Giant Thoracocervical Lymphangioma with Multivisceral Involvement: A Different Concept

ABSTRACT

Background: Lymphangiomas are benign lymphatic malformations frequently seen in head and neck region. Most reported cases are cervical lesions with mediastinal extension.1,2 Most are conceived as due to failure of fetal lymph sacs. Synchronous or metachronous lymphangiomatosis with cystic hygroma of the neck is also reported.3

Case details: Massive multicompartmental mediastinal lymphangioma with cervical extension and concomitant visceral involvement is presented. It is proposed as a hamartomatous tumor rather than maldevelopment of fetal jugular lymphatic sacs.2 Solid intrathoracic component with cystic neck extension supports mediastinal origin. Cell culture lines at 4 months were positive for endothelial cell lines positive for factor VIII antigen.

Conclusion: Giant thoracocervical lymphangioma is more likely to be a tumor rather than simple fetal failure of lymphatic sac fusion. Surgical excision is curative.

Keywords: Giant thoracocervical lymphangioma, Multivisceral involvement, Magnetic resonance imaging.

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INTRODUCTION

Lymphangiomas are benign lymphatic malformations frequently seen in head and neck region. Most reported cases are cervical lesions with mediastinal extension.1,2 Synchronous or metachronous lymphangiomatosis with cystic hygroma of the neck is also reported.3 Massive multicompartmental mediastinal lymphangioma with cervical extension and concomitant visceral involvement is presented. It is proposed as a hamartomatous tumor rather than maldevelopment of fetal jugular lymphatic sacs.2 Solid intrathoracic component with cystic neck extension supports mediastinal origin. Magnetic resonance imaging (MRI) and other radiological findings were correlated with clinical, surgical and histological features.

CASE REPORT

A 17-year-old boy presented with painless swelling in left side of the neck associated with heaviness and discomfort in the chest of 4 years’ duration. There was no history of dyspnea, dysphagia or repeated chest infections.

The swelling in the neck was supraclavicular and deep to sternocleidomastoid. It was soft, cystic, fluctuant and transilluminant. Lower pole of mass could not be reached (Fig. 1A).

Chest radiograph lateral view (Fig. 1B) revealed large homogeneous opacity occupying anterior and middle mediastinum with compression of left lung posteriorly. Ultrasound scan of neck showed a 4.5 × 1.5 cm cystic lesion with regular margins, multiple internal septae, avascular signal on Colour Doppler extending intrathoracically (Fig. 1C). The mass was not compressing the barium filled esophagus (Fig. 1D). Magnetic resonance imaging revealed a thoracocervical mass of 25 × 14 × 11 cm size. It was occupying the anterosuperior, anterior and middle mediastinal compartments compressing the lung posteriorly. It extended from the left hemidiaphragm to lower neck without displacement of major mediastinal vessels (Fig. 2A).

The lesion extended to left half of the neck through the thoracic outlet. Superficial subplatysmal component and deep substernomastoid posterior triangle component formed a dumbbell. Internal jugular vein was surrounded by the mass (Fig. 2B). The mass was multilocular, septate with smooth contours and thick walls. It was isointense (to skeletal muscle) on T1-weighted images with intermittent hyperintense signals signifying solid cystic nature; T2-weighted images showed hyperintensity. Subplatysmal component was homogenous as against intrathoracic component. Mass was separated from the surrounding structures without compression.
Further MRI also showed T2 hyperintense lesions in the spleen, vertebral bodies, ribs, the upper and lower extremity bones and bones of pelvis (Fig. 2C) without bone marrow edema, periosteal reaction or accompanying soft-tissue mass. Computed tomography (CT) scan of the chest (Fig. 2D) revealed low density lesion with attenuation values ranging between 18 and 36 HU with multiple internal septae. No calcification was seen within the lesion.

Surgical exploration revealed superficial subplatysmal component and deep substernomastoid posterior triangle component having a dumbbell connection. Retrosternocleidomastoid swelling was compressing the IJV medially. Extension into mediastinum behind manubrium sterni did not show pressure effect on trachea and esophagus. Mass was large and solid occupying entire left hemi-thorax necessitating additional left anterior thoracotomy as solid nature prevented decompression. Mass was excised sparing the phrenic nerve (Fig. 2E).

Recovery was uneventful and left lung expanded completely. Histopathology was hamartoma with neural component. Cultured cells from mass at 4 months showed a viable primary endothelial cell line positive for factor VIII-associated antigen.

DISCUSSION

Cystic lymphangiomas, first described by Rodenber in 1828, are rare benign lymphatic tumors with equal distribution among genders and races. Ninety percent of lymphangiomas develop on body surface, 75% in neck, mostly posterior triangle and 20% in axilla. Most are detected before 2 years of age. One percent of cystic lymphangiomas have mediastinal localization. Frequent locations are the anterior and upper mediastinum (50-60%) with rare occurrence in the middle and posterior
mediastinum. Cystic lymphangioma of the mediastinum in the right paratracheal, paracardiac, anterior and posterior mediastinal regions have been reported. Less frequent sites are in the adrenal gland, kidney, bone, omentum, gastrointestinal tract, retroperitoneum, spleen, liver and pancreas. They grow proportional to patient’s body growth. There are only few reports of giant mediastinal lymphangiomas. Whether lymphangiomas are true malformations or tumors is not addressed comprehensively in literature.

Lymphangiomas are conventionally hypothesized to be hamartomas representing sequestered lymphatic sacs which fail to communicate with the remainder of the lymphatic or venous system, during the development in 8th week of gestation. Subsequently, sequestered lymphatic tissues lead to cystic morphology.

The expression of different growth factors in the endothelium of lymphangiomas supports tumor origin. Bowman et al hypothesized that cystic hygroma represents an expanding proliferating endothelial growth process and not simply a sequestered lymphatic receptacle.

Lymphangiomas traditionally are divided into capillary, cavernous or cystic, and macrocystic, micro-cystic or combined, on gross morphology.

Histopathology showed lymphangioma with single layer of flattened endothelium lined spaces filled with eosinophilic proteinaceous material between fat, fibrotic and neural and lymphatic structures. Immunochemistry confirms the endothelial line of differentiation in lymphangiomas. Cells cultured from the mass yielded an endothelial cell line positive for factor VIII-associated antigen.

Solid mediastinal component with skeletal muscle intensity on MR and inability to decompress at surgery indicate tumor behavior in the present case.

Site of primary focus (chest or neck) is a controversy ignored in previous publications. All cervicothoracic lesions except superficial lesions extend from the neck to the thorax through the thoracic inlet. In the present case, we strongly believe in an intrathoracic origin because of greater intrathoracic component (> 95%). Only the cystic component extended along neurovascular planes into the neck. If it was the other way round, chest component also would have been cystic. Normally, when lymphangiomas extend, they go behind the lung. Solid intrathoracic component was anterior, pushing the lung posteriorly. Imaging findings also support below upwards migration; more of chest component extending into neck due to lack of space.

The issue of disseminated lymphangiomatosis vs diffuse lymphangiomatosis with associated giant lymphangioma also deserves mention. The former represents a proliferation of lymph vessels involving soft tissue skin and the skeletal system. Clinical presentation depends on the organ involved and extent of the mass. Diffuse lymphangiomatosis occurring synchronously or metachronously with cystic hygroma of the neck is occasionally reported.

Giant intrathoracic solid component and cystic cervical part with patchy multivisceral involvement distinguishes the present case from disseminated lymphangiomatosis.

Integrating the ultrasound (US), CT and MRI scan findings yields comprehensive anatomical details. The common feature in the diagnosis of lymphangiomas are uni- or multilocular predominantly cystic masses, clear borders, septations with lobular contours, contiguous extension along neurovascular planes of mediastinum without mass effect. Magnetic resonance imaging in cystic hygromas is fundamental and indispensable because of its advantages. Multplanar, nonionising, high contrast resolution with tissue-specific signal intensity provides accurate anatomy and internal structure of the lesion.

Lymphangiomas are hyperintense to fat on T2-weighted MR images, and are isointense to muscle on T1-weighted. High signal intensity on T1 represents elevated protein content in the fluid or combination of fluid, solid tissue and fat.

In the present case, MRI showed a cystic-solid septate lesion with detailed anatomy, in particular, the close proximity to the brachiocephalic and common carotid arteries. A large predominantly solid intrathoracic lymphangioma with cystic extension into neck presented as a complex mass. Hence, a tumor origin with solid components is preferred to conventional fetal nonunion of lymph sacs.

The treatment options of intrathoracic lymphangiomas also range from surgical excision to intralesional sclerosant to radiotherapy. Surgical treatment is the gold standard and preferred by many.

The bigger the lesion, the greater is the technical difficulty of excision. Surgeon’s mindset regarding challenging dissection around the vital structures supported the use of intralesional sclerosant agents. Surgical excision was preferred in the present case due to solid cystic nature of the mass.

Visceral involvement (splenic/bony) are kept under observation for growth and symptoms expecting spontaneous resolution.

CONCLUSION

Giant solid cystic intrathoracic lymphangioma with extension into neck with multivisceral involvement is a rare complex presentation. Tri-dimensional MRI with extension into neck with multivisceral involvement is a rare complex presentation. Tri-dimensional MRI with extension into neck with multivisceral involvement is a rare complex presentation.
sequestered fetal lymphatic channels and lead to option of surgery.

Immunohistochemistry confirms the endothelial line of differentiation in lymphangiomas. Tissue cultures of explants yield a primary endothelial cell line positive factor VIII-associated antigen.

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REFERENCES