

Rare cause of an anterior mediastinal mass causing airway compression in a young child

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DESCRIPTION

A 1-year-old female child presented with recurrent chest infections and a tracheal cough. She had been previously well with no known tuberculosis contacts. The child had distended neck veins but not any facial plethora. Chest X-ray demonstrated a large anterior mediastinal mass extending into the right hemithorax. The trachea was compressed and displaced to the left, while the carina was depressed and splayed (figure 1A). Post-contrast axial CT scan (figure 2A, B) demonstrated a homogenous, non-enhancing soft tissue mediastinal mass of visibly lower density than muscle, predominantly in

the right paratracheal position. There were no calcifications noted. The mass encased the superior vena cava and right subclavian artery, as well as displacing the trachea and carina from their position on the thoracic spine. The trachea was significantly narrowed ('circumferentially') as compared to the right and left main bronchi. The thymus appeared visibly normal. A soft tissue biopsy was performed via an anterior thoracotomy. This specimen was confirmed to be thymic tissue. The symptoms persisted without any improvement, and bronchoscopy confirmed a 50% compression of the upper and middle trachea. This prompted removal of the mass

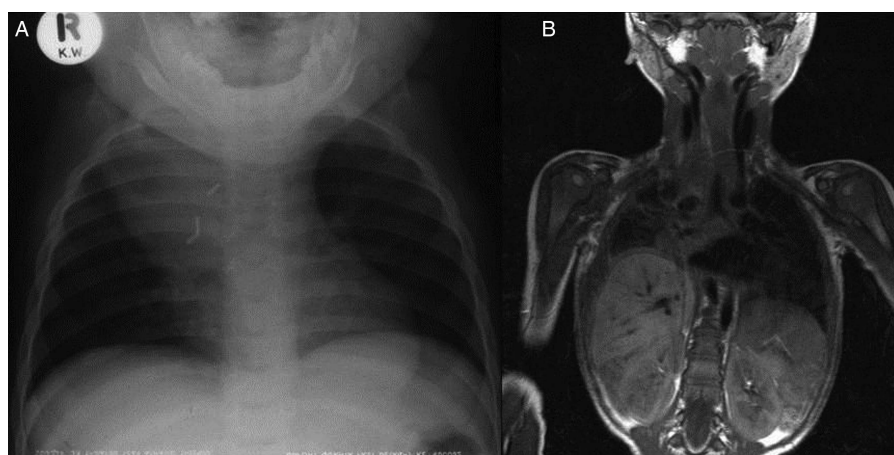


Figure 1 (A and B) Postbiopsy plain radiograph and corresponding postoperative chest MRI. Chest X-ray after biopsy (note the surgical clips) demonstrating a large anterior mediastinal mass extending into the right hemithorax. The trachea is displaced to the left and is compressed while the carina is depressed and splayed. Corresponding T1-weighted coronal MRI confirms the right paratracheal, subcarinal and paracardiac soft tissue mass. There is an elevated right hemidiaphragm that is related to phrenic nerve injury (as a result of the surgical biopsy).

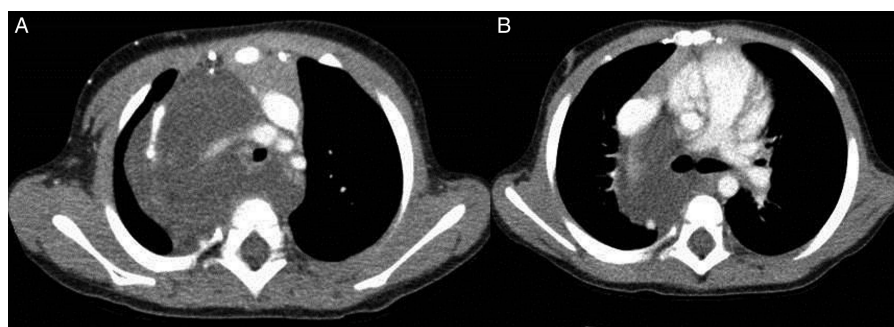


Figure 2 A 1-year-old female child with a lymphangioma of the mediastinum presenting as an anterior mediastinal mass. Postcontrast axial CT of the chest at the level of the major branches of the aortic arch (A) and at the level of the carina (B) demonstrating a homogenous, non-enhancing, soft tissue mediastinal mass of visibly lower density than muscle, predominantly in a right paratracheal position. There are no calcifications noted. The mass encases the superior vena cava and right subclavian arteries, and also separates the trachea and carina from their position on the thoracic spine. The trachea is significantly narrowed ('circumferentially') as compared to the right and left main bronchi. The thymus is visible and normal.



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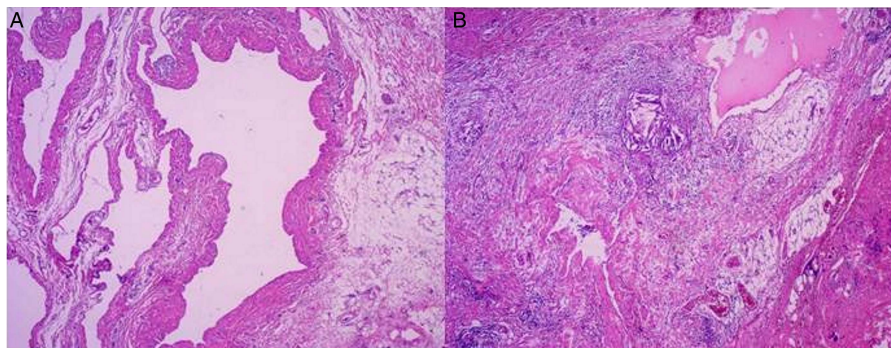


Figure 3 Microscopic examination. (A) H&E stain, $\times 100$, demonstrating numerous large and small vascular spaces, lined by endothelium. These lymphatic spaces have either very thin walls or a small amount of musculocollagenous tissue in their walls. In areas there are small lymphoid aggregates adjacent to the endothelial lining. (B) H&E stain, $\times 40$, demonstrating the intervening stroma between the lymphatic channels consisting of fibrous tissue, fat and nerves. Foci of old haemorrhage with cholesterol clefts are also present. These histological features are consistent with a lymphangioma.

via a lateral right-sided thoracotomy. At surgery, the endotracheal tube was placed bronchoscopically above the carina to make ventilation easier during surgery. At microscopy, there were numerous large and small vascular spaces present, lined by endothelium. These lymphatic spaces had either very thin walls or small amounts of musculocollagenous tissue in their walls. In areas there were small lymphoid aggregates adjacent to the endothelial lining (figure 3A). The intervening stroma between the lymphatic channels consisted of fibrous tissue, fat and nerves. Foci of old haemorrhage with cholesterol clefts were also present (figure 3B). These histological features were consistent with a lymphangioma. Because this lymphangioma encased the mediastinal structures, the phrenic nerve was injured during its removal. Follow-up MRI (figure 1B) demonstrated a residual soft tissue mass in the right paratracheal, subcarinal and paracardiac regions, as well as an elevated right hemidiaphragm related to phrenic nerve injury. Lymphangiomas are congenital malformations of the lymphatic system and represent embryological remnants of lymphatic tissue. Ninety per cent manifest before the end of the second year of life. The majority of these lesions appear in the cervical area and only 2–3% are associated with an intrathoracic extension. An isolated mediastinal lymphangioma without a cervical component is a very uncommon occurrence.¹ Most thoracic lymphangiomas remain asymptomatic and only cause symptoms once they compress vital structures.²

Learning points

- ▶ Lymphangiomas are congenital malformations of the lymphatic system.
- ▶ A mediastinal lymphangioma can cause airway compression.
- ▶ Phrenic nerve injury can occur during removal of a lymphangioma.

Contributors PG and RG were involved in the diagnosis and management of the patient. SA was responsible for the radiological interpretation, and PS for the histological diagnosis.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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