

Short reports

Primary leiomyoma of the pulmonary vein

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Abstract

The first case of a primary leiomyoma in a pulmonary vein in an adult is presented. Although rare, leiomyoma and leiomyosarcoma of the lung and great vessels should be included in the differential diagnosis of intrathoracic space occupying lesions.

Leiomyomas are common benign neoplasms of soft tissue in adults, especially in women, the most frequent site being the uterus. Other sites are less common and a venous primary site is very rare.¹ Of those reported, the commonest site is the inferior vena cava.² The only case of leiomyoma occurring in a pulmonary vein was described in a 3 year old child.³ We report the first case of a leiomyoma occurring in a pulmonary vein in an adult.

Case report

A 54 year old woman presented with a six month history of cough, occasional haemoptysis and mild dyspnoea on exertion. She complained of right sided chest discomfort of three months' duration but denied weight loss, anorexia, or bone pains. Twenty five years previously she had undergone hysterectomy for menorrhagia. She smoked 20 cigarettes per day.

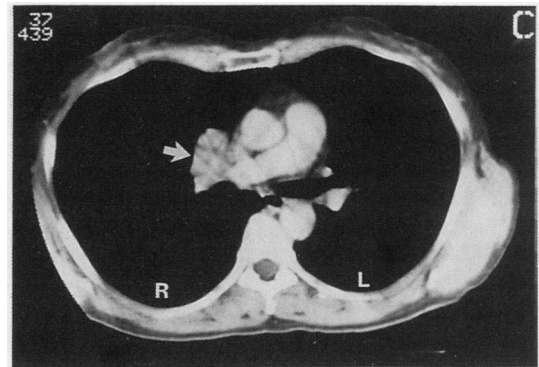


Figure 1 Computed tomogram of the thorax (mediastinal setting, with contrast), showing a right sided soft tissue mass in the aortopulmonary window (arrow).

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The results of clinical examination and routine haematological and biochemical tests were normal. Chest radiography showed a dense right hilum consistent with a soft tissue mass, which was larger than on a radiograph taken six months previously; a chest radiograph taken six years earlier had been normal. Computed tomography of the chest confirmed the presence of a right hilar mass (fig 1). No evidence of malignancy or metastatic disease was found on computed tomograms of the abdomen or brain or at fiberoptic bronchoscopy. After a preliminary cervical mediastinoscopy a right thoracotomy was performed; there was a hard mass in the right superior pulmonary vein. Although frozen section showed no evidence of malignancy in the mass, a right upper lobectomy was performed in view of the clinical findings. The patient's recovery was uneventful and she was discharged home well.

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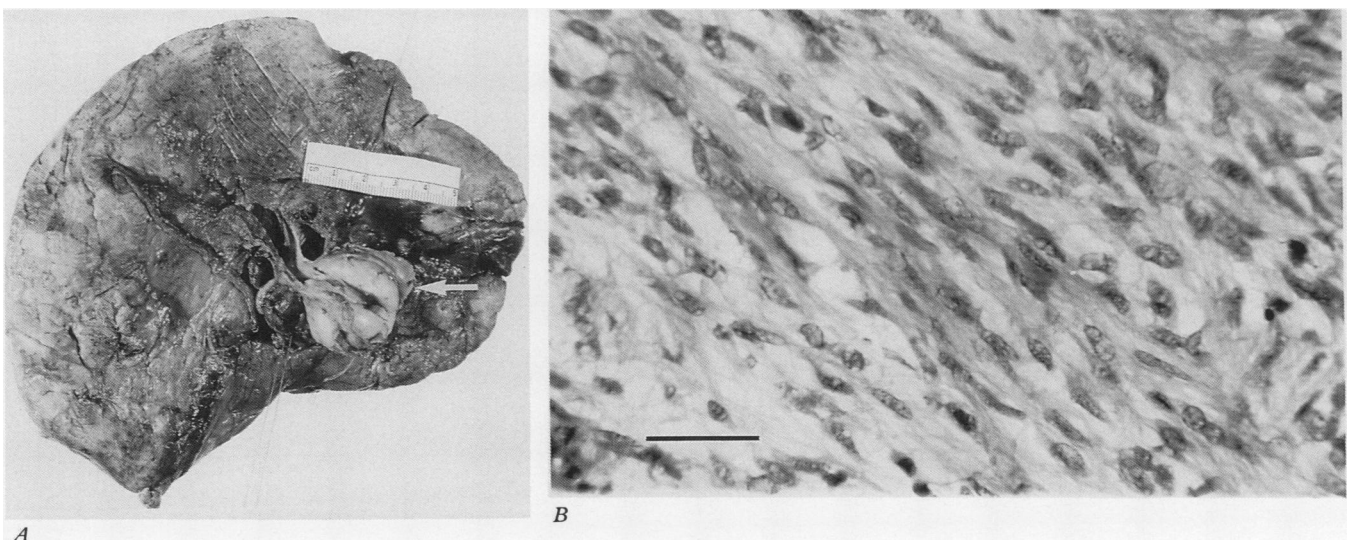


Figure 2 A—Macroscopic view of the resected lung lobe, showing the leiomyoma in the opened pulmonary vein (arrow). B—Histological appearance of the tumour, showing bundles of smooth muscle fibres containing uniform spindle shaped nuclei. There is no mitotic activity. (Haematoxylin and eosin; bar = 40 µm.)

Pathological findings

A hard, well circumscribed tumour, measuring about 3.5 × 2.5 × 2.5 cm (fig 2A), arose from the superior pulmonary vein. The tumour had a bosselated surface appearance and a homogeneous white aspect when cut. Histological examination showed that the mass contained haphazard bundles of smooth muscle cells with uniform spindle nuclei in the absence of any mitotic activity (fig 2B). No necrosis or haemorrhage was observed; considerable areas of fibrosis, however, were seen throughout the tumour with areas of chronic inflammation. Immunocytochemical studies showed that the tumour cells were positive for desmin and vimentin but negative for CAM 5.2, confirming that the tumour was of muscle origin. A diagnosis of leiomyoma was made; none of the lymph nodes contained tumour.

Discussion

This is the first reported case of a primary leiomyoma in a pulmonary vein in an adult. McAllister and Fenoglio⁴ in their comprehensive review of primary tumours of large veins recorded only 16 cases of leiomyoma (the first published in 1869) and commented that about 16% of all smooth muscle tumours of veins were benign. The only case these authors cite of a leiomyoma in a pulmonary vein occurred in a 3 year old child.³ In common with other tumours,^{5,6} this child's lesion was classified as a "leiomyofibroma," emphasising the presence of extensive fibrosis, a morphological feature seen in our case. We speculate that persistent trauma by blood turbulence engenders chronic inflammation and subsequent fibrosis.

The differential diagnosis of a leiomyoma presenting in a vein in a female patient includes intravenous leiomyomatosis and benign

metastasising leiomyoma. Both arise from uterine lesions. In the former case smooth muscle fibres are seen to infiltrate into uterine veins and, in some cases, retrograde extension may occur and the infiltration may extend to thoracic organs.⁷ In the latter case solitary or multiple cytologically benign leiomyomas are seen in retroperitoneal lymph nodes or in the lungs.⁸ The patient's operation for menorrhagia prompted a thorough review of the hysterectomy specimen for fibroids or intravenous leiomyomatosis, but these were not present. We concluded that her pulmonary vein leiomyoma was unrelated and therefore a primary manifestation.

Primary and secondary leiomyomas are rare and should be included in the differential diagnosis of intrathoracic space occupying lesions.

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