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Lymphatic malformations: Diagnosis and management

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ABSTRACT

Lymphatic malformations are benign vascular lesions that arise from embryological disturbances in the development of the lymphatic system. They encompass a wide spectrum of related abnormalities, including cystic lymphatic lesions, angiokeratoma, lymphatic malformations that occur in bones (Gorham–Stout Syndrome), lymphatic and chylous leak conditions, and lymphedema. This article will focus only on lymphatic malformation mass lesions, whereas other related disease entities will be covered elsewhere in this journal issue. Lymphatic malformations occur frequently in lymphatic-rich areas such as the head and neck region, but they can also be found on any anatomical site in the body. In general, lymphatic malformations are categorized into macrocystic, microcystic, or combined depending on the size of the cysts contained within the lesion. Lymphatic malformations can cause both deformation of the anatomical site involved and functional deficits. The goal of this article is to discuss the etiology, epidemiology, treatment modalities, and comorbidities associated with lymphatic malformations.

Lymphatic malformations are benign vascular lesions that arise from embryological disturbances in the development of the lymphatic system. They encompass a wide spectrum of related abnormalities, including cystic lymphatic lesions, angiokeratoma, destructive lymphatic malformations that occur in bones (Gorham–Stout Syndrome), lymphatic and chylous leak conditions, and lymphedema. This article will focus only on lymphatic malformation mass lesions, whereas other related disease entities will be covered elsewhere in this journal issue. Symptoms of lymphatic malformations are related to the anatomical location of these lesions, as well as to the extent of involvement of the local anatomical structures.

Lymphatic malformations can be seen in any anatomic region but are more commonly seen in lymphatic-rich areas, such as the head and neck (45–52%), axilla, mediastinum, groin, and retroperitoneum.[1,2] These malformations are thought to be the result of abnormal development of the embryonic lymphatics or lymphatic jugular sacs, with failure of these structures to connect or drain into the venous system. In some patients, ectatic adjacent veins can be seen in association with a cystic lymphatic lesion. There are three morphologic types of cystic lymphatic lesions: macrocystic, microcystic, and combined. Macrocystic lesions are large, compressible or non-compressible, smooth, and translucent masses under normal or bluish skin. Most macrocystic lesions are multilocular structures consisting of numerous cysts that vary in size (Figure 1). Macrocystic lesions are often located below the level of the mylohyoid muscle and involve the anterior and posterior cervical triangles.[3] Microcystic malformations present as clear, tiny vesicles that permeate the subcutaneous tissue and muscles (Figure 1). Microcystic lesions are commonly found above the level of the mylohyoid muscle and involve the oral cavity, oropharynx, tongue, parotid gland, submandibular gland, and pre-epiglottic space. These vesicles are often firm and may give the impression of a brawny edema. Combined lesions are more common below the head and neck and contain a mixture of micro- and macro-cysts.

Histologically, cystic lymphatic malformations are composed of vascular spaces filled with eosinophilic and protein-rich fluid (Figure 2). A single layer of flattened endothelium lines the walls of lymphatic channels. The fibrovascular vessel walls are of variable thickness, with abnormally formed smooth muscle. Collections of lymphocytes are common throughout the contained connective tissue. Hemorrhage within the cystic spaces is common, indicating recent trauma or spontaneous intralesional bleeding.

Large cystic lymphatic lesions can be diagnosed *in utero* using ultrasound as early as the beginning of the second trimester,[4,5] but lesions are more commonly noted at birth; the vast majority are evident by 2 years of age. Unlike hemangiomas, lymphatic malformations persist throughout life, grow proportionately with the size of the patient, and do not undergo involution as does

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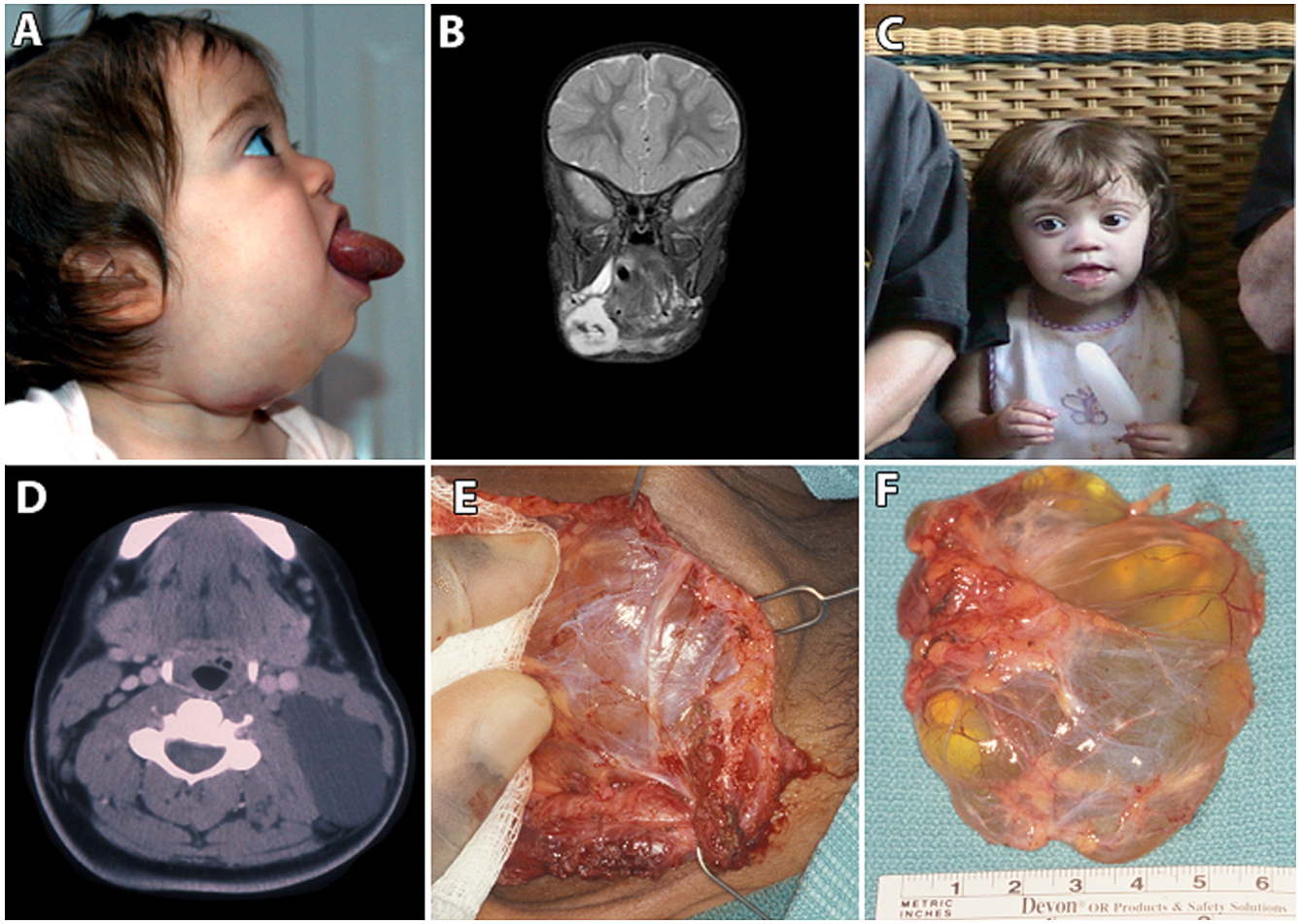

Fig. 1. (A) A photograph and (B) an MRI of a 2-year-old girl with a large cervical–facial predominantly microcystic lymphatic malformation. The lymphatic malformation occupies the right submandibular gland, floor of mouth, and tongue. (C) The same patient after surgical excision of the tongue and neck lymphatic malformation. (D) An MRI of a 15-year-old girl with a left neck focal macrocystic lymphatic malformation. (E and F) Appearance of the macrocystic lymphatic malformation intraoperatively and after excision. Notice the large cystic cavities filled with a yellow clear fluid. (Color version of figure is available online.)

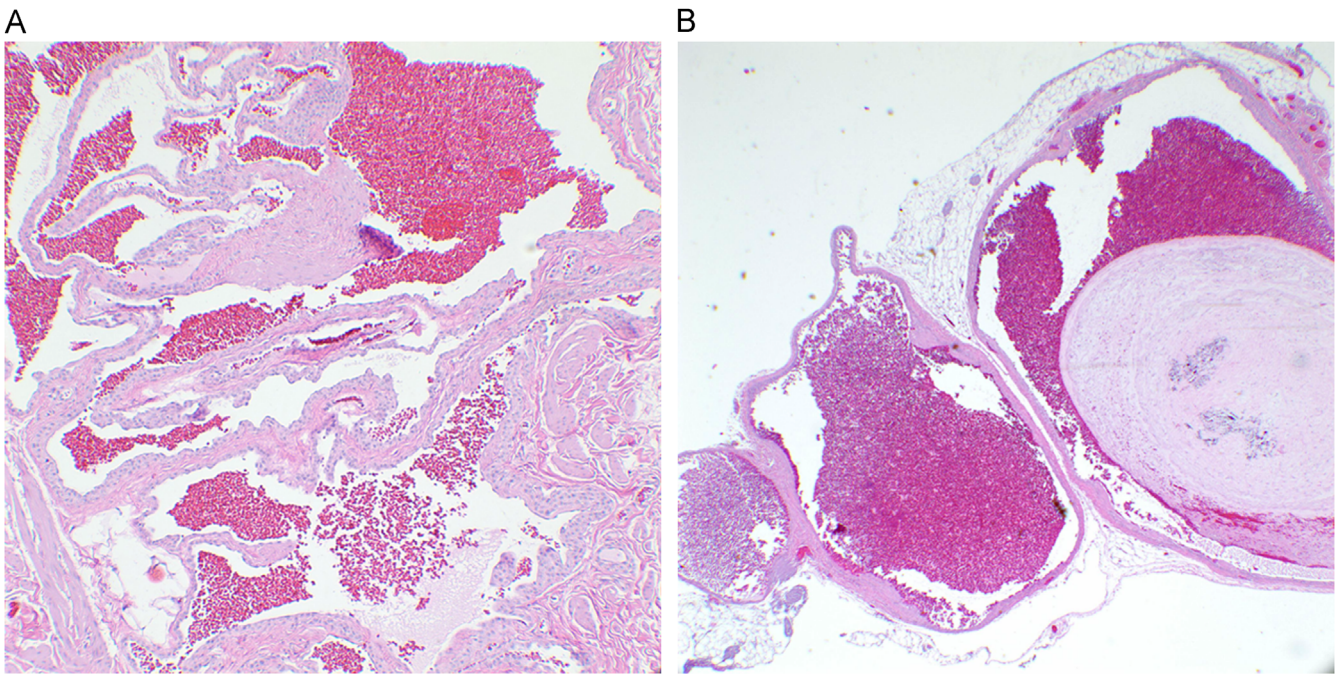

Fig. 2. Photomicrographs of a lymphatic malformation. (A) Lymphatic malformations are composed of vascular channels of varying sizes interspersed with lobules of fibroadipose tissue (Hematoxylin & Eosin; original magnification, 40 ×). (B) Small- to medium-sized channels lined by flat lymphothelium and patchy smooth muscle. The lumina are filled with pink, proteinaceous material and lymphocytes (Hematoxylin & Eosin; original magnification, 400 ×). (Color version of figure is available online.)