

## SHORT COMMUNICATIONS

## Successful Treatment of Recurrent Epithelioid Sarcoma-like Hemangioendothelioma with Mohs Micrographic Surgery

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### INTRODUCTION

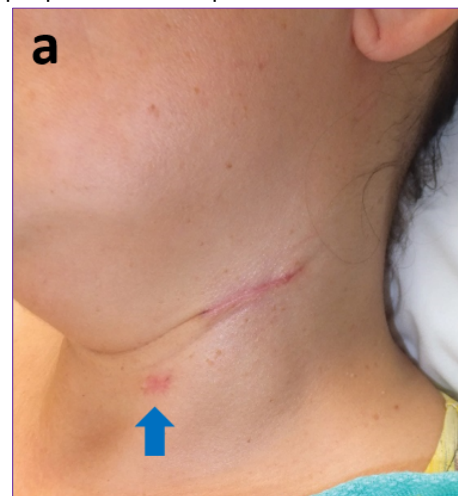
Epithelioid sarcoma-like hemangioendothelioma (ES-H), also known as pseudomyogenic hemangioendothelioma, is an unusual vascular tumor that often presents as a deep-seated subcutaneous nodule. We present the case of a locally recurrent ES-H treated with Mohs micrographic surgery.

### CASE PRESENTATION

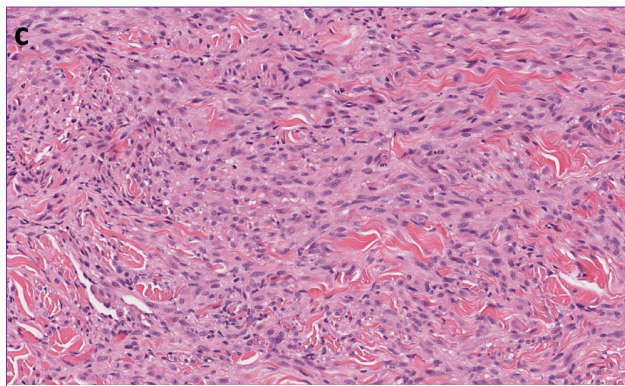
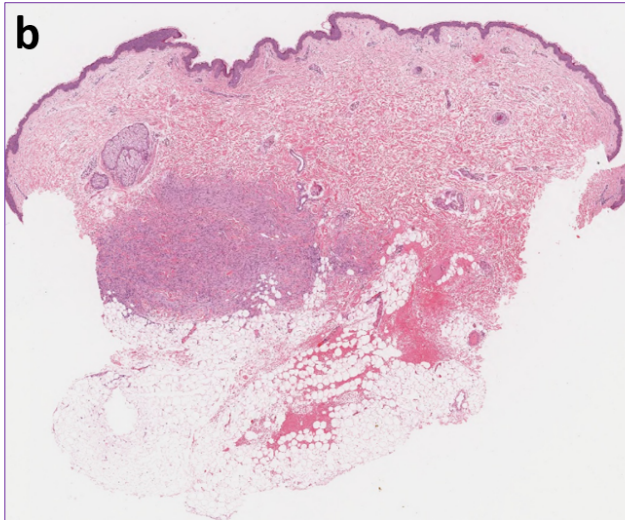
A 42-year-old woman presented with a flesh-colored nodule along her left anterior neck that doubled in size over a four-month period. This lesion was previously excised by an outside provider one month prior but had rapidly recurred. Histopathology from the prior excision was consistent with ES-H. Her medical history was otherwise unremarkable. Review of systems was negative for fevers, chills, night sweats, or weight changes, and was otherwise unremarkable. On examination, the patient had a 3cm x 0.8cm pink, indurated plaque along her prior excision scar and a 1.2 cm pink plaque anterior to the scar on the left neck (Figure 1A). The remainder of the physical examination was unremarkable. Laboratory testing was unremarkable. A biopsy of the anterior 1.2cm lesion revealed

sheets and nests of plump, spindled cells with dense eosinophilic cytoplasm at the deep dermis-subcutaneous junction with rare scattered neutrophils and peripheral collagen trapping (Figure 1B, C). Immunohistochemical stains for Cytokeratin AE1, AE3, and CD31 were diffusely positive within the involved cells and negative for SMA, desmin, EMA, Factor XIIIa, and S100, consistent with a local recurrence of ES-H. CT scans of the neck, chest, abdomen, and pelvis were unrevealing. The lesion was excised with Mohs micrographic surgery with negative margins, and the patient remained without evidence of recurrence at 9 months' follow-up.

**Figure 1. (A)** Clinical image of the subcutaneous nodular lesions on the lateral left neck. The arrow indicates the 1.2cm plaque that was biopsied.



**Figure 1. (B) and (C),** skin biopsy specimen revealing spindle cells at the dermis-subcutis junction, scattered neutrophils, and collagen trapping. **(B)** – Original Magnification 4x. **(C)** – Original Magnification 100x.



## DISCUSSION

ES-H is an uncommon low-grade vascular tumor most commonly presenting as a deep-seated firm dermal to subcutaneous nodule with occasional involvement of the overlying epidermis.<sup>1-3</sup> The most common primary sites are in the lower extremities, although it can involve a variety of locations including bony structures.<sup>3,4</sup>

Microscopically, ES-H lesions demonstrate sheets and fascicles of round, epithelioid cells infiltrating soft tissues. These tumor cells typically have abundant strongly eosinophilic cytoplasm, large vesicular nuclei, prominent nucleoli, and lack morphologic signs of vascular

differentiation. Immunohistochemical stains are positive for keratin AE1/AE3, ERG, FLI1, and FOSB, variable staining for CD31, and negative stains for desmin, CD34, S100, CD68, and Factor XIII. Occasional cases may stain focally positive for SMA.

ES-H is typically indolent with as a low chance of metastasis to regional or distant locations and a generally favorable prognosis.<sup>1,3</sup> Treatment generally consists of surgical excision with clear margins although local recurrence may occur in up to 60% of cases.<sup>3,4</sup> Radiotherapy and chemotherapy have also been trialed and recent reports suggest that ES-H may respond to mTOR inhibitors like everolimus or sirolimus.<sup>5,6</sup> To our knowledge, this is the first reported case of ES-H successfully treated with Mohs micrographic surgery, and the patient remains without clinical evidence of recurrence 9 months after treatment. Given the high recurrence rates of ES-H and excellent margin control that Mohs micrographic surgery offers, it should be considered as a treatment modality for ES-H.

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