Growing Subcutaneous Mass on the Thigh

Catherine S. Yang, MD; Leslie Robinson-Bostom, MD; Helena O. Taylor, MD, PhD; Lionel Bercovitch, MD; Shoshana Landow, MD



A 55-year-old woman with a history of basal cell carcinoma and hypothyroidism presented with a changing red patch on the right upper thigh. The patch had been present since birth without symptoms until 3 months prior to presentation when the patient noted that her pants fit tighter, despite stable weight. The lesion was not painful, pruritic, or hyperhidrotic. On examination the right thigh appeared larger than the left thigh. There was a 12×14-cm red patch overlying a 12×18-cm rubbery lobulated swelling on the lateral aspect of the right thigh. Ultrasonography was performed and was normal. Magnetic resonance imaging showed a vascular malformation superficial to fascia.

What's the diagnosis?

- a. angiosarcoma
- b. eccrine angiomatous hamartoma
- c. epithelioid hemangioendothelioma
- d. glomuvenous malformation
- e. kaposiform hemangioendothelioma

From the Warren Alpert Medical School of Brown University, Providence, Rhode Island. Drs. Yang, Robinson-Bostom, Bercovitch, and Landow are from the Department of Dermatology; Dr. Robinson-Bostom also is from the Division of Dermatopathology; and Dr. Taylor was from the Department of Plastic and Reconstructive Surgery. Dr. Taylor currently is from the Department of Surgery, Division of Plastic Surgery, Mount Auburn Hospital, Cambridge, Massachusetts.

The authors report no conflict of interest.

Correspondence: Catherine S. Yang, MD, Department of Dermatology, Rhode Island Hospital, 593 Eddy St, APC 1018, Providence, RI 02903 (catherine_yang@brown.edu).

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The Diagnosis: Eccrine Angiomatous Hamartoma

iven the progression of symptoms 3 months prior to presentation, an excisional biopsy was performed (Figure 1). Hematoxylin and eosin staining showed prominent eccrine sweat glands and vessels surrounded by superficially located adipose tissue in the mid and deep dermis (Figure 2).

Eccrine angiomatous hamartoma (EAH) is an uncommon benign tumor typically located on the arms and legs or trunk. It is usually solitary, though cases with multiple lesions have been reported. Most cases are diagnosed in childhood as either congenital or acquired lesions. However, EAHs can develop in adulthood and have been described in patients up to 70 years of age. The median age of diagnosis is 10 years, indicating that EAH is primarily a pediatric tumor. There is no gender predilection.

Approximately 35% to 66% of patients report pain, pruritus, or hyperhidrosis associated with EAHs, though this incidence may be overrepresented because patients tend to present when the lesions become symptomatic.²⁻⁵ The pain is attributed to nerve fibers infiltrating the tumor. Hypertrichosis also has been described and is thought to be due to hair follicles within the hamartoma.

Histologically, EAHs are characterized by normal-appearing eccrine glands mingled with venules and capillaries. Additional variable pathologic findings include lipomatous, pilar, lymphatic, or mucinous features.² Other vascular anomalies

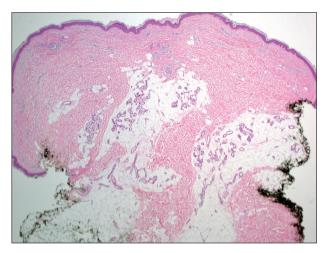
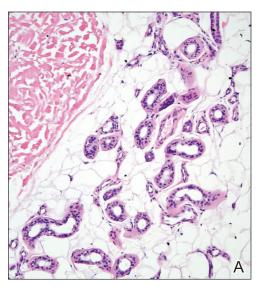


Figure 1. An excisional biopsy demonstrated prominent eccrine glands and vessels surrounded by adipose tissue in the mid and deep dermis (H&E, original magnification ×4).

such as hemangiomas or arteriovenous malformations occasionally have been described in association with EAH. The vessels stain for ulex europaeus 1 and factor VIII. Eccrine glands stain for S-100 protein, carcinoembryonic antigen, epithelial membrane antigen, and cytokeratin CAM 5.2.



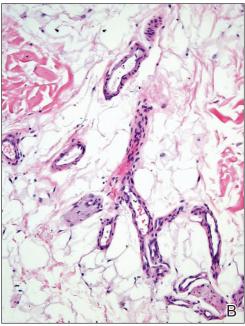


Figure 2. Eccrine sweat glands (A) and capillaries and venules (B) appeared normal (both H&E, original magnification $\times 10$).

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In light of a publication proposing that EAH is a lymphatic proliferation,⁶ a D2-40 stain was performed on the specimen and was negative.

Eccrine angiomatous hamartoma has been reported to grow mainly during childhood, puberty, or pregnancy, presumably due to hormonal influences. There are few reports of EAH enlarging in middle-aged adults, and even fewer without pain during the growth phase. It is unclear what triggered the growth in our otherwise healthy postmenopausal patient.

Eccrine angiomatous hamartoma does not have malignant potential and thus treatment is optional and based on relief of symptoms. Simple excision of the EAH usually is curative, but recurrences can occur.⁴ Botulinum toxin also has been used to treat hyperhidrosis in tumors that are too large for resection. Treatment with lasers such as the pulsed dye laser and Nd:YAG laser has not been successful.⁸ A case of spontaneous regression has been reported.¹

Liposuction was considered in our patient given the substantial adipose tissue on biopsy. The patient ultimately declined treatment. This case highlights that EAH can present in adulthood and should be considered in the differential diagnosis of an enlarging but otherwise asymptomatic vascular tumor.

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