Cervical Lymphangioma in Adult

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ABSTRACT

Lymphangiomas are uncommon congenital lesions of the lymphatic system which are usually present in childhood. We report a case of adult lymphangioma, localized in the neck, and discuss the presentation, diagnosis and management of this tumor.

Keywords: Head and neck, Lymphangioma, Children, Diagnosis, Treatment.

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INTRODUCTION

Lymphangioma is an uncommon benign pathology, usually reported in children and rarely in adults. Cervical lymphangioma involves congenital and cystic abnormalities derived from lymphatic vessels with a progressive and painless growth. Imaging techniques can aid in the precise mapping of the lesion and in defining its boundaries with the surrounding vital structures, improving therapeutic success. Various therapeutic options are reported in literature, but complete surgical excision is still considered the best approach. Complete removal is more easily achieved in adults than in children.

CASE REPORT

An 18-year-old male presented to our hospital with a slow-growing right neck mass since 2 years. Upon presenting to our hospital, the patient noted only slight discomfort in his neck, which occurred with movement. He had no history of trauma, shortness of breath, dysphagia, dysphonia or pain. On physical examination, the mass was soft and fluctuating. It extended from below the angle of the mandible to the supraclavicular region by tracking along the anterior border of the sternocleidomastoid muscle and anteriorly upto thyroid lamina (Fig. 1). Oral cavity was normal. Magnetic resonance imaging (MRI) of the neck axial and coronal sections revealed a right-sided submandibular cystic lesion in subcutaneous plane extending medially from retromandibular region and abuts and insulates the right vallecula with its obliteration. It also extended anteriorly over the thyroid lamina in subcutaneous plane with its displacement. Inferiorly, it extended upto C6-C7 vertebral level and superiorly upto inferior margin of parotid gland.

There was no enhancing portion, and the lesion measured approximately 6 × 4 × 3 cm. It was profoundly hyperintense on T2W1 images (Fig. 2). It showed multiple incomplete thin septi with communicating loculi.

Surgery

The mass was reached through a 7 cm transverse skin incision in the submandibular region. Cystic mass was seen in subcutaneous plane. Mass was dissected from tail of parotid laterally then from behind posterior belly of digastric muscle till submandibular gland medially to vallecula in superomedial plane. It was touching internal jugular vein (IJV) and common carotid artery posteriorly. Cystic mass was draining into the IJV via small vessel (Fig. 3). That vessel was ligated near the IJV. Although the lesion was in close proximity to nerves, vessels and muscles, there was no...
invasion into these structures, and 6 × 4 × 3 cm mass was circumferentially dissected and removed in its entirety (Fig. 4). Histopathology revealed a lymphangioma composed of large irregular lymphatic channels lined by bland endothelial lining filled with eosinophilic proteinaceous fluid, i.e. lymph (Fig. 5).

**DISCUSSION**

Lymphangiomas are rare congenital benign lesions occurring mainly in the head, neck and oral cavity. Three theories have been proposed to explain the origin of this abnormality. (1) Blockage or arrest of normal growth of the primitive lymph channels occurs during embryogenesis. (2) Primitive lymphatic sac does not reach the venous system. (3) During embryogenesis, lymphatic tissue lays in the wrong area.

A commonly used classification classifies these lesions into capillary lymphangioma or lymphangioma simplex, cavernous lymphangioma and cystic lymphangioma or cystic hygroma. When a lymphangioma is confined to fairly dense tissue, such as the tongue, it presents as a cavernous lymphangioma, but when it develops in the relatively loose fascia of the neck, a cystic lesion occurs. These three types are frequently found together in a same patient, depending on the severity of the disease. Cystic hygromas, however, account for approximately 90% of the lymphangiomas in the head and neck region. Other common sites, outside the head and neck, include the axilla, shoulder, chest wall, mediastinum, abdominal wall and thigh lymphangiomas of the head and neck are sometimes referred to as cystic hygromas and result from abnormal growth of the lymphatic vessels. Lymphangiomas are most commonly found in children and rarely observed in adults. Most lymphangiomas are congenital, but they have also been reported to arise from infections, neoplasms, trauma and iatrogenic injuries.

Lymphangiomas are benign lymphatic malformations that occur most commonly in children. Some authors refer to these lesions as cystic hygromas, a term first used by Wernher in 1843. The term cystic hygroma refers to the cystic nature of these lesions and their mass effect; hygroma means ‘moist tumor’. Lymphangioma, however, is the more appropriate nomenclature because it denotes the tissue of origin. Lymphangiomas are thought to arise from an embryologic defect that consists of an abnormal connection from the jugular sacs to the peripheral lymphatic system.

Lymphangiomas are relatively rare congenital lesions that result from abnormal growth of the lymphatic vessels. It is rare for lymphangiomas to make their initial presentation during adulthood, and fewer than 100 such cases have been reported. Approximately, 60% of lymphangiomas are present at birth, and up to 90% are
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detectable by 2 years of age.\(^5\) Although more than 90% of lymphatic malformations are clearly congenital, later presentations may occur as a result of trauma, infection, neoplasms or iatrogenic injuries. The pathophysiology of adult lymphangiomas is not clearly understood, but these lesions may occur secondary to induction of dormant rests of embryonic lymphatic tissue that are stimulated to differentiate and grow. Lymphangiomaticous lesions are rare congenital malformations of the lymphatic system that occur throughout the body with greater frequency in the cervicofacial area. Cystic lymphangiomas have been reported, in the literature, to be present in up to 67% of all cystic lymphangiomas.\(^6\) Introduction of prenatal USG allows detection of disease \textit{in utero}, in approximately the same percentage. However, almost all are detected before the age of 2 years. Histologically, these lesions are composed of dilated lymphatic channels with one or two endothelial layers with or without an adventitial layer. These dilated lymphatics can vary in size.

The incidence of lymphangiomas has been reported to range from 1.2 to 2.8 per 1,000 newborns. They have no gender predilection and present as a painless mass that progressively enlarges. The most prominent sign or symptom of all lymphangiomas is the presence of a mass. Typically, the mass is soft, nontender and ill-defined. The mass may be small and unnoticed at birth only to present later with an upper respiratory tract disorder or incidental trauma at the site. Most lesions, however, are recognized early on account of their size and associated symptoms of respiratory obstruction and problems with feeding, which are the second and third most common presenting symptoms. Difficulty in swallowing results from lymphangiomas extending to involve the oral cavity, oropharynx and/or the hypopharynx. Isolated tongue involvement can lead to macroglossia with dysphagia and airway obstruction. In adult patients, neoplasm can switch to squamous cell carcinoma.\(^7\) Cervical lymphangiomas have not been shown to lateralize to any particular side of the neck. In adults, the main complaint is cosmetic unacceptability of mass.

Ultrasoundography and computed tomographic (CT) scanning have been used extensively to evaluate the anatomy of lymphangiomas. On ultrasound examinations, these lymphatic malformations appear as thin-walled, multiseptate, multicystic, hypoechoic masses. Lymphangiomas appear as multiloculated cystic lesions on CT scanning. On T2-weighted MRI, lymphangiomas appear isointense to cerebrospinal fluid, whereas their intensity varies on T1-weighted images due to variable protein content.\(^8\)

Observation is frequently the first step for small, nonexpanding lesions. Those that persist continue to grow, or present with obstructive symptoms should be resected. To avoid injuring adjacent neurovascular structures, surgical planning is crucial before resection. Some surgeons prefer to use MRI to facilitate resection because it allows improved anatomical demarcation. Ultrasonography has limited ability to delineate the anatomical planes of resection or the extent of structural involvement.

Complications of resection include infection, bleeding, hematoma and postoperative seromas. Injury to facial, hypoglossal, glossopharyngeal, recurrent laryngeal and lingual nerves has been reported.

Complete excision of a cystic hygroma has been shown to have an 81% cure rate. When only part of the lymphatic malformation is excised, there is an 88% recurrence rate. Several studies have shown an increase in rates of recurrence, morbidity and complications for lymphangiomas located in the suprahyoid versus infrahyoid region.

Sclerosing agents and radiation therapy have not been shown to play a role in the primary treatment of cystic hygromas. They may be indicated in lymphatic malformations that are macrocystic and not amenable to surgical resection.\(^9\) Sclerotherapy with OK-432, a lyophilized, low-virulence SU strain of group A \textit{Streptococcus pyogenes}, has been suggested as a possible therapy for macrocystic lesions. To date, however, the efficacy of OK-432 has not been proven in prospective, controlled trials.\(^10\)

**CONCLUSION**

Lymphangioma of the head and neck are benign neoplasms which are easy to diagnose. Surgical intervention represents the treatment of choice. Lesion extension and involvement of vital structures can reduce, in some cases, the possibility of complete resection.

**REFERENCES**


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