

Epithelioid hemangioendothelioma of the subcutaneous tissues of a finger

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ABSTRACT

Epithelioid hemangioendotheliomas (EHEs) are known to have a variable malignant potential. EHEs are rarely seen in the hand and there is no consensus about their management. The options include excision, excision followed by adjuvant radiotherapy and amputation. In this paper, we report a case of EHE of a finger that was treated by excision. Although the tumor had ill-defined borders and there was histological evidence of tumor extension to all resection margins, no local recurrence or metastasis were noted during the 3 years of followup. The literature is reviewed and an argument is made that EHEs of the hand may have a more benign behavior compared with EHEs of the lower limbs and viscera.

Key words: Finger, hand, vascular tumor, epithelioid hemangioendothelioma

MeSH terms: Fingers, tumor, hemangioma, epithelioid

INTRODUCTION

Epithelioid vascular tumors are rare tumors that are characterized by the presence of epithelioid endothelial cells. There are three distinct types of epithelioid vascular tumors: Epithelioid hemangioma, epithelioid hemangioendothelioma (EHE) and epithelioid angiosarcoma.^{1,2} The former type is benign and the later behaves as a malignant angiosarcoma. The behavior of EHE is intermediate between the other two types and there is no consensus about the management of these tumors in hand. In hand, management varied from amputation^{3,4} to en-block or wide excision.⁵⁻⁸ Some authors have also used adjuvant radiotherapy following excision of EHE of the hand.^{9,10}

We report a case of EHE of a finger that was treated by excision alongwith review of literature.

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CASE REPORT

An 8-year-old girl presented with a 6-month history of slowly growing mass on the volar aspect of the right middle finger [Figure 1a]. An incisional biopsy was done at a local hospital, but it was not conclusive and the specimen was not available for reexamination. Clinically, the mass was fixed to the dermis and the flexor sheath. However, there was no numbness and there was full range of motion of the finger joints. A magnetic resonance imaging showed an enhancing lesion that is adherent to the flexor sheath [Figure 1b]. Excisional biopsy was done through a zigzag volar incision. Intraoperatively the tumor was found to have ill defined borders. It was adherent to the dermis, flexor sheath and both digital nerves. Part of the flexor sheath was resected and the tumor was shaved off from the dermis by sharp dissection. No skin excision was done and both digital nerves were preserved. The tumor size was 2 × 1.5 cm [Figure 1c]. Postoperatively, a small area of the skin flap became necrotic and this was treated conservatively [Figure 1d]. Histological examination showed all the classic features of EHE. The tumor consisted of numerous epithelioid cells which are arranged in clusters, trabeculae and cords in a myxoid background [Figure 1e]. Another characteristic histological feature of this tumor is the presence of intracytoplasmic vacuoles [Figure 1f]. The mitotic rate was 3/10 high power fields (HPF) with evidence of focal pleomorphism. The tumor cells were extended to all resection margins. A panel of immune histochemical stains was done and

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Figure 1: (a) Preoperative clinical photograph showing the tumour mass (b) preoperative magnetic resonance imaging showing an enhancing lesion that is adherent to the flexor sheath (c) intraoperative view showing the excised tumour. (d) early postoperative view showing a small area of skin necrosis (e) histology showing the epithelioid tumor cells (H and E, $\times 200$) (f) the arrow points at intracytoplasmic vacuoles (H and E, $\times 600$) (g) positive immune stain to CD34 (brown color, $\times 200$) (h and i) Clinical photographs 3 years after surgery showing no recurrence full flexion

showed positive staining of tumor cells with CD34, CD31, and factor VIII [Figure 1g]. Tumor cells were negative for other markers confirming the endothelial origin of the tumor. We considered the malignant potential of the tumor and the positive resection margins and we offered the patient further management in the form of wide resection of volar soft tissues and flap reconstruction. The family refused and elected for followup. The followup protocol was local examination and radiological workup for metastasis every 6 months. The patient is now 3 years after surgery with no evidence of local recurrence or distant metastasis [Figure 1h and i].

DISCUSSION

Most reported cases of EHE were found in the lower limbs and viscera.² Outside the hand, EHE are known to have a local recurrence rate of 13%, a metastasis rate of 31%

and a mortality rate of 13%.¹¹ A review of the literature of EHE of the hand³⁻¹⁰ showed the rare occurrence of local recurrence⁸ and zero rates of metastasis and mortality. It is well known that the malignant potential of visceral EHE is higher than EHE of the lower limbs.¹¹ Our review of the literature indicates that EHE of the hand may have a more benign behavior and this is supported by our case report. Another factor in the behavior of soft tissue tumors with malignant potential is the size and the degree of mitoses. According to World Health Organization,¹² excellent prognosis is expected if the size of the tumor is less than 3 cm and the mitotic figures were ≤ 3 mitoses per 50 HPF. Our tumor was 2 cm in size, but the mitotic rate was 3/10 HPF.

EHE of the hand may be classified in three subtypes: Bony, intravascular and soft tissue types. EHE of the bones of the hand may either be unicentric or

multicentric,³ and the tumor usually involves the phalanges. Simple curettage and bone grafting is usually curative³ and if the tumor recurs it can be management by en block excision and autogenous reconstruction.⁸ Intravascular EHE of the hand are very rare and are treated by excision of the involved segment of the vessel and revascularization of the involved digits (if required).⁹ EHE of the soft tissues of the hand usually present as a slowly growing infiltrative mass (similar to our case)⁵⁻⁷ or it may have a scar like appearance within the nail bed.⁴ Most hand surgeons have treated these soft tissue tumors with excision preserving the adherent vital structures.⁵⁻⁷ No postoperative radiotherapy was used and all patients did not have local recurrence or metastasis.⁵⁻⁷ Our case is an important addition to the existing body of literature because we document mitotic figures/pleomorphism and positive resection margins and yet no local recurrence or metastases were noted. We believe that EHEs of the hand have a more benign behavior. Hence, excision preserving adherent vital structures is a viable option of management of soft tissue EHEs of the hand; especially when complete excision requires an amputation or significant loss of function. However, careful followup for local recurrence and metastasis is still mandatory.

The authors are aware that based on one case experience, one cannot recommend a treatment plan of marginal excision in lesions that have a potential aggressive behavior. However, the main argument behind this case report is the suggestion that the behavior of EHE of the soft tissues is probably much more benign in the hand compared with other anatomical sites. This suggestion is supported not only by the current case report but also by all other previous cases reported in hand.⁵⁻⁷ The concept of tumor behaving differently in the hand versus other anatomical sites is not new and is well described for giant cell tumors of bone. Giant cell tumors of the jaws have a very benign behavior as long as the lesion is less than 5 cm with no cortical perforation.¹³ Hence, simple curettage or even intralesional triamcinolone injections are considered acceptable forms of management for such jaw tumors.¹⁴ In contrast, giant cell tumors of the small bones of the hand are usually more aggressive in behavior and hence more aggressive treatment is recommended.¹⁵ The authors believe that such an anatomically based behavior may also exist for EHE.

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