Lymphangioma

Lester D.R. Thompson, MD

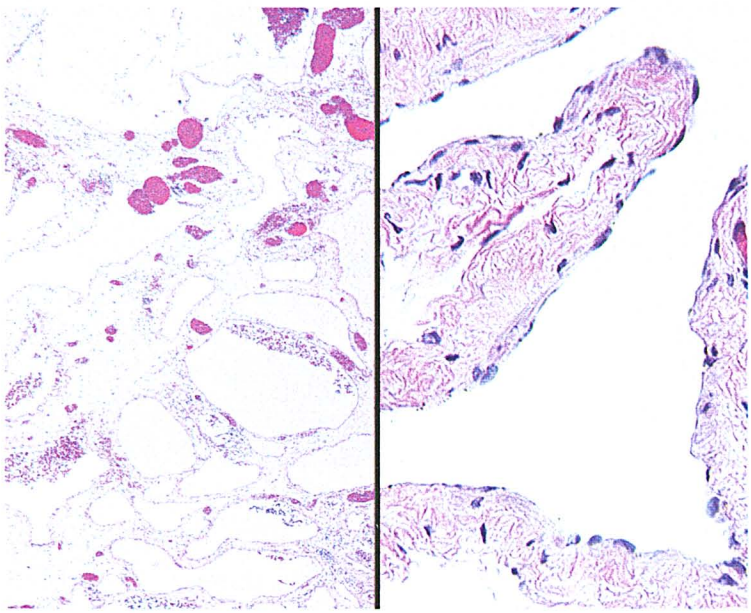

Figure 1. Patulous, dilated vascular channels are filled with proteinaceous fluid. The endothelial cells lining the cystic spaces are bland.

Lymphangiomas are rare congenital tumors, with up to 70% reported in the head and neck. They are separated into three types: *cystic* (cystic hygroma), *capillary*, and *cavernous*. Lymphangiomas account for approximately 25% of all vascular neoplasms in children and adolescents. About 25% of cervical cysts are lymphangiomas.

Roughly two-thirds of all lymphangiomas are noted shortly after birth, and 95% are present by the end of the second year of life. Cystic hygroma may also be detected in utero by ultrasonography. Cystic hygroma is commonly associated with fetal hydrops and Turner syndrome. In general, symptoms relate to pressure caused by the enlarging mass in the posterior neck, although the tumor may extend into the anterior compartment, upward into the cheek, or down into the mediastinum or axilla. When located superior to the hyoid bone, the tumor may cause dysphagia or airway compression. The appearance of cystic hygromas varies from a single soft mass with a pseudocontour to lobulated multicystic masses.

Lymphangiomas (specifically, the cystic type) are made up of dilated thin-walled spaces that are filled with eosinophilic proteinaceous fluid and lined with flat endothelial cells (figure 1). The intervening stroma contains scattered lymphoid aggregates and wisps of smooth muscle fibers (figure 2). Fibrosis may be increased in lesions that have been present for a long duration. Endothelial markers (e.g., factor VIII-RAg, CD31, and CD34) will be positive. The most common differential diagnosis is cavernous hemangioma, which is filled with red blood cells and lacks valve structures. Metastatic papillary carcinoma of the

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

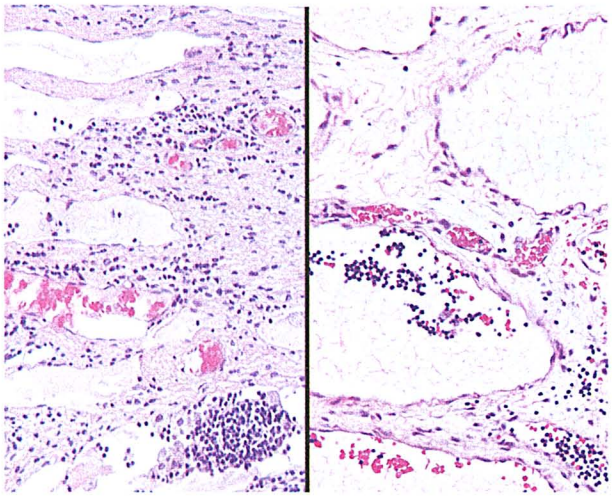

Figure 2. A variable amount of smooth muscle, fibrosis, and lymphocytes can be present between the vascular channels. Note the proteinaceous fluid within the cystic spaces. The small veins contain erythrocytes.

thyroid may exhibit flattened cells along the spaces, but TTF-1 or thyroglobulin will be positive and a lymph node architecture should be seen.

Recurrence rates range from 15 to 50%. Mortality rates are between 3 and 7%, specifically related to pressure destruction of vital structures of the neck. Lymphangiomas may occasionally become infected. Surgery is the treatment of choice, while laser treatment and injected sclerosing agents are alternate therapies.

Suggested reading

Coffin CM, Dehner LP. Vascular tumors in children and adolescents: A clinicopathologic study of 228 tumors in 222 patients. Pathol Annu 1993;28:97-120.

Weiss SW, Goldblum JR. Tumors of lymph vessels. In: Weiss SW, Goldblum JR, eds. Enzinger and Weiss's Soft Tissue Tumors. 4th ed. St Louis: Mosby, 2001:955-83.