum E, Jadeja B, Xu B, et al. Prognostic stratification of clinical and molecular epithelioid hemangioendothelioma subsets. Mod Pathol. 2020;33(4):591–602. doi: 10.1038/s41379-019-0368-8.

[10] Kikuchi K, Watanabe M, Terui T, et al. Nail-destroying epithelioid haemangioendothelioma showing an erythematous scar-like appearance on the finger. Br J Dermatol. 2003; 148(4):834–836. doi: 10.1046/j.1365-2133.2003.05254.x.

[11] Patel MR, Srinivasan KC, Pearlman HS. Malignant hemangioendothelioma in the hand: a case report. J Hand Surg Am. 1978;3(6):585–588. doi: 10.1016/s0363-5023(78)80010-0

[12] Bruegel M, Waldt S, Weirich G, et al. Multifocal epithelioid hemangioendothelioma of the phalanges of the hand. Skeletal Radiol. 2006;35(10):787–792. doi: 10.1007/s00256-005-0943-6.

[13] Clarke LE, Lee R, Militello G, et al. Cutaneous epithelioid hemangioendothelioma. J Cutan Pathol. 2008; 35(2):236–240. doi: 10.1111/j.1600-0560.2007.00790.x.

[14] Castelli P, Caronno R, Piffaretti G, et al. Epithelioid hemangioendothelioma of the radial artery. J Vasc Surg. 2005; 41(1):151–154. doi: 10.1016/j.jvs.2004.10.036.

[15] Hampers DA, Tomaino MM. Malignant epithelioid hemangioendothelioma presenting as an aneurysm of the superficial palmar arch: a case report. J Hand Surg Am. 2002; 27(4):670–673. doi: 10.1053/jhsu.2002.33707.

[16] Atalar H, Armangil M, Uzüm N, et al. Solitary epithelioid hemangioendothelioma of the metacarpal. Eklem Hastalik Cerrahisi. 2013;24(3):182–186. doi: 10.5606/ehc.2013.39.

[17] Kitagawa Y, Ito H, Iketani M, et al. Epithelioid hemangioendothelioma of the phalanx: a case report. J Hand Surg Am. 2005;30(3):615–619. doi: 10.1016/j.jhsa.2004.11.008.

[18] Park SM, Kim HS, Ko HC, et al. Cutaneous epithelioid hemangioendothelioma treated with mohs micrographic surgery. Int J Dermatol. 2017; 56(1):97–99. doi: 10.1111/ijd.13299.