Available online at www.sciencedirect.com

ScienceDirect

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

Case Report

Pseudomyogenic (Epithelioid sarcoma-like) hemangioendothelioma with bone invasion

Ahmet Karakasli ^{a,*}, Ahmet Karaaslan ^b, Mehmet Erduran ^a,
Sercan Capkin ^a, Emine Burcin Tuna ^c, Hasan Havitcioglu ^a

^a Dokuz Eylul University, Faculty of Medicine, Department of Orthopedics and Traumatology, 35340 Izmir, Turkey

^b Sifa University, Faculty of Medicine, Department of Orthopedics, 35100 Izmir, Turkey

^c Dokuz Eylul University, Faculty of Medicine, Department of Pathology, 35340 Izmir, Turkey

ARTICLE INFO

Article history:

Received 11 February 2014

Accepted 29 June 2014

Available online 30 July 2014

Keywords:

Phalanx

Bone invasion

Hemangioendothelioma

ABSTRACT

Aim: Pseudomyogenic (epithelioid sarcoma-like) hemangioendothelioma is a rare local aggressive vascular tumor. Herein we present a 54-year-old male patient with a tumor on his index finger.

Case: The patient presented with a 1-year history of pain and swelling that progressively exacerbated. Bone invasion was observed on the middle phalanx via direct radiography. Histopathological examination findings were compatible with epithelioid sarcoma-like hemangioendothelioma.

Conclusion: To the best of our knowledge is the first case report of epithelioid sarcoma-like hemangioendothelioma with bone invasion.

Copyright © 2014, Professor P K Surendran Memorial Education Foundation. Publishing Services by Reed Elsevier India Pvt. Ltd. All rights reserved.

1. Introduction

Epithelioid sarcoma-like hemangioendothelioma (ES-H) is a rare vascular tumor with a low degree of aggressiveness.¹ It often originates in the peripheric vessels.² Clinically, it imitates epithelioid sarcoma,³ and can be differentiated from epithelioid sarcoma on the basis of immunohistological and histochemical differences.⁴ ES-H can exhibit local aggressive behavior, but has never been observed to metastasize.⁴ ES-H is not specific to any particular location and can develop in any soft tissue of the body⁵; however, bone invasion is an extraordinary situation that might necessitate aggressive surgical treatment.

2. Case report

A 54-year-old male presented to Izmir Dokuz Eylul University, Orthopedics and Traumatology Department with a 1-year history of a painful mass on the right index finger that progressively increased in size. On physical examination an immobile lesion with irregular borders was observed on the volar surface of the proximal and middle phalanges of the right index finger. The lesion was painful upon palpation and hypoesthesia was noted on the ulnar side. Direct radiography showed the soft tissue shadow of a bone-destroying lesion located on the volar aspect of the proximal and middle phalanges of the index finger (Fig. 1b). MRI showed a hyperintense

* Corresponding author.

E-mail address: akarakasli@mailcity.com (A. Karakasli).

<http://dx.doi.org/10.1016/j.jor.2014.06.019>

0972-978X/Copyright © 2014, Professor P K Surendran Memorial Education Foundation. Publishing Services by Reed Elsevier India Pvt. Ltd. All rights reserved.

Fig. 1 – a. MRI of the epithelioid sarcoma-like hemangioendothelioma. b. X-ray of the pseudomyogenic hemangioendothelioma.

12 × 22-mm lesion that was destroying the middle phalanx of the index finger (Fig. 1a).

The tumor was surgically removed and the surgical borders were irregular in shape. The tumor had extended along the neuromuscular band and invaded the neuromuscular band on the ulnar side (Fig. 2a and b). The entire tumor was surgically removed, while concentrating on protecting the neuromuscular band. During the removal of the tumor a bone specimen was obtained from the middle phalanx ulnar cortex. Macroscopically, the tumor measured 25 × 25 × 15 mm, the external surface had lobular contours and was smooth, and it had a solid irregular appearance. After serial sections were performed, it was determined that the tumor was grayish white and had a lobular appearance.

Histopathological examination showed that the tumor consisted of eosinophilic cytoplasmic cells that were generally round epithelioid in appearance with irregular borders. The existence of collagen attracts attention among the cells. Epithelioid cells were, in places, accompanied by cells with spindle-shaped morphology (Fig. 3a). In the light of the initial

microscopic findings, hemangioendothelioma-like epithelioid sarcoma, epithelioid sarcoma, epithelioid hemangioendothelioma, and epithelioid schwannoma were differential diagnostic considerations. Immunohistochemically, the neoplastic cells were S100-, CK7-, CK19-, and Cyclin D1-positive. Additionally, these cells exhibited focal staining with CD31 and creatine, with high molecular weight, but did not stain with CD117, desmin, CD34, HMB-45, calretinin, Melan-A, or CEA. K67 proliferative activity was approximately 30%. The final diagnosis of epithelioid sarcoma-like hemangioendothelioma (ES-H) was based on simultaneous evaluation of the morphological and immunohistochemical findings.^{3,4}

Despite removal of the entire tumor, 7 months post surgery the patient had recurrence and index finger ray amputation was recommended; however, the patient did not agree to undergo the procedure. Consequently, the recurrent mass was surgically removed, but 3 months later the tumor recurred again at the same location and radiological examination showed that bone invasion had increased; therefore, index finger ray amputation was performed with the patient's

Fig. 2 – a. Pre-operative view of epithelioid sarcoma-like hemangioendothelioma in the right index finger. b. Intra-operative view of the epithelioid sarcoma-like hemangioendothelioma.

Fig. 3 – a. Histopathologic view of the epithelioid sarcoma-like hemangioendothelioma (H&E, 20×). b. Black arrows indicate bone invasion of the epithelioid sarcoma-like hemangioendothelioma.

consent. Histopathological examination showed that bone invasion of the hemangioendothelioma-like epithelioid sarcoma (Fig. 3b).

3. Discussion

ES-H is a low degree vascular tumor similar to epithelioid sarcoma in terms of clinical and histopathological characteristics⁶; however, it is not associated with distant metastasis and can be differentiated from epithelioid sarcoma based on some immunohistochemical indicators.^{6–8} The differential diagnosis has, together with epithelioid sarcoma, hemangioendothelioma, as well.^{7,8} Although CD31 positivity is focal, due to positivity of other determinants the tumor is thought to be compatible with hemangioendothelioma-like epithelioid sarcoma (ES-H).

Local recurrence developed in the presented case 7 months after surgical excision of the tumor, and the mass was surgically removed again; however, it recurred again 3 months later and his right index finger underwent ray amputation. To the best of our knowledge the literature includes 10 cases,^{1–4} in which the lesion had one localization in seven cases (the chest wall, scalp, cubital fossa, knee, and calf). In one case the lesion originated from the radial artery.⁸ Among these 10 cases, in two the lesion had two localizations: one in the elbow and forearm, one in the forearm and index finger, and two lesions in the thigh in the third case.

As reported, 3 of the 10 earlier cases had > one lesion and recurrence occurred in 3 of the 10 cases, but distant metastasis did not occur in any of the cases nor did bone invasion. There was a lesion destroying the middle phalange of the right index finger in the presented patient. During the patient's post ray amputation follow-up recurrence and distant metastasis did not occur.

According to the 10 previously reported cases and the case presented herein, ES-H doesn't exhibit a gender predisposition (7 males and 4 females). In terms of tumor localization, ES-H can occur in any soft tissue region of the body. The presented case had a similar presentation to that reported by Watabe et al,² who reported a lesion on the dorsal base of the right index finger and on the flexor aspect of the right forearm. Local recurrence developed in their case and the presented

case, both consequently undergoing amputation. The presented case differs from the earlier reported cases in that bone destruction was observed. We think that these lesions might not be as innocuous as generally thought and that treatment of ES-H might at times be extremely complicated. Angiographic embolization and stereotaxic radiosurgery should be considered in the treatment of lesions of vascular origin; however, these methods can damage peripheral vessels.

In conclusion, ES-H should be diagnosed immunohistochemically, and clinicians must be aware that bone destruction can occur in association with lesions localized at the end regions of the extremities, which due to a high recurrence rate might require amputation.

Conflicts of interest

All authors have none to declare.

REFERENCES

1. Billings SD, Folpe AL, Weiss SW. Epithelioid sarcoma like hemangioendothelioma. *Am J Surg Pathol.* 2003;27:48–57.
2. Watabe A, Okuyama R, Hashimoto A, et al. Epithelioid sarcoma-like haemangioendothelioma: a case report. *Acta Derm Venereol.* 2009;89:208–209.
3. Tokyol C, Uzüm N, Kuru I, Uluoglu O. Epithelioid sarcoma-like hemangioendothelioma: a case report. *Tumori.* 2005 Sep–Oct;91:436–439.
4. Weiss SW, Goldblum JR, Enzinger, Weiss's. *Soft Tissue Tumors.* 5th ed. St. Louis: Mosby-Elsevier; 2008:698–701.
5. Chase DR, Enzinger FM. Epithelioid sarcoma: diagnosis, prognostic indicators, and treatment. *Am J Surg Pathol.* 1985;9:241–263.
6. Miettinen M, Fanburg-Smith JC, Virolainen M, Shmookler BM, Fetsch JF. Epithelioid sarcoma: an immunohistochemical analysis of 112 classical and variant cases and a discussion of the differential diagnosis. *Hum Pathol.* 1999;30:934–942.
7. Traweek ST, Kandalaft PL, Mehta P, Battifora H. The human hematopoietic progenitor cell antigen (CD34) in vascular neoplasia. *Am J Clin Pathol.* 1991;96:25–31.
8. Castelli P, Caronno R, Piffaretti G, Tozzi M. Epithelioid hemangioendothelioma of the radial artery. *J Vasc Surg.* 2005;41:151–154.