ENT PATHOLOGY CLINIC

Angiolymphoid hyperplasia with eosinophilia

Lester D.R. Thompson, MD

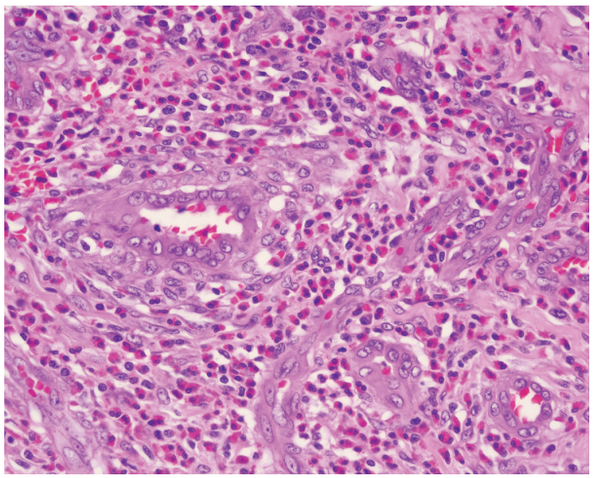

Figure 1. Multiple lobules composed of immature capillaries show enlarged, epithelioid endothelial cells. Note the remarkable number of eosinophils

Angiolymphoid hyperplasia with eosinophilia (ALHE), sometimes called *epithelioid hemangioma,* is a benign vascular tumor. It features immature blood vessels lined by epithelioid endothelial cells with a prominent inflammatory infiltrate, frequently showing a conspicuous eosinophil component. There is controversy about whether this lesion is a reactive or benign neoplastic condition.

Patients present over a wide range of ages, with a peak in the third to fifth decades, and with females affected more often than males. The head is most commonly affected (scalp and ears), with the digits involved second most frequently. There is usually a subcutaneous nodule/mass that may be painful and/or pruritic, with pink to red-brown, dome-shaped papules or nodules, which may coalesce. This lesion *does not* involve lymph nodes (i.e., not Kimura disease). While peripheral serum eosinophilia may be seen, IgE levels are not elevated.

There is an excellent prognosis with excision, but recurrences or persistence after surgery is common, requiring close clinical follow-up.

In general, the lesions are small and may resemble lymph nodes because of circumscription and peripheral inflammation. The surface epithelium is usually intact, although excoriation may be seen. There are multiple lobules composed of immature capillary to medium-sized vessels, usually without well-developed lumina (figure 1). Larger vessels may be seen. Occasionally, the lesions are solid. The endothelial cells are enlarged and appear epithelioid or histiocytic (figure 2). Cytoplasmic vacuolization may be present. The vascular proliferation is invested by a rich inflammatory infiltrate, including lymphocytes, mast cells,

From the Department of Pathology, Southern California Permanente Medical Group, Woodland Hills Medical Center, Woodland Hills, Calif.

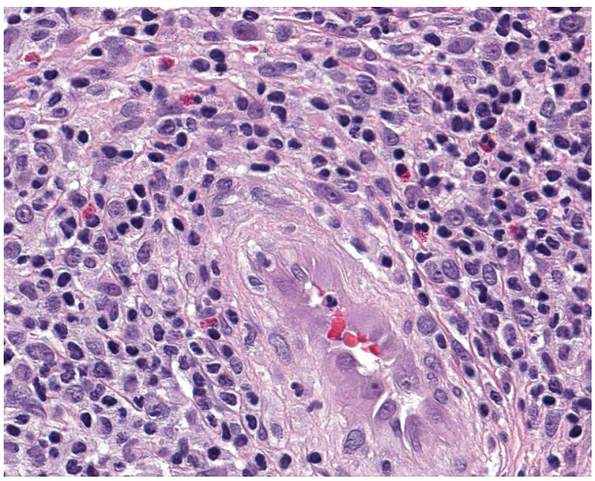

Figure 2. This high-power image shows a central vessel with very high epithelioid endothelial cells lining the space. The surrounding lymphoid infiltrate shows scattered eosinophils

and eosinophils, the latter varying in density within and between lesions (figures 1 and 2). Uncommonly, germinal centers may be sparse and poorly formed.

The differential diagnosis includes Masson vegetant or papillary endothelial hyperplasia (a reactive endothelial proliferation associated with organizing thrombus/clot); Kimura disease (lymph node disorder, most often in Asian men, who have peripheral serum eosinophilia, reactive lymphoid follicles, follicular lysis, eosinophilic microabscesses, polykaryocytes, and IgE deposition); and angiosarcoma (freely anastomosing vessels, atypical endothelial cells, increased mitoses, necrosis).

Suggested reading

Chen H, Thompson LD, Aguilera NS, Abbondanzo SL. Kimura disease: A clinicopathologic study of 21 cases. Am J Surg Pathol 2004;28(4):505-13.

Sun ZJ, Zhang L, Zhang WF, et al. Epithelioid hemangioma in the oral mucosa: A clinicopathological study of seven cases and review of the literature. Oral Oncol 2006;42(5):441-7.

Tosios K, Koutlas IG, Papanicolaou SI. Intravascular papillary endothelial hyperplasia of the oral soft tissues: Report of 18 cases and review of the literature. J Oral Maxillofac Surg 1994;52(12):1263-8.

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occurs more frequently in women (female-to-male ratio: 1.2:1) between the fifth and sixth decades of life, with the most common complaint being that of a growing mass with local pain and facial paralysis.[4]

Optimal imaging studies for perineural invasion can be obtained with magnetic resonance imaging (MRI), which demonstrates abnormal enhancement of the cranial nerves involved, extending into the base of the skull and neural foraminal erosions.[4]

Histologically, ACC is divided into three types: *cribiform, tubular,* and *solid*.[1] The *solid* type is associated with distant metastasis to lungs, bone, viscera and brain.[5] In most cases there is an incidence of about 60% of lung involvement as the first manifestation of delayed distant metastasis.[5,6] Lung metastases secondary to ACC may be asymptomatic longer than metastases from other primary carcinomas. The average time from detection of lung metastases until death is 32.3 months.[6] Tumor size of more than 3 cm is generally another highly predictive factor for distant metastasis.[5]

The most common biological targets in ACC, in descending order of expression, are: proto-oncogene c-kit (78 to 92%), epidermal growth factor receptor (36 to 85%), and human epidermal growth factor receptor 2 (2 to 36%).[6] Clinically in ACC, lymphatic spread to the neck plays a minor role in distant metastasis and is considered a delay manifestation, occurring within 5 and 10 years after treatment.[1]

Overall, the recurrence-free survival rates for ACC are 65% at 5 years, 52% at 10 years, and 30% at 15 years.[7] Unfortunately, aggressive therapy for the primary tumor in the presence of metastatic disease, including radiotherapy and neoadjuvant chemotherapy, does not influence survival.[1]

References

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3. Bradley PJ. Adenoid cystic carcinoma of the head and neck: A review. Curr Opin Otolaryngol Head Neck Surg 2004;12(2):127-32.
4. Ayadi K, Ayadi L, Daoud E, et al. Adenoid cystic carcinoma of the parotid with facial nerve invasion. Tunis Med 2010;88(1):46-8.
5. Schwentner I, Obrist P, Thumfart W, Sprinzl G. Distant metastasis of parotid gland tumors. Acta Otolaryngol 2006;126(4):340-5.
6. Guzzo M, Locati LD, Prott FJ, et al. Major and minor salivary gland tumors. Crit Rev Oncol Hematol 2010;74(2):134–48.
7. Khan AJ, DiGiovanna MP, Ross DA, et al. Adenoid cystic carcinoma: A retrospective clinical review. Int J Cancer 2001;96(3):149-58.