

Case report

Child with Mediastinal cavernous lymphangioma

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Abstract

Mediastinal cavernous lymphangioma is a benign rare lesion originating from lymphatic system. It is usually asymptomatic. We have presented a 2 year old male child with fever for seven days. Opacity was found in chest X-ray in the upper part of right hemithorax, merged with the mediastinum. CT scan of chest was performed, which revealed a large, lobulated, smoothly marginated non-enhancing, low density, mediastinal mass, involving right half, extending from root of neck. Finally, pathological examination of the surgical sample indicated 'Cavernous lymphangioma'.

Key words: Child, imaging, cavernous lymphangioma

Introduction:

Mediastinal lymphangioma is a histologically benign proliferation of interconnecting lymphatic vessels and sacs. It may grow in an infiltrative fashion^{1,2}.

Mediastinal lymphangioma is rare (less than 1%) and when confined to the mediastinum, they are usually asymptomatic³. Fewer than 10% of cases occur in the posterior mediastinum. Lymphangioma usually appears in the childhood (90% of cases present at <2 years of age)⁴. Above all cavernous type of mediastinal lymphangioma is very rare⁵. We have presented a case of childhood asymptomatic mediastinal cavernous lymphangioma, involving the mediastinal compartments.

Case presentation:

A two year old male child presented with fever for seven days. He did not have cough, sputum, chest pain or anorexia. The parents gave history of right sided neck swelling of the child since birth, which was diagnosed as cystic hygroma and underwent sclerotherapy injection bleomycin, 1 year back. No history of malignancy was found in his family. Physical examinations revealed no abnormality.

In laboratory findings, complete blood count and arterial blood gas analysis were within normal limit. Chest X-ray showed opacity in upper part of right hemithorax.

Then CT scan of chest without and with I/V contrast was done. The CT scan shows a large, lobulated low density mass lesion extending from the root of the neck, chest wall and all compartment of right sided mediastinum, predominantly anterior and middle mediastinum, down to right principal bronchus. Few of the areas are of fluid density.

No calcification is seen. The lesion is non-enhancing. The vessels are not compressed, rather the mass insinuates through the vessels. The trachea and bronchi are not compressed by the mass. No evidence of any nodule, consolidation or pleural effusion is seen on either side. Right postero-lateral thoracotomy was performed. The mass was resected and sent for pathological examination. Pathological examination reported dilated empty spaces, lined by endothelial cells and intervening lymphoid tissue. The lymphoid tissue was composed of small lymphocytes and occasional follicles.

The pathological diagnosis was: Cavernous lymphangioma.

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Fig I: Large lobulated low density mass in anterior, middle and posterior mediastinum.

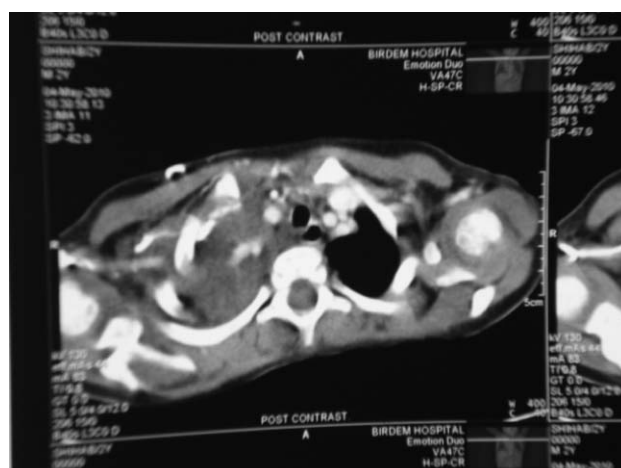


Fig II: The non enhancing mass has insinuated through vessels

Discussion:

Lymphangiomas are benign tumours of mesodermal origin consisting of proliferating lymph vessels of varying sizes. Lymphangiomas have been classified into three groups: lymphangioma simplex, composed of capillary sized thin walled lymphatic channels, Cavernous lymphangioma, composed of dilated lymphatic channels, often with fibrous adventitia coats, and cystic lymphangioma or cystic hygroma, composed of cysts that range from a few millimeter to several centimeter in diameter⁶. Cavernous type is microscopic thin walled lymphatic channels with associated stroma that consist of varying amount of spindle shaped smooth muscle cells, collagen bundles, fibroblasts, and lymphocytes^{5,7,8} similar to our pathological report. Cavernous lymphangioma of the thorax are extremely rare⁹.

Mediastinal and cervicomediastinal lymphangiomas

are rare lesions that can be treated with surgical excision. Prognosis appears to be excellent because no difference in survival was found between patients and age and sex matched control. M:F ratio is 1:3, dyspnoea being most common symptom¹⁰.

Approximately, 1% of all lymphangiomas are confined to the chest⁹. Most lymphangiomas are discovered in fetuses, neonates, or young children. Fifty percent are present at birth, and 90% are discovered by two years of age^{1,11}. Ninety five percent involve the neck or axilla, and 10 percent extend into the superior aspect of the anterior mediastinum¹² or less commonly other mediastinal compartments¹³. In Japanese medical literature, only seven cases of cavernous mediastinal lymphangioma were reported¹⁴. Most of them were infants or young children. Although the majority of lymphangiomas present in the first two years of life, there has recently been an increased recognition of lymphangiomas in adults^{15,16}.

Mediastinal lymphangiomas may insinuate into surrounding organs. As incomplete resection can result in recurrence, complete tumour removal should be performed, based on accurate preoperative evaluation¹⁷.

Radiological imaging can help in differential diagnosis of mediastinal masses, to determine its exact location, extension and content of mediastinal lymphangiomas. There are few literatures describing the evaluation of imaging and radiological findings in mediastinal lymphangiomas especially cavernous type. CT scan is the golden standard to detect and evaluate mediastinal tumours¹⁸. In 1997 Pue et al. in a study evaluated ultrasound, CT scan and MR images of 18 patients of lymphangioma and found all of these valuable¹⁹. Miyake et al. presented CT results in three cases of mediastinal lymphangiomas. One case showed multiloculated, water-density mass enveloping mediastinal structures, without their displacement or compression; one case was well-defined, homogeneous waterdensity mass in right paratracheal region, and one was associated with haemorrhage into cysts and significant increase in size over five years. According to them, CT is useful for diagnosis and evaluation of extent or contents of mediastinal lymphangioma²⁰. Unusual features of

thoracic lymphangiomas were calcifications, speculated margins, and homogeneous soft tissue attenuation¹². Briefly we presented this case because mediastinal lymphangiomas are rare, especially cav-

ernous type. It is usually asymptomatic. In order to determine more exact radiological characteristics of mediastinal lymphangiomas, more cases must be reported and reviewed.

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