

**Picture Story**

## Mediastinal lymphatic venous malformation mimicking thymic shadow in an infant

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### Introduction

Mediastinal lymphatic venous malformation (LVM) was first described in 1983<sup>1</sup>. LVMs are tumour-like lesions combining dysplastic lymphatic and blood vessel structures<sup>2</sup>. Pathogenesis of LVM is unknown<sup>3</sup>. Clinical manifestations of mediastinal LVMs vary depending on size, location, expansion of mass and the proportion of lymphatic and venous components<sup>3</sup>. LVMs are rare and to our knowledge, there are no case reports of infants in English medical literature. We report a case of mediastinal LVM which was seen as a thymic shadow on the chest x-ray in an infant.


### Case report

A 6.5-month-old, previously well-baby girl presented with high-grade fever (102 °F) associated with a moist cough and cold for 2 days and no other symptoms. Examination revealed a normal respiratory system. Investigations showed increased inflammatory markers (white blood cell count was 25,000/ $\mu$ L with 75% neutrophils and C-reactive protein was 102 mg/dl). Chest x-ray revealed a homogenous opacity over the thymic region (Figure 1).

She was started on intravenous antibiotics. An ultrasound scan of the chest revealed a well-defined multi-loculated cystic lesion measuring 3.5×4.6×5.4cm in size in the anterior mediastinum at the level of ascending aorta in the right paracardiac location suggestive of an infected bronchogenic cyst.

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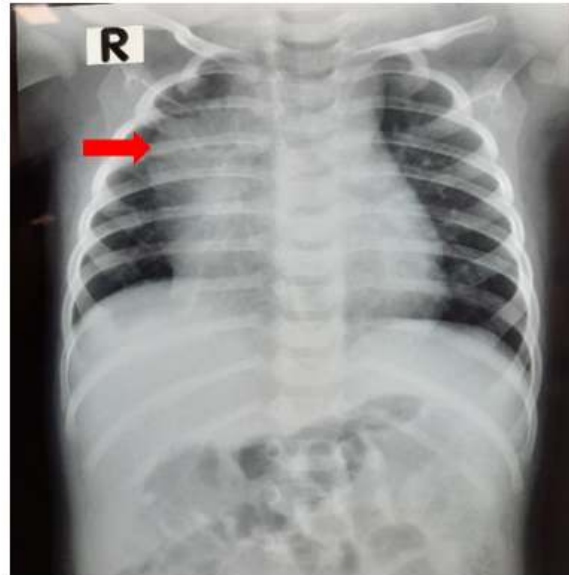
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**Figure 1: Chest x-ray showing homogeneous opacity over thymic region**

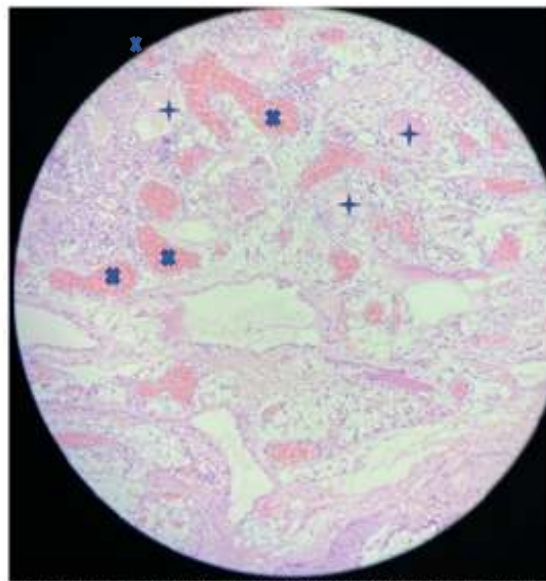
The fever continued despite treatment. On day 4 of illness a contrast-enhanced computed tomography (CECT) scan of chest (Figure 2) revealed a multiloculated cystic lesion in the anterior mediastinum measuring 3.8 (AP) ×4.2 (Tr) ×4.6 (cc) cm. It had a thin wall and abutted thymus on the left lateral side. The lesion extended along the right paratracheal region inferiorly up to the diaphragm. There was no significant mass effect. Peripherally, the lesion extended up to the anterior chest wall. CECT findings were suggestive of a mediastinal cystic lymphangioma or thymic cyst.

On the eighth day of the illness, the child underwent laparoscopy-assisted excision of the lesion. Multi-cystic lymphatic malformation within the superior and anterior mediastinum was found and almost 80% of the lesion was excised. The histology report was compatible with a lymphatic venous malformation (Figure 3).

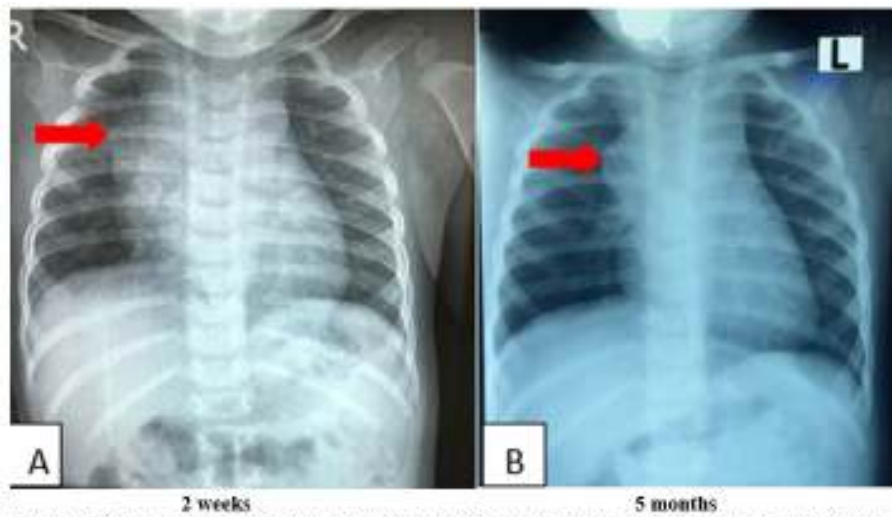
The postoperative period was uncomplicated. She was sent home on day 10 of admission after completing intravenous antibiotics for 7 days. The child was symptom-free and thriving well on follow-up. Follow-up chest x-rays after 2 weeks and 5 months were satisfactory without evidence of recurrences. (Figure 4)



**Figure 2:** Contrast-enhanced computed tomography (CECT) of chest. The horizontal arrow indicates a multiloculated cystic lesion with a thin wall. The vertical arrow indicates the aortic arch.



**Figure 3:** Microscopically, there are dilated vascular channels interconnected with dilated lymphatic channels. Vascular channels are filled with red blood cells (B) and lymphatic vessels are filled with pale eosinophilic material (A).



**Figure 4:** Post-operative chest x-rays showing insignificant debulking with minimal residual changes

## Discussion

LVMs, slow-flow vascular malformations are one of the combined types, which are defined as two or more vascular malformations found in one lesion<sup>2</sup>. The International Society for the Study of Vascular Anomalies (ISSVA) classifies vascular anomalies into two major categories, vascular tumours, and vascular malformations<sup>2</sup>. Vascular tumours are vascular anomalies with a proliferative component<sup>2</sup>. Vascular malformations are relatively static lesions attributed to inborn errors of vascular morphogenesis<sup>2</sup>. Vascular malformations can be subclassified into four types based on their predominant vessel involvement: simple, combined, of major named vessels, and associated with other anomalies<sup>2</sup>. Simple vascular malformations are further classified based on the main types of vessels: capillary, venous, lymphatic, arteriovenous, and arteriovenous fistula<sup>2</sup>.

LVMs are usually found in the cervical, axillary, head, and neck region<sup>3</sup>. LVM in the mediastinum is a rare occurrence and only account for 0.5% of mediastinal tumours<sup>4</sup>. It commonly occurs in the anterior mediastinum and 50% of those patients are asymptomatic, while the other 50% have symptoms of cough, dyspnoea, or haemoptysis. Expanded lesions may compress the surrounding structures<sup>5</sup>.

It was found in the anterior and superior mediastinum in our patient and presented with symptoms of cough and there were no signs of compression on surrounding structures. The growth of these malformations generally commensurate with the age of the patient<sup>2</sup>. However, they may grow rapidly due to hormonal stimulation such as in puberty and pregnancy, or complications such as haemorrhages (often with minor trauma) and infection<sup>2</sup>. In our patient infection may have caused the rapid growth of the tumour.

Radiological findings in computed tomography (CT) and magnetic resonance imaging (MRI) of LMV depend on the proportions of the lymphatic and venous components<sup>3, 4</sup>. MRI is useful for the diagnosis of mediastinal LVM as it can effectively detect slow-flow components and small vessels such as venous connections to the superior vena cava<sup>3</sup>. MRI for our patient was not done due to financial constraints. CT scan is excellent for delineating the lesions, and their relationships to adjacent structures and may reveal signs of infiltration<sup>3</sup>. Imaging studies are helpful in guiding both pre-operative differential diagnosis and subsequent plans of treatment<sup>3</sup>. The definitive diagnosis of LVM was made by pathology<sup>3, 4</sup>.

Surgery and sclerotherapy are the treatment choices for vascular malformations<sup>4</sup>. Surgery is most successful in isolated and symptomatic LMV<sup>4</sup>. Complete excision is difficult in some cases<sup>4</sup>. Recurrence is more common in diffuse form and incomplete excision<sup>4</sup>. As our patient underwent near total excision, close follow-up is essential to see the recurrences.

## Conclusion

VMs are rare congenital tumours which can be misdiagnosed as the thymus in a chest X-ray. The tumour gets more prominent due to haemorrhage, or infection. Therefore, a high index of suspicion is paramount in reaching a definitive diagnosis.

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