Case Report

Isolated Mediastinal Lymphangioma Herniating Through the Intercostal Space

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Lymphangiomas are congenital malformations of the lymphatic system, and 90% have manifested by the end of the second year of life. While 75% of these are located in the cervical region, only 2% to 3% are associated with an intrathoracic extension. An isolated mediastinal lymphangioma without a cervical component is an uncommon occurrence. Presented here is an isolated mediastinal lymphangioma that herniated through the intercostal space to present as a cystic mass in the parasternal region, which has not been reported so far. [Asian J Surg 2004;27(3): 241–2]

Introduction

Cystic hygroma is a congenital malformation due to maldevelopment of the lymphatic system. About half present in the first year of life, and 90% manifest by the end of the second year of life. Lymphangiomas predominantly occur in the cervical region (75%) and the axillary region (20%). Of cervical lymphangiomas, only 2% to 3% may be associated with an intrathoracic extension. An isolated mediastinal lymphangioma without a cervical component is an uncommon occurrence that can pose a diagnostic dilemma. Presented here is an isolated mediastinal lymphangioma that herniated out through the intercostal space to present as a cystic mass in the parasternal region.

Case report

A 4-year-old boy presented with a history of a progressively enlarging lump in the left parasternal region of 2.5 years' duration. The lump was not associated with pain. There was no history of local trauma. Examination revealed a cystic, non-tender 4×4 cm lump in the left parasternal region over the third intercostal space (Figure 1).

The swelling was transilluminant; on coughing, the parasternal cystic mass became tense and turgid, which gave the clinical indication of the mass having an intrathoracic component. Chest X-ray did not reveal any abnormalities. Ultrasound showed a multicystic lesion in the thorax anterior to the main pulmonary artery and right ventricle with a small extension into the subcutaneous tissue through the intercostal space. Computed tomography showed a 5×3×2 cm cystic lesion in the anterior mediastinum located anterior to the vascular structures and tracheobronchial tree, with a small extension into the anterior chest wall (Figure 2).

A midline sternotomy was performed. Both lobes of the thymus were delineated and the cystic mass separated from it. The mediastinal mass along with its subcutaneous component protruding through the intercostal space was excised. Histopathology confirmed it to be a lymphangioma. Two years' follow-up showed no evidence of recurrence.

Discussion

An isolated mediastinal lymphangioma without a cervical component is an uncommon occurrence. Such tumours may be asymptomatic or may have a diverse presentation.
They have been reported to manifest as acute respiratory distress,\textsuperscript{1,2} respiratory failure\textsuperscript{3} and hoarseness.\textsuperscript{4} Mediastinal lymphangioma has also been reported to mimic teratomas.\textsuperscript{5} In the mediastinum, lymphangiomas are usually located in the anterior mediastinum, but may be located in the posterior mediastinum.\textsuperscript{6}

Presentation of an isolated mediastinal lymphangioma as a cystic mass in the parasternal region due to herniation of the cysts through an intercostal space has not been reported so far. Magnetic resonance imaging of the thorax is essential in cases of mediastinal and intrathoracic lymphangiomas because it is extremely valuable in the assessment of the extent of the disease as these tumours are often infiltrating. Unlike cervical lymphangiomas, treatment with intratantal OKT-432 is a limited option since a reactionary increase in the size of the cysts can compromise respiration. These tumours can also be treated using thoracoscopy.\textsuperscript{7} However, a surgical approach via a midline sternotomy is easy and provides excellent exposure for the meticulous dissection of all the cysts, which is essential to prevent recurrence. Not only the vascular structures but also the phrenic nerves must be delineated and safeguarded.

References