Case Report

Dumbbell–shaped lymphangioma of neck and thorax

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ABSTRACT

Vascular malformations consist of a spectrum of lesions involving all parts of the body. They have different terminologies like vascular tumors, vascular malformations, vascular anomalies, and so on, which create a lot of confusion in understanding and treating these pathologies. Of late, classification on the basis of cellular kinetics and clinical behavior has been devised. Hemangioma is the most common vascular tumor. Vascular malformations are of either lymphatic or capillary and venous origin. Sometimes they are of a mixed origin. Lymphangiomas are common in the face and neck area. They are also not unusual in the mediastinum. We present a case where a huge lymphangioma occupied the right supraclavicular area of the neck, extending to nearly the entire right thoracic cavity, compressing the whole lung. The patient had occasional symptoms of cough. Ultrasonography (US) and computed tomography (CT) images clearly described the lesion as cystic, occupying both the supraclavicular and thoracic cavity compressing the lung parenchyma. Surgical excision was planned and we approached the lesion via both cervical and thoracotomy incisions. Complete excision was done, and the lung expended fully after the surgery.

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Introduction

Lymphangiomas are unusual malformations deriving from the detachment of lymph sacs from the venous drainage systems.[1] Lymphatic malformations are usually present at birth (65 to 75%), but sometimes they become evident during childhood or adolescence.[2] There are two clinical types of lymphatic malformations: The macrocystic type (cyst size of more than 2 cm^3), misnamed cystic hygroma, and the microcystic type (cyst size of less than 2 cm^3), which is more infiltrative, with clear or hemorrhagic vesicles on top.[3] Histologically, lymphatic malformations consist of chyle-filled cysts lined with endothelium.[4] Combined lateral marginal veins (LMVs) are common. A cystic variant of lymphangioma is included in the general classification of lymphangiomas according to Landing and Farber[5] enclosing the capillary and venous variant, characterized by dilatation of the capillary and sinusoidal lymphatic vessels, respectively, (the latter with continued growth of the stromal component), which remain connected to the lymphatic network. The cystic variant instead, is unconnected with the lymphatic network and is characterized by several spaces filled with proteinaceous and chylous material, which contains lymphocytes and sometimes red blood cells.[6] Patients suffering from cystic lymphangioma of the neck and mediastinum extending into thorax are often asymptomatic, but because of lesion growth they may present with cough, respiratory disorders, dysphagia or vascular compression syndromes. Complications arising from nonsurgical resection include hemorrhages and infections.[7]

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An 18-year-old boy presented with a large swelling in the right supraclavicular area and occasional cough

[Figure 1]. On clinical assessment the lesion was cystic and the lower border of the lesion was not reachable.

The plain x-ray was suggestive of a vague swelling on the right upper thorax and lower neck [Figure 2]. A computed tomography (CT) scan suggested nonvascular swelling making a probable diagnosis of lymphangioma occupying both the thoracic cavity and lower neck [Figure 3]. The patient underwent surgery, which necessitated both thoracic and neck incisions for completion of the procedure of resection. Thoracotomy was performed first and the neck component of the lesion was delivered through the thorax, however, it needed an incision over the neck [Figure 4].

Discussion

Cystic lymphangioma or cystic hygroma is a low-flow, vascular malformation, developing where the lymph sacs are separated from the venous drainage system. The most common location is the neck region (75%), especially in the posterior triangle and in the back cervical cavity, but it can be also found in the axillary region (20%) and infrequently in the retroperitoneum, mesentery, omentum, colon, pelvis, groin, bone, skin, scrotum, and spleen;[8] Only 1% of cystic lymphangiomas have mediastinal localization and the most frequent locations are the anterior and upper mediastinum (50-60%), while the middle and posterior mediastinum occur more rarely (20%). In literature, they are described cases of cystic lymphangioma of the mediastinum localized in the right paratracheal, paracardiac,[9] anterior,[10] and posterior mediastinal[11-14] regions.

The present case deals with a cystic hygroma that originated from the lower neck and reached the upper mediastinum in an anteroinferior direction. Approximately 70% of cystic hygromas are already present at birth and more than 90% manifest within the first two years of life; Symptoms depend primarily on the location of the lesion and the possibility for the mass to grow rapidly, for the onset of hemorrhages or infections. Patients with lymphangioma of the neck and mediastinum are usually asymptomatic, but they may present with chest pain, cough, dyspnea, dysphagia or vascular compression syndromes, if the lesion reaches dimensions large enough to cause compression of the adjacent structures. Just for the tendency to expand locally

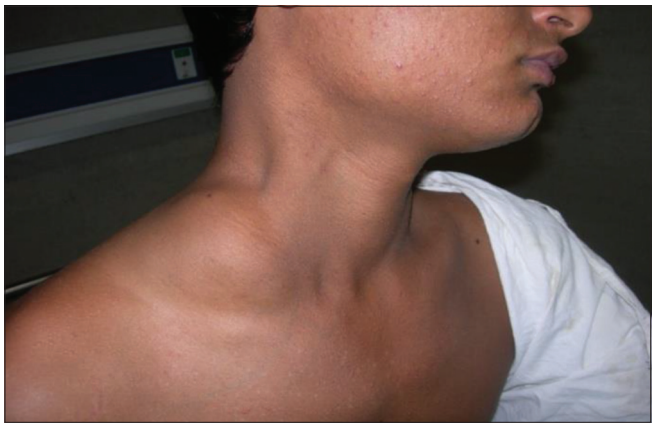

Figure 1: Swelling in the right supraclavicular area

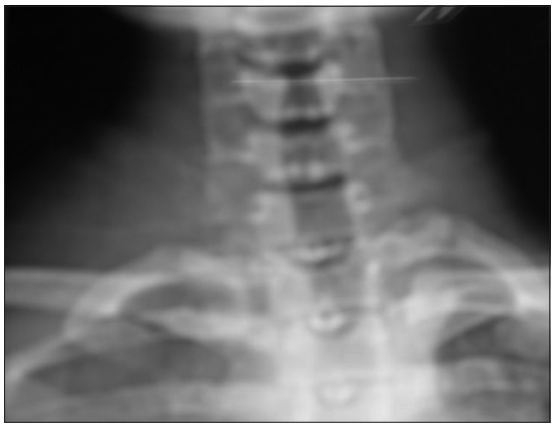

Figure 2: Plain X-ray showing soft tissue density in the supraclavicular area

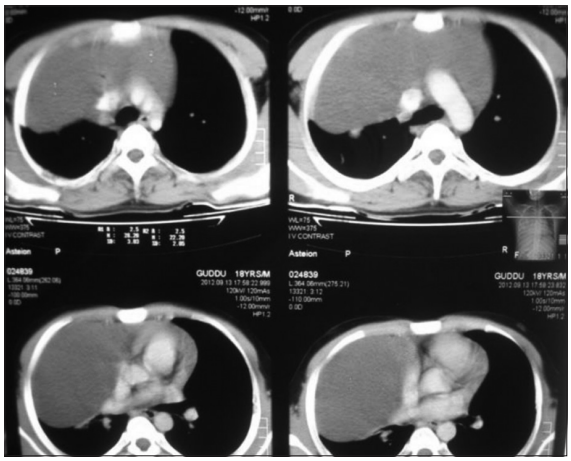

Figure 3: A CT scan thorax showing the intrathoracic lesion

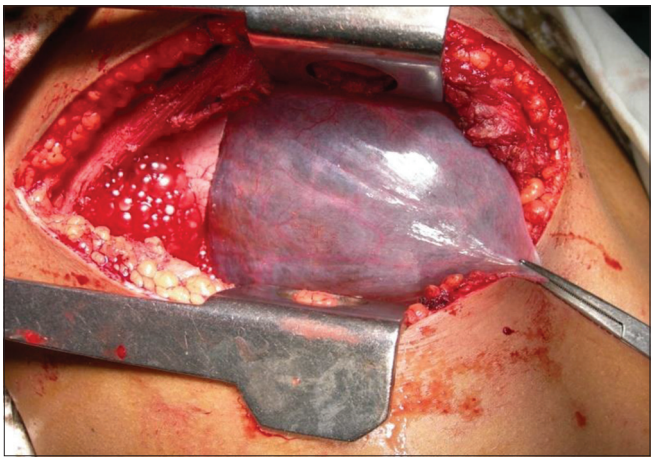

Figure 4: Intraoperative image showing the intrathoracic large cyst

and for the risk of complications, surgical resection is the treatment of choice for a cystic hygroma. In our case the patient had cough due to irritation of the compressed and collapsed lung parenchyma. Even if a cystic lymphangioma can occur with atypical findings, such as, heterogeneous content, irregular borders, and calcification, the diagnosis may be obvious in cases of typical features, such as, fluid content and regular margins. Imaging has a fundamental role in the detection and characterization of the cystic component in the diagnosis of cystic lymphangioma with ultrasonography (US), CT, as well as with magnetic resonance (MR). Moreover, imaging can help in the differential diagnosis between cystic lymphangioma and other cystic masses, including, cystic thymoma, cystic teratoma, hematomas, necrotic tumors, and abscess.[1,6] US is the first level tool to investigate a suspected mass of the neck because of its noninvasiveness, low cost, and non-use of ionizing radiation. In our case the lesion was accurately characterized by US, with demonstration of fluid content and internal septations, without significant vascularization. However, US had to be integrated with CT and MR scans because of its non-panoramic view and for obtaining more information about the structural features and internal as well as peripheral enhancement patterns. A CT scan has been currently performed with the multislice technique that allows volumetric acquisition of the selected anatomic region with multiplanar reconstruction using different methods, such as, *maximum intensity projection* (*MIP*), multiplanar reconstruction (*MPR*), *shaded surface display* (*SSD), and volume rendering* (*VR*). However, CT imaging is performed with radiation exposure. Conversely, MR can give specific information about the fluid content even when the CT shows greater intralesional attenuation values for the presence of a proteinaceous material. Moreover, MR is helpful in surgical planning, on account of its multiplanarity and high-contrast resolution. The definitive diagnosis of cystic lymphangioma is histological after surgical resection; however, percutaneous needle biopsy is considered conclusive in cases of retroperitoneal localization.[15] In our case, the patient underwent surgery and histopathology was done to confirm the diagnosis.

Conclusion

In conclusion, integrated as well as correlative diagnostic imaging is useful for the evaluation of cystic lymphangioma, clearly demonstrating lesion structure and morphology. In particular, US and MRI, avoiding CT radiation exposure, should be preferred, as patients need repeated scans before surgery or during the follow-up, when surgical treatment is refused or postponed; Furthermore, the tri-dimensional imaging evaluation using advanced modalities is fundamental to assess lesion anatomy and local spread.

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