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Medical Imaging Quiz – Case 74

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ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2023, 40(4):575–576

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A 25-year-old female presented to the emergency department due to a painful enlarged neck mass. She referred having a small neck mass since she was a child. Physical examination revealed a compressible mass and a neck computed tomography (CT) revealed pathological finding (fig. 1).

Comments

Lymphangiomas are benign malformations of the lymphatic system. Cystic hygromas known as macrocystic lymphatic malformations (LMs) are macrocystic lymphangiomas that most commonly occur in the cervicofacial regions, particularly at the posterior cervical triangle in infants.

Lymphangiomas may also be classified on the basis of their radiographic morphology into three categories: macrocystic (lymphatic structures with diameter >2 cm), microcystic (lymphatic structures with diameter <2 cm), and mixed. Treatment varies according to morphology, with each form posing unique management challenges.

The World Health Organization (WHO) has recognized three types of lymphangiomas: capillary, cavernous, and cystic. The first term is a synonym for microcystic lymphangioma, and the second and third concern macrocystic lymphangiomas.

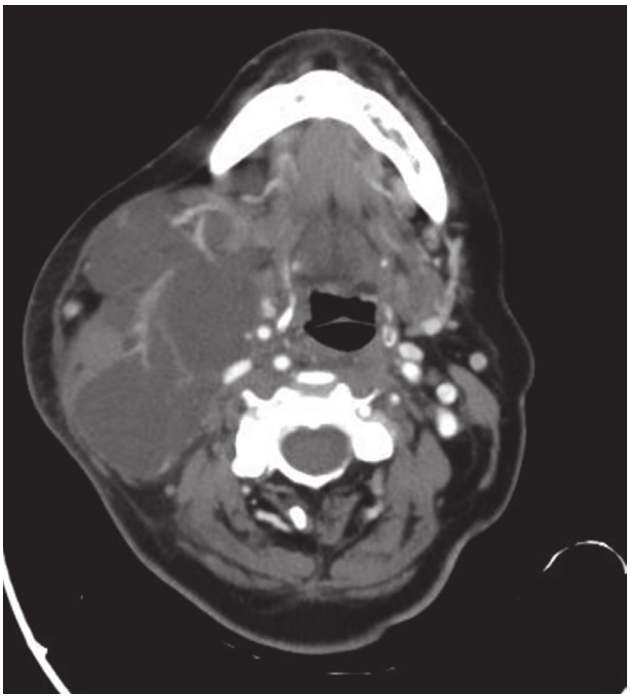

Figure 1. Neck computed tomography (CT) with intravenous contrast enhancement revealed an enlarged right hypoattenuating neck cystic mass.

LMs can be either congenital or acquired. Acquired LMs can arise from trauma (including surgery), inflammation, or lymphatic drainage pathway obstruction. LM tends to be more common in patients with Turner syndrome, Klinefelter syndrome, trisomy 21, and trisomy 18 (as well as 13), though these are not considered to be causative. Karyotypic abnormalities are present in 25–70% of children with LMs.

Incidence of LM is estimated to be 1 per 6,000–16,000 live births. Intrauterine alcohol exposure has been associated with the development of LM. Most LMs (50–65%) are evident at birth, and 80–90% are present by age 2 years. Some authors believe that all LMs are present at birth, even though they may not have fully manifested at that time. The sex distribution is equal. Most series have reported no racial predominance, though a decreased incidence in African Americans has been described. Patients in the infantile or pediatric population can present with pain, dyspnea, infection, hemorrhage, or respiratory compromise. It is compressible and transilluminable.

They are thought to arise from delayed development, failure of the lymphatic system to communicate with the venous system of the neck. Microscopically, they are comprised of endothelium-lined cystic spaces with scanty stroma. They can vary significantly in size. Lymphatic vascular malformations may be mixed with other forms of vascular malformation, including capillary or venous. They are usually well-circumscribed and are of fluid density. Cystic hygromas may also have an infiltrative appearance and may be uni- or multilocular. The density can also be variable with a combination of fluid, soft-tissue density and fat.

CT commonly reveals a hypoattenuating ill-defined neck cystic mass. Magnetic resonance imaging (MRI) reported signal characteristics include T1: predominantly low signal unless there are hemorrhagic components, T2: predominantly high signal, T1 C+ (Gd): no enhancement on any component except occasional faint enhancement of rim.

Management may be by surgical excision and surgical drain-

age to limit recurrent lymphatic collection. Watchful waiting for LMs should be considered only in patients who are asymptomatic.

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