Cutaneous Angiolipomatous Hamartoma

Nima Amini, MD; Syed Abedi, MD; Silvija P. Gottesman, MD; Thinh Chau, BS; Jacqueline M Junkins-Hopkins,MD

Departments of Pathology & Laboratory Medicine and Dermatology, University of California Davis, Sacramento, CA

Ackerman Academy of Dermatopathology, New York, NY

Introduction

Cutaneous vascular anomalies are classified as follows: hamartomas, malformations, dilatations of pre-existing vessels, hyperplasias, or neoplasms (benign or malignant). Hamartoma is defined as a disorganized arrangement of tissues indigenous to an organ. Cutaneous vascular hamartomas are rare, and include phakomatosis pigmentovascularis (co-existence of capillary malformation and melanocytic lesion) and eccrine angiomatous hamartoma (EAH), which is a dermal proliferation of eccrine glands and thin-walled blood vessels, usually noted at birth or during infancy. PTEN hamartoma of soft tissue (PHOST) is a distinctive lesion, usually intramuscular or subcuticular, in patients with PTEN hamartomatous tumor syndrome (PHTS). PHTS is characterized by hamartomatous elements of adipose tissue and vessels, predominantly with a fibromyxoid stroma and occasionally other elements (lymphoid follicles, neural tissue, and bone). We report a case of a hamartomatous-appearing vascular lesion, which does not fulfill histologic criteria for the aforementioned entities.

Case History

A 70-year-old man presented with a 2-year history of a 1.5cm cystic lesion on the left temple. Histologically, there were variably dilated thick-walled blood vessels intimately associated with smaller vessels and adipocytes in the dermis and subcutis, with entrapment of anatomic eccrine glands. (Figure 1, 4) Immunostain against CD31 and SMA highlighted well-formed vessels; smooth muscle bundles were absent. No distinct spindle cell component was highlighted by SMA staining. Scattered eccrine glands were seen.

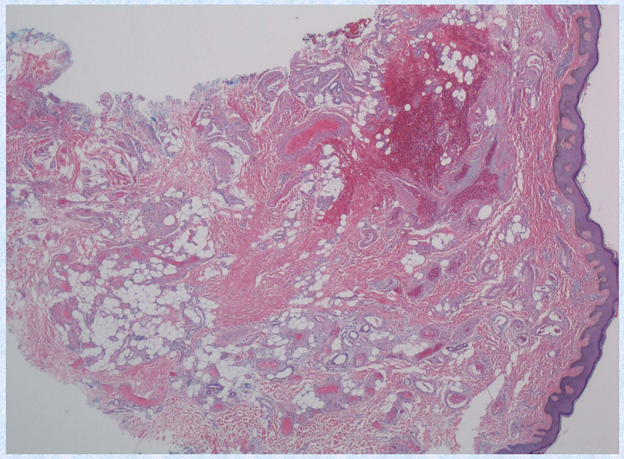

Figure 1. Histopathology of the left temple lesion on H&E stain (20X)

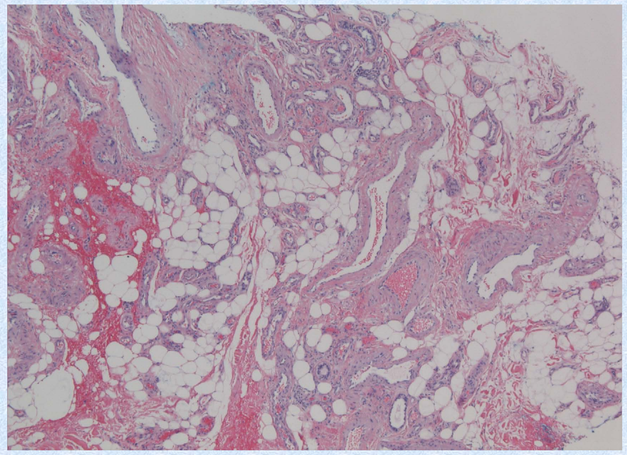

Figure 2. Overlapping histologic features of a hamartoma and a vascular neoplasm were observed on H&E stain (40X)

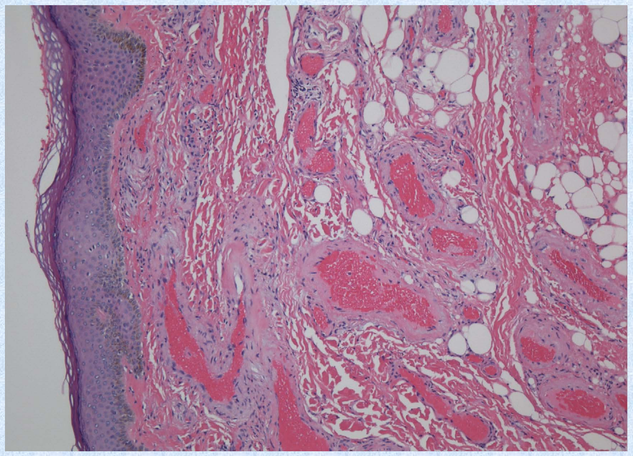

Figure 3. Overlapping histologic features of a hamartoma and a vascular neoplasm were observed on H&E stain (100X)

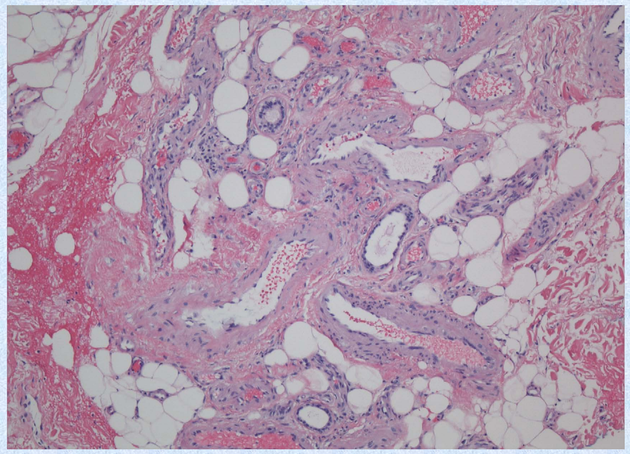

Figure 4. Histopathology of the lesion with vessels and adipose tissue on H&E stain (100X)

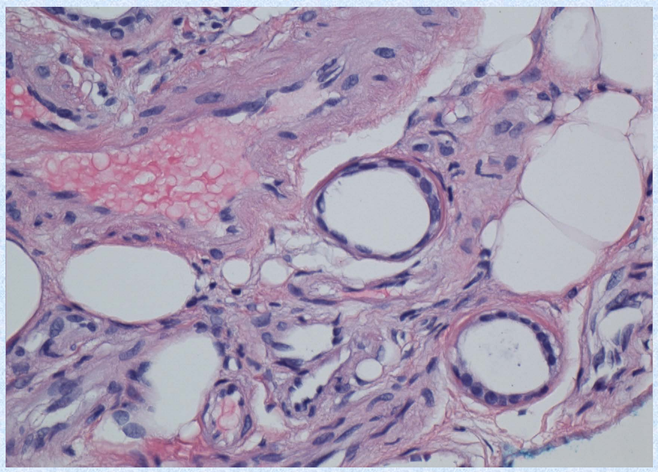

Figure 5. Entrapped eccrine glands (200X)

However, only anatomic structures were immunoreactive to AE1/AE3. A benign hamartomatous vascular neoplasm composed of blood vessels and adipose tissue was ultimately favored.

Discussion

The main challenge of this unique case is that certain histologic features fit those of a hamartomatous vascular entity whereas others resemble those of a vascular neoplasm. Hamartomas encompass several combined vascular and melanocytic proliferations, grouped as phakomatosis pigmentovascularis and so-called eccrine angiomatous hamartoma which exhibits both eccrine and vascular proliferations.

Eccrine angiomatous hamartoma (EAH), a rare cutaneous hamartoma, share similar histologic features to our case. However, our lesion is distinct based on late clinical onset, absent eccrine proliferation and smooth muscle bundles, and prominent subcutaneous extension. Multiple lesions are typical in patients with EAH whereas our patient presented with a solitary lesion.

Although our case bears histologic resemblance to PHOST, a fibromyxoid stroma was absent in our case. The occasional lymphoid follicles, neural tissue and bone were not also observed.

Immunohistochemical studies were crucial to distinguishing this new entity from previously discussed conditions. AE1/AE3 highlighted a pattern favoring entrapped anatomic eccrine glands in contrast to eccrine proliferation, a characteristic feature of EAH.

References

1. Requena L, Sangueza OP. Cutaneous vascular anomalies. Part I. Hamartomas, malformations, and dilation of preexisting vessels. Journal of American Academy of Dermatology. 1997 October;37(4):523-49; quiz 549-52.

Contact info: nimamini@ucdavis.edu