

# DERMATOPATHOLOGY UNIT

## UPP | Department of Dermatology

UPMC Dermatopathology "Case of the Month" Presentations

UPP - Department of Dermatology, Dermatopathology Unit

5230 Centre Avenue (412) 623-2614

Pittsburgh, PA 15232 (412) 682-6450 FAX

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*Authors: Philina M. Lamb, M.D., Michael Zhang, M.D., PhD., and Drazen Jukic M.D., PhD.*

### OCTOBER 2003 CASE OF THE MONTH

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#### FINDINGS

An 84-year old white female presented to the surgery clinic for re-excision of a melanoma-in-situ involving her left upper arm. Incidentally, she was noted to have a 0.7 x 0.4 cm dark blue nodule on the left elbow. The patient did not recall how long the lesion had been present; however, she states it was not present during childhood. The lesion had no associated enlargement, pain, or hyperhidrosis. Because the patient had a history of melanoma-in-situ involving the same extremity, the nodule was elliptically excised and sent for histologic examination to rule out a primary malignant melanoma, a metastatic malignant melanoma, or a hematoma.

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#### Figures & Images

- 1. Click on the Figure number you wish to review.*
- 2. Click on the image to enlarge*

- **Histologic examination:**

The epidermis was unremarkable [Figures 1,2]. In the mid-to-deep reticular dermis, there was a nodular proliferation of capillaries surrounded by an increased number of mature eccrine glands [Figures 3, 4] . These vessels were noted to be either dilated or collapsed due to the enlarged eccrine glands [Figures 5,6]. The lesion was well demarcated but not encapsulated.

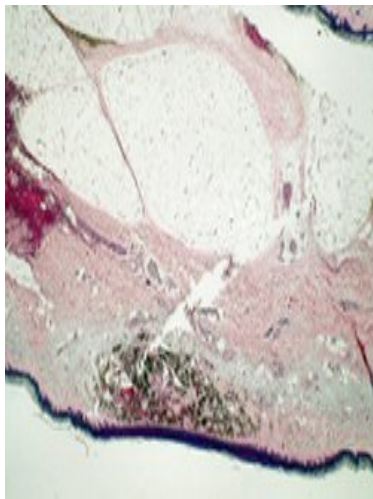


Figure 1

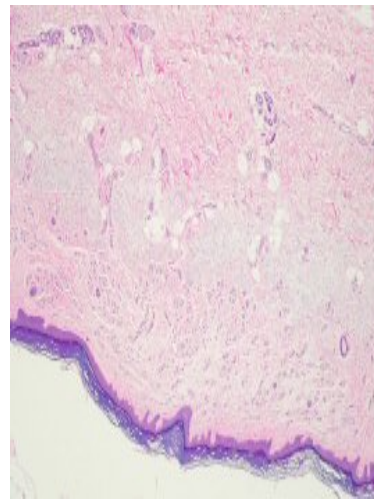


Figure 2

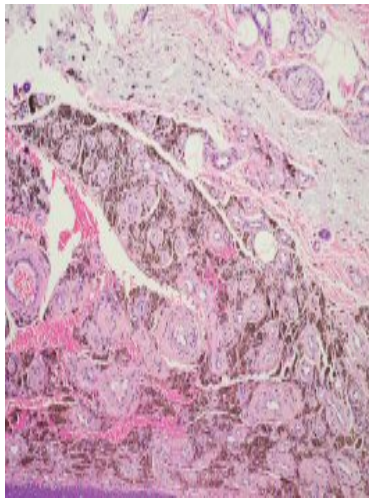


Figure 3

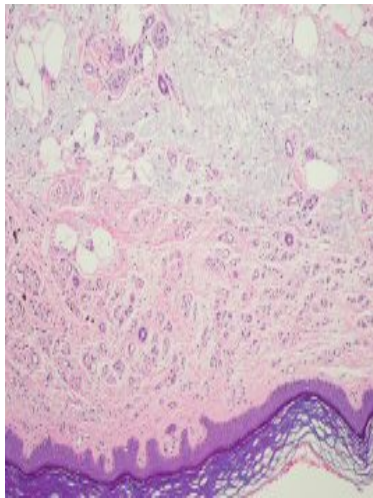


Figure 4

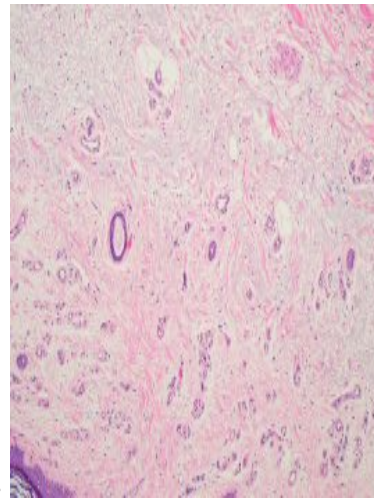


Figure 5

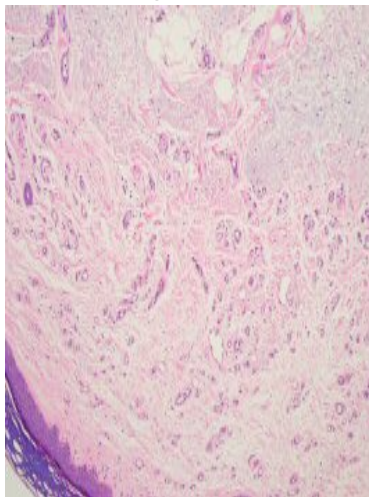


Figure 6

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**Immunohistochemistry:**

• Vascular immunohistochemistry:

The nodular proliferation of capillaries stained positively for CD31 [Figures 7,8].



Figure 7



Figure 8

• **Eccrine ductal immunocytochemistry:**

The secretory component of the eccrine ducts stained positively for S100 [Figures 9, 10, 11].

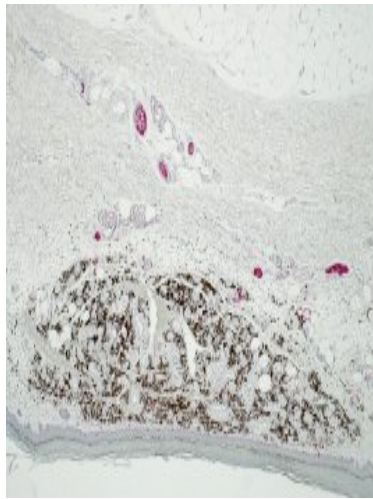


Figure 9

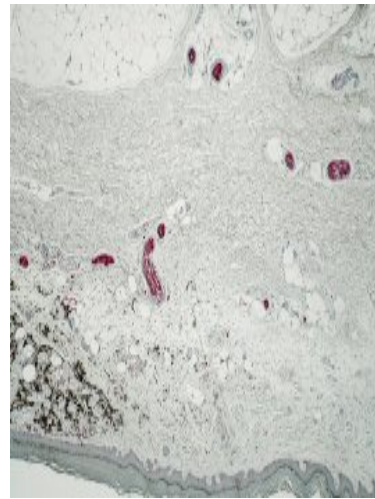


Figure 10

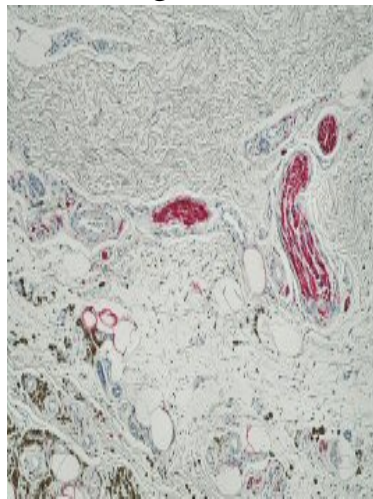


Figure 11

The ductal components of the eccrine ducts stained positively for cytokeratin 1 (high molecular weight cytokeratin) [Figure 12].

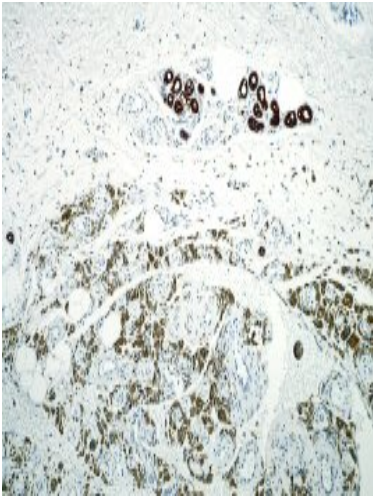


Figure 12

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### OCTOBER 2003 CASE OF THE MONTH

#### DISCUSSION & DIAGNOSIS

##### Diagnosis:

***Eccrine angiomatous hamartoma***

##### Discussion:

Eccrine angiomatous hamartoma (EAH) is a benign cutaneous hamartoma. It is characterized by the proliferation of eccrine and capillary structures within the dermis. Lotzbeck first described this entity in 1859 as an angiomatous lesion on the cheek of a child. Since its first description, there have been approximately 37 cases reported in the literature.

Typically, EAH occurs in children or young adults; however, congenital and adult-onset cases have been reported.<sup>2</sup> The oldest reported case of EAH occurred in a 60-year-old woman. EAH usually occurs as a solitary lesion, but multiple lesions have been described in several patients.<sup>2</sup> Characteristically, the nodule may be flesh-colored, blue-brown, or red. Lesions tend to occur most frequently on the extremities, but lesions have been reported on the trunk, face, and diffusely over multiple areas. There is no gender predilection. EAH is usually asymptomatic, but the most commonly associated symptoms include pain, hyperhidrosis, and hypertrichosis. Although these lesions may grow, they grow in proportion to the growth of the patient.

The diagnosis of EAH is made histologically, since the clinical picture can be variable. Histologic findings include a dermal proliferation of enlarged, mature eccrine glands in close association with dilated and collapsed blood vessels. These vessels are typically thin-walled or capillary-in-nature and exist as either ill-defined angiomatous foci or aggregated into capillary lobular proliferations. Histologic variants have been described and these include the infiltration of lipomatous tissue,<sup>4-6</sup> the presence of

pilar structures, 7-9 and dermal mucin.6, 10 Typically, the epidermis is normal<sup>2, 8</sup> but it may be hyperkeratotic, <sup>11</sup> papillomatous, <sup>3,7</sup> or acanthotic.<sup>11</sup> Immunohistochemical studies demonstrate that the secretory portions of the eccrine glands within EAH are positive for carcinoembryonic antigen, S100, epithelial membrane antigen, and Cam5.2. The ductal portion of the eccrine gland stained positively with carcinoembryonic antigen and cytokeratin 1.<sup>12</sup> In addition, the endothelial cells expressed anti-*Ulex europaeus* and anti-Factor VIII antigens.<sup>5, 12</sup> Mitotic figures and cytologic atypia have not been demonstrated in lesions of EAH.<sup>4, 9</sup>

The etiology of EAH is unknown. Hamartoma formation is believed to be due to an abnormal induction of heterotypic dependency during early organogenesis. Altered chemical interactions between differentiating epithelium and mesenchyme leads to hamartoma growth, producing an abnormal proliferation of vascular and eccrine structures.<sup>7</sup>

No treatment is needed for EAH. Excision is curative and can be performed for pain palliation or for cosmetic purposes. Recurrence of the primary lesion has not been documented.<sup>13</sup>

The clinical differential diagnosis of EAH includes eccrine nevus, blue-rubber bleb syndrome, tufted angioma, glomus tumor, smooth muscle hamartoma, and macular telangiectatic mastocytosis. Eccrine nevus is distinguished from EAH by the generation of localized hyperhidrosis clinically and the lack of capillary proliferation, histologically. Sudoriparous angioma is a histologic entity, which is very similar to EAH. In lesions of sudoriparous angioma, the angiomatous features are more prominent and the vessels are predominantly large-caliber. <sup>14,15</sup> In addition, the eccrine glands are dilated but not hyperplastic in sudoriparous angioma. <sup>14</sup> The above remaining lesions in the differential are easily distinguished histologically.

## **SUMMARY:**

We present an interesting case of eccrine angiomatous hamartoma, which was incidentally found. To date, this patient has the latest age of presentation of EAH. Eccrine angiomatous hamartoma is a benign proliferation of eccrine glands and capillary vessels. It tends to be a painful lesion on the extremity and may be associated with hyperhidrosis. Histopathology is essential for diagnosis. Excision is curative and generally indicated in painful lesions or for cosmesis.

## **References:**

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