LETTERS TO JCP

# A case of solitary pulmonary lymphangioma

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Solitary pulmonary lymphangiomas are rare benign lesions thought to result from the development of abnormally proliferating lymphatic vessels. This report describes a case of solitary pulmonary lymphangioma resected under video assisted thoracoscopic surgery and diagnosed using histological and immunohistochemical investigations.

A 53 year old woman was admitted to our hospital for further examination of a nodule in the right lower lung field discovered on chest radiography. Physical examination and laboratory studies revealed no abnormalities. Computed tomography (CT) revealed a lobulated nodule with a well defined border in the lower lobe of the right lung. The CT number of the nodule was approximately 10 Hounsfield units, and contrast enhancement on CT was not seen, consistent with a cyst (fig 1). On magnetic resonance imaging (MRI), the lobulated nodule was of low signal intensity on T1 weighted images and very high signal intensity on T2 weighted images. The nodule displayed ring-like enhancement after injection of Gd-DTPA (diethylene triamine penta-acetic acid). Based on these findings, an intrapulmonary cystic lesion such as a bronchogenic cyst was strongly suspected, although hamartoma was also considered because of the lobulated configuration of the lesion and very high signal intensity on T2 weighed MRI images. Bronchoscopic transbronchial lung biopsy did not help make a definitive preoperative diagnosis for this lesion. We chose to resect the lesion because of concern that it could be malignant. The operation was performed via a left lateral approach under general anaesthesia. The nodule was identified in the lower lobe as a cystic lesion with a well defined margin and removed in a wedge fashion under video assisted thoracoscopic surgery (VATS). The postoperative course was uneventful and the patient was discharged nine days after surgery. One year later, no recurrence had been detected.

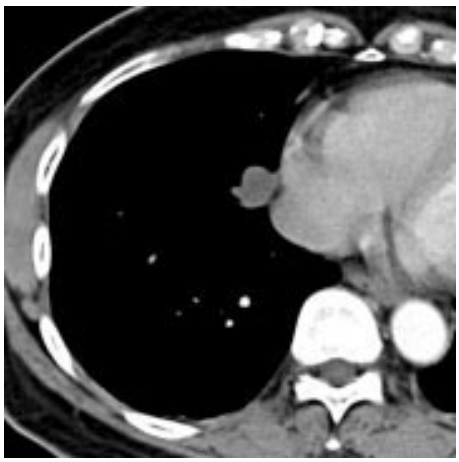

**Figure 1** Computed tomography (CT) revealed a lobulated nodule with a well defined border in the right lower lobe. The CT number of the nodule was approximately 10 Hounsfield units, and contrast enhancement was not seen.

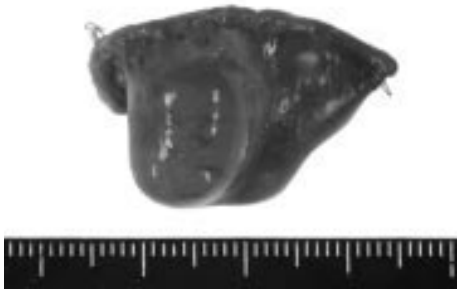

**Figure 2** Macroscopically, the lesion was 1.5 × 1.3 × 1.6 cm. The cut surface revealed a cystic lesion containing sanguinous fluid.

> "We chose to resect the lesion because of concern that it could be malignant"

Macroscopically, the lesion was 1.5 × 1.3 × 1.6 cm, and the cut surface revealed a cystic lesion containing sanguinous fluid (fig 2). Histopathologically, it was a unilocular cystic lesion, with a single layer of flat cells lining the inner cyst wall. The cyst wall was made up of fibrous connective tissue containing collagen fibres, elastic fibres, and smooth muscle cells. Several foci of dense lymphoid infiltrations were present (fig 3). At the periphery of the main cyst, small cystic areas were seen forming anastomosing cavernous spaces. No other cystic change was noted in the lung parenchyma. Immunohistochemically, lining cells were positive for factor VIII related protein (fig 4A), CD31, and proliferating cell nuclear antigen. These cells were negative for CD34, epithelial membrane antigen, AE1/AE3 (fig 4B), p53, and Ki-67. Smooth muscle cells in the cyst wall showed a positive reaction for α smooth muscle actin and a negative reaction for HMB-45 and melan-A. Based on these macroscopic and microscopic features, the lesion was diagnosed as localised solitary lymphangioma.

## DISCUSSION

Lymphangioma is a benign lesion that can involve any organ within the body. The lesion is believed to result from abnormal development of the lymphatic system. Intrathoracic lymphangiomas have occasionally been known to arise within the mediastinum, but pulmonary lymphangiomas are extremely rare. Since Wada *et al* first reported a case of lymphangioma of the lung,[1] nine cases, including our present one, have been documented as pulmonary lymphangioma in the English

**Abbreviations:** CT, computed tomography; MRI, magnetic resonance imaging; VATS, video assisted thoracoscopic surgery

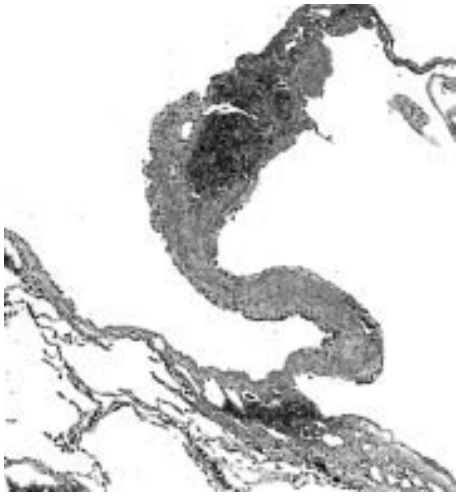

**Figure 3** The cyst wall contained smooth muscle and a lymphoid cell infiltration (haematoxylin and eosin stain; original magnification, ×50).

literature.[1–4] Most adult patients with a pulmonary lymphangioma experience no symptoms and patients usually present with localised cystic lesions for which limited resection is performed.

> "Because lymphangioma and lymphangiectasis are both thought to result from prolonged and excessive development of the lymphatic network in the early embryonic period, differentiating between these two entities is sometimes difficult"

Preoperative diagnosis of pulmonary lymphangioma based on diagnostic imaging is difficult. CT images reveal a cystic or multilobulated mass with either a well defined border and homogeneous density in the localised type, or diffuse involvement.[2 3] A recent paper indicated that MRI findings for thoracic lymphangioma in adults may include a cystic component isointense or hypointense to muscle on T1 weighted images and hyperintense to fat on T2 weighted images.[4] Although findings in both CT and MRI are very helpful in delineating the extent of the disease, the diagnosis cannot be made on the basis of radiological studies alone.

The final diagnosis was confirmed postoperatively under pathological examination. In the diagnosis of lymphangioma, it is necessary to rule out cystic epithelial tumour, congenital lymphangiectasis, and lymphangioleiomyomatosis.[3] Because lymphangioma and lymphangiectasis are both thought to result from prolonged and excessive development of the lymphatic network in the early embryonic period,[1] differentiating between these two entities is sometimes difficult. Recent immunohistochemical data confirmed a common origin of these separate diseases.[5] In our case, the cyst lining cells and the endothelial cells of the capillaries were positive for factor VIII related protein. The cyst lining cells were negative for keratin, in contrast to the alveolar lumina lined by reactive type II and type I pneumocytes, which showed strong reactivity. These results confirmed that it was a vascular lesion and not a malignant neoplasm.

**Take home messages**

- We report a case of pulmonary lymphangioma resected under video assisted thoracoscopic surgery and diagnosed using histological and immunohistochemical investigations
- Although preoperative computed tomography and magnetic resonance imaging can delineate the extent of the lesion, they cannot rule out the presence of a tumour so that surgical resection is the safest option

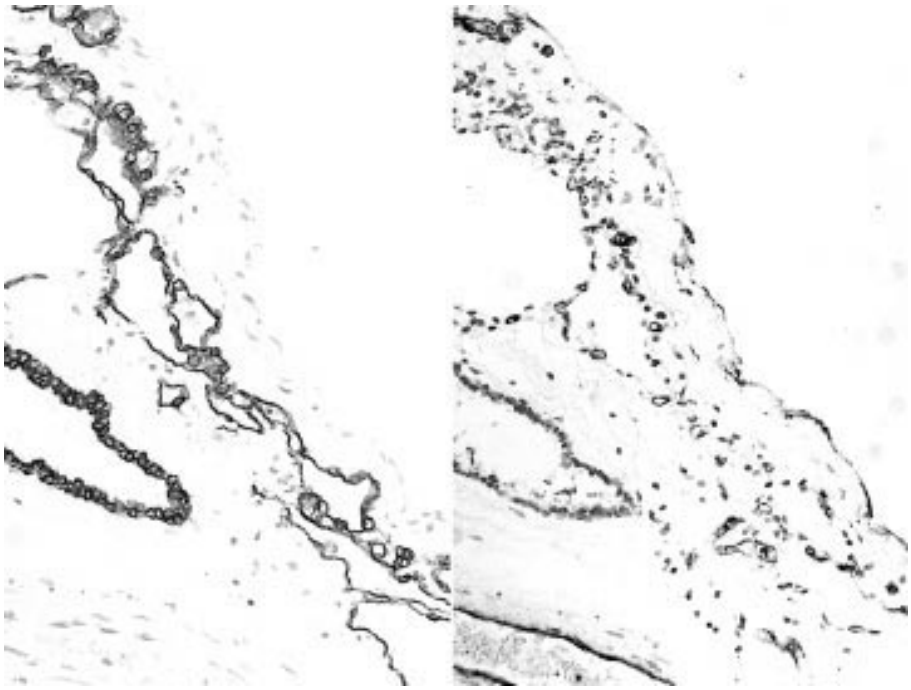

**Figure 4** (A) The cyst lining cells and endothelial cells of the capillaries showed a thin layer of positivity for factor VIII related protein (F-VIII immunohistochemistry; original magnification, ×50). (B) The cyst lining cells were negative for keratin, in contrast to the alveolar lumina, which was lined by reactive type II pneumocytes and type I pneumocytes that showed strong reactivity (AE1/AE3 immunohistochemistry; original magnification, ×50).

In summary, an asymptomatic pulmonary nodule was resected under VATS and a diagnosis of pulmonary lymphangioma was confirmed by immunohistochemical examination. Although preoperative CT and MRI can delineate the extent of the lesion, surgical resection should be considered, because radiological findings cannot rule out the presence of a tumour.

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