Annals of Otology, Rhinology & Laryngology 117(11):824-826.

Cystic Lymphatic Malformation of the Middle Ear

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Objectives: We review the clinical, radiologic, and histopathologic features of cystic lymphangioma of the middle ear, and discuss the developmental etiology and management of such a lesion.

Methods: We present an unusual location for the development of a cystic lymphangioma with emphasis on etiology, clinical implications, and current treatment.

Results: A 10-year-old girl presented with a mass involving the medial surface of the right tympanic membrane. T2-weighted magnetic resonance imaging demonstrated a hyperintense lesion in the anterior-superior middle ear cavity without evidence of vascular abnormalities.

Conclusions: To our knowledge, this is the only report of lymphangioma involving the middle ear represented in the English-language literature. Such a lesion has been demonstrated to arise from abnormalities in growth factors that contribute to the tightly regulated process of lymphangiogenesis. Lymphatic malformations can be diagnosed presumptively by virtue of magnetic resonance imaging in combination with a detailed physical examination. The treatment of choice for lymphangiomas located in the middle ear is surgical excision. Definitive diagnosis of the lesion is then made by identifying specific histopathologic characteristics. Although rare and histologically benign, middle ear lymphangiomas may produce significant patient discomfort and ultimately a conductive hearing loss. Therefore, these lesions warrant early recognition and treatment.

Key Words: lymphangioma, lymphatic malformation, middle ear.

INTRODUCTION

Often referred to as lymphangiomas, lymphatic malformations are benign, typically congenital malformations of the lymphatic system. The majority of cases are detected before the third year of life, and they primarily involve the head, neck, and axilla. A small percentage of lesions remain undetected beyond the third year of life and come to medical attention only when symptoms develop. We recently encountered a lymphatic malformation involving the middle ear cavity and a portion of the tympanic membrane in a pediatric patient. To the best of our knowledge, this is the first reported case in the English-language literature. The clinical, radiologic, and histopathologic features of this lesion are presented.

CASE REPORT

A 10-year-old girl presented with a 10-month history of slowly progressive right ear discomfort, tinnitus, and aural fullness. She denied hearing loss, vertigo, otorrhea, or recurrent otitis media. On physical examination, the right ear demonstrated an erythematous mass in the middle ear space, involving the upper posterior quadrant of the tympanic membrane and investing the ossicles. The middle ear cavity was otherwise well visualized, and there was nothing on the promontory. Her facial motor function was intact and symmetric. The findings of the complete physical examination were otherwise unremarkable and were without evidence of any syndromic process.

An audiogram confirmed normal hearing in both ears. Computed tomography scanning revealed a small soft tissue mass adjacent to the scutum. There did not appear to be any obvious erosion of the ossicles or scutum. T1- and T2-weighted magnetic resonance imaging demonstrated a linear bright lesion in the superior middle ear cavity, abutting the scutum.

The patient underwent uncomplicated excision of the mass through a right postauricular-transcanal approach. The lesion was easily separated from the tympanic membrane and malleus. Tumor in the epi-

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Presented at the 2007 Southern Section Meeting of The Triological Society, Marco Island, Florida, February 15-17, 2007.

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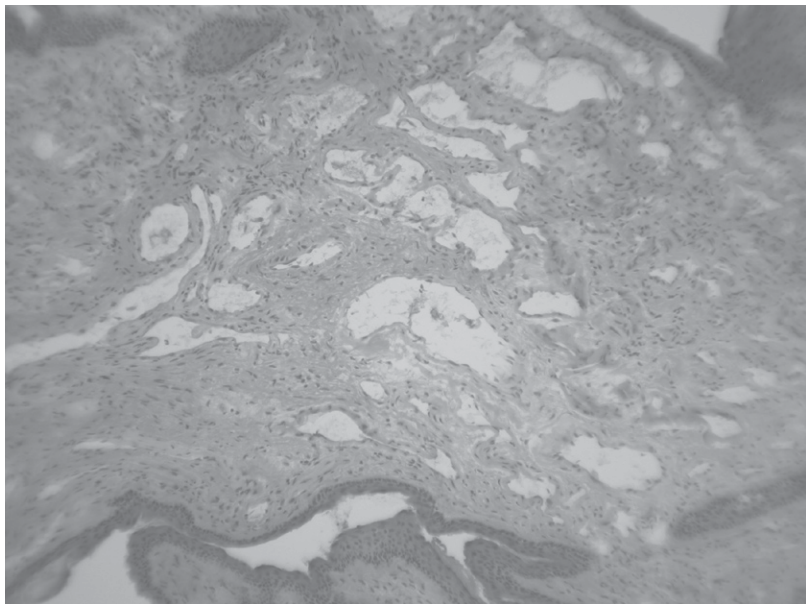

Irregular dilated lymphatic channels of various sizes exist in loosely collagenous stroma.

tympanum was removed by curettage. During the operation, we estimated the lesion to be 8 mm. No significant bleeding was encountered. Temporalis fascia was used for underlay grafting.

Histologic evaluation of the excised middle ear mass demonstrated irregular dilated lymphatic channels of various sizes in an irregular arrangement (see Figure). The channels were lined by endothelial cells, and the lumina contained sparse mononuclear cells and no red cells. The dilated channels had peripheral strands of a collagen network and thin fascicles of smooth muscle. The stroma was loosely collagenous. A noninflamed metaplastic squamous epithelium was overlying the lymphangioma. On the basis of the histologic architecture and the endothelial cytologic pattern, the diagnosis of the middle ear mass was a lymphatic malformation. Without evidence of any event that would lead to the development of an acquired lymphatic malformation, the lesion was considered to be congenital.

The patient had no postoperative complications, including facial motor dysfunction, imbalance, and subjective or objective hearing loss. The patient has been serially evaluated for 18 months since the resection and continues to demonstrate no recurrence or complication. The last audiogram was performed at 12 months after resection and was normal.

DISCUSSION

Lymphatic malformations were first described by Redenbacker in 1828.[1,2] As they were originally considered a vascular neoplasm, the common label lymphangioma continues to be seen in the literature today. The term lymphatic malformation has gained favor, however, as it is considered by experts to be a more accurate description of the congenital pathologic process without suggesting neoplasia.[2]

Over time, the understanding of lymphatic malformations has evolved, and multiple theories detailing the pathogenesis of lymphatic malformations have been proposed. Sabin[3] describes our lymphatic system as originating from 5 primitive venous buds. It is theorized that during this process, lymphatic tissues extend from the buds abnormally, becoming sequestered and forming lymphatic malformations.[1,3] Other theories have been proposed, including the centripetal theory of lymphatic development described by McClure and Huntington.[1,2,4] Acquired lymphatic malformations have also been reported to occur after infection, trauma, or chronic lymphatic compression.[1,4] Independent of cause, the pathologic implication of the lymphatic malformation is the failure of lymphatic vessels to drain their contents into the venous system.[1,5]

The middle ear and mastoid mucosa are drained by lymphatic channels lying adjacent to the eustachian tube. The deep retropharyngeal and jugular lymph nodes then drain these lymphatic channels. The development of this lymphatic network occurs as mesenchymal cells differentiate into lymphangioblasts and subsequently into lymphatic endothelium and lymphatic vessels. The process is tightly regulated by specific growth factors, by signaling molecules, and by the receptors of each. Vascular endothelial growth factor (VEGF) has been suggested to have a significant role in the early stages of lymphangiogenesis. Christison-Lagay and Fishman[6] described the use of transgenic mice to demonstrate the role of VEGF and certain integrins in the process of lymphatic development. Mice with overex-

pressed VEGF-C and VEGF-D isoforms developed hyperplastic lymphatic vessels. Similarly, deletions or mutations in certain subtypes of integrins were shown to result in abnormal lymphatic development, as the role of integrins is to act as cellular adhesion molecules and signal transducers within the extracellular matrix.[6]

Multiple classification schema exist that categorize lymphatic malformations according to histologic appearance, therapeutic implications, or disease location.[1,4,7] Wernher described one of the first histologic classification systems, dividing lymphatic malformations into 1 of 3 categories: cystic (cystic hygroma), cavernous, or capillary.[1,2,4] Another system divides cystic lymphatic malformations on the basis of size. Lesions 2 cm^3 or greater are labeled macrocystic, and those under 2 cm^3 are microcystic.[1] A combination of classification systems is commonly used to describe a lymphatic malformation and predict a response to treatment. The histopathologic description of the lesion in the patient described herein, including dilated thin-walled spaces lined with flat endothelial cells, is consistent with that of a cystic lymphatic malformation. Independent of differences in histology, location, or size, each category of lymphatic malformation is believed to be the product of the same pathologic process.[1]

The evaluation of a lymphatic malformation includes a detailed history and physical examination. When the lesion is located in the middle ear, its differential diagnosis is extensive and may include hemangioma, rhabdomyosarcoma, cholesteatoma, neuroendocrine tumors, or schwannoma. In addition to solitary lesions, lymphatic malformations are commonly present in association with specific syndromes, including Turner, Klinefelter, Fryns, and Noonan syndromes and certain trisomies.[1] For this reason, the physical examination must include the entire body, and not be limited to the head and neck.

Imaging studies are equally important in the diagnosis of a lymphatic malformation. Computed tomography, ultasonography, and magnetic resonance imaging have all been implemented. Magnetic resonance imaging is generally the preferred imaging modality, as it has satisfactory soft tissue resolution. Computed tomographic scanning, however, has been touted for its ability to assist in surgical planning.[1,4,8]

Treatment options for a lymphatic malformation include observation, injection of sclerosing agents, and surgical excision. Controversy exists in the literature regarding the effectiveness of observation as a treatment option for lymphatic malformations. Of 46 described head and neck lymphatic malformations, Kennedy et al[4] reported that 7 of 8 observed patients (87.5%) underwent complete spontaneous resolution. The rate of spontaneous regression has also been reported to be as high as 15% by Giguère et al.[1] In contrast, Naidu and McCalla[2] recommended prompt surgical intervention in all adults with head and neck lymphatic malformations to avoid a difficult excision following infection or acute enlargement of the mass.

Surgical excision is considered by many authors to be the treatment of choice for the removal of lymphatic malformations.[1,2,4,5,8] The ability to adequately resect a lymphatic malformation depends on both the size and location of the tumor. Lesions in the tongue, pharynx, and larynx have been shown to have higher rates of recurrence than those involving the upper aerodigestive tract. Bilateral lymphatic malformations and lesions greater than 5 cm are unlikely to be resectable in entirety without postoperative functional impairment.[1] Authors agree that surgery should not be excessive, and significant nerves and vessels should not be sacrificed.[1,5] Complications from surgical excision include infection, nerve paralysis, and excessive blood loss, predominantly in the pediatric population.[1,5,8] Although there is a persistent debate regarding the treatment for lymphatic malformations of the head and neck, most authors agree that the presence of worsening symptoms, rapid tumor growth, or airway compromise precludes an observational approach.[1,4,5,8]

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