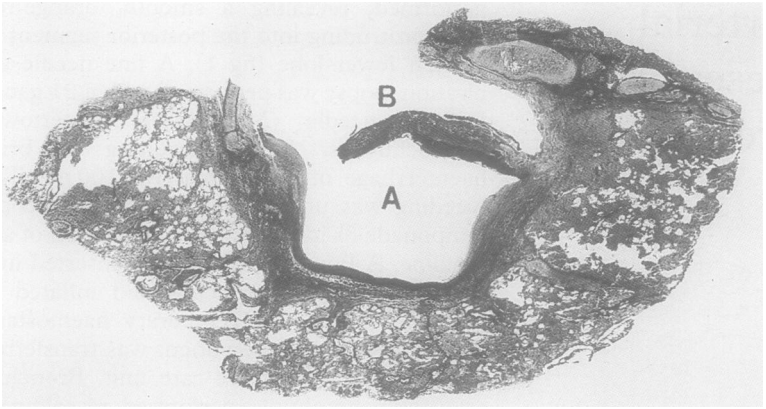

Figure 2 Paraffin embedded cross section of aneurysm at point of rupture into bronchial lumen; A = aneurysmal lumen, B = bronchial lumen. Elastic stain, magnification × 3.

parenchyma and fail to impinge directly on a bronchus, it is not surprising that few have been detected by fibreoptic bronchoscopy.

We have found only one previous description of the bronchoscopic appearance of such a lesion. St Christov *et al*[3] described a patient with a 2·5–3 cm reddish, tumourlike mass filling the left main bronchus. This lesion bled spontaneously after a few seconds and resulted in the patient's death. Pathological examination revealed a bulbous aneurysm encasing the bronchus in a cuff-like fashion, with medial necrosis of the vessel wall. Erosion into the bronchi must almost certainly occur in many proximal aneurysms, given the frequency of fatal haemoptysis, but we found no other report of bronchoscopic detection of an aneurysm.

The current case is notable for its mimicry of a submucosal tumour. The bland, orange-red bronchoscopic appearance belied the vascular nature of the lesion, unlike the more telltale, plexiform red pattern seen in well vascularised tumours such as a central carcinoid. Although transection of normal pulmonary vessels has been reported as a complication of transbronchial biopsy,[4] such vessels are unlikely to protrude into the airway lumen.

Previous reports[5] have cautioned against percutaneous biopsy of peripheral coin lesions without prior radiological examination, such as rapid sequence CT scan with contrast, to rule out aneurysms. This case suggests that similar caution needs to be extended to selected bronchoscopic procedures, and that the bronchoscopist would do well to add this lesion to the list of endobronchial masses in which a biopsy is to be assiduously avoided.

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Preoperative diagnosis of a pulmonary artery sarcoma

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Abstract

A pulmonary artery sarcoma was diagnosed preoperatively by magnetic resonance imaging enhanced with gadolinium and confirmed by percutaneous computed tomographic guided needle biopsy. Accurate preoperative diagnosis allowed planned curative surgery with removal of the right ventricular outflow tract and reconstructive surgery using a cryopreserved homograft.

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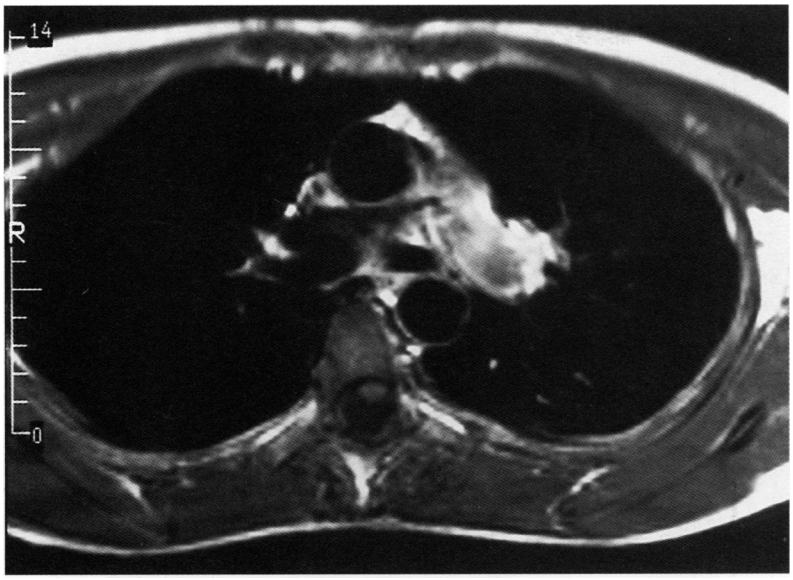
Keywords: pulmonary artery sarcoma, diagnosis.

Pulmonary artery sarcoma is a rare tumour of the cardiovascular system, only about 120 cases having been reported.[1] The tumour is most often mistaken for pulmonary embolism,[2] and the surgical treatment may have to be improvised if the diagnosis is established only upon exploration of the pulmonary artery, which may lead to incomplete resection, palliative surgery, or imperfect reconstruction.[3] The advantage of a precise preoperative diagnosis is obvious. We report a case of pulmonary artery sarcoma which was diagnosed preoperatively using two readily available radiological techniques that allowed planned and appropriate surgery.

Case report

A 37 year old man was admitted for suspected pulmonary embolism. He had voluntarily lost 7 kg in weight over the previous three months and presented with a dry cough and intermittent fleeting pain in the left chest. A chest radiograph showed reduced vascularisation of the left lung and a ventilation perfusion scan showed absence of perfusion to the left lung. The patient was not in distress and physical examination and blood gas analysis breathing air were normal. An ascending venogram showed no thrombi in the leg veins and vena cava. The pulmonary artery pressure was normal. A computed tomographic (CT) scan of the thorax confirmed the presence of an obstructed



Gadolinium enhancement of the left pulmonary artery mass shown with magnetic resonance imaging.

left pulmonary artery and showed a mass or thrombus in the left lateral aspect of the pulmonary truncus. Three densities in the left lung were consistent with pulmonary infarction but metastases could not be excluded. Magnetic resonance imaging with gadolinium showed uptake by the pulmonary artery mass, indicating neovascularisation, compatible with a malignant process (figure). A percutaneous CT guided needle biopsy of the left pulmonary artery was performed and showed a malignant mesenchymal tumour consistent with a sarcoma. The slides were reviewed by a second pathologist who diagnosed a pleomorphic spindle cell sarcoma. A CT scan of the abdomen showed no primary tumour and the diagnosis of primary pulmonary artery sarcoma was made.

A cryopreserved homograft was ordered (National Heart Hospital, London). Seventy two hours after the percutaneous biopsy the patient was operated on through a median sternotomy with extracorporeal circulation. The heart was emptied and left beating during the entire procedure. The pulmonary artery was palpated. The left pulmonary artery was hardened and no softer area was found on dissecting into the hilum. A longitudinal incision on the median aspect of the pulmonary artery showed a gelatinous tumour on the left anterior cusp of the pulmonary valve, extending to the lateral aspect of the pulmonary truncus and totally obstructing the left pulmonary artery. The right ventricular outflow tract was transected at the level of the pulmonary valve and the right pulmonary artery was transected behind the aorta. A left pneumonectomy was performed. Reconstruction was achieved using the 26 mm cryopreserved pulmonary homograft with the left pulmonary artery branch oversewn.

The postoperative course was uncomplicated and the patient was discharged on the tenth postoperative day. Echocardiography revealed a normally functioning homograft and no pulmonary regurgitation. Adjuvant radiation therapy was started six weeks later.

Final pathological analysis confirmed the diagnosis of sarcoma of the pulmonary truncus totally obstructing the left pulmonary artery. The pulmonary masses seen on the CT scan were zones of pulmonary infarction.

Discussion

Only a few cases of sarcoma of the pulmonary artery have been diagnosed before resection. Fitzgerald[4] described a needle biopsy of the pulmonary artery, but the histological findings were not diagnostic. Experience with this procedure is not extensive; however, the risk of bleeding from the thin walled pulmonary artery is minimal if it is totally occluded. A more recent report has described a diagnostic procedure similar to ours and definite preoperative diagnosis.[5]

Magnetic resonance imaging (MRI) with gadolinium enhancement has been advocated for the diagnosis of malignancies and its value has previously been reported in a case of pulmonary artery sarcoma.[6] It is increasingly being used for the evaluation of perfusion and metabolic activity of tissues. It has been limited by lack of a good contrast material to show neovascularisation, but this is now possible with gadolinium enhancement. We suggest that the combination of a perfusion lung scan, MRI, and CT guided needle biopsy provides a means of reaching the correct diagnosis preoperatively.

Pulmonary artery sarcoma has been treated surgically by several approaches including thoracotomy with pneumonectomy and "end-arterectomy" of the tumour plug,[7] or resection of the pulmonary artery using extracorporeal circulation and reconstruction with a valved conduit or a homograft.[8] In view of the extension of the tumour in our case, we chose the latter approach which allowed wide excision of the tumour filled pulmonary artery. Even so, microscopic infiltration of the right section was found. For this reason, radiotherapy was given using multiple opposed fields to a CT constructed mediastinal target. There is no consensus on the efficacy of radiotherapy in this rare tumour, but it has been used in similar cases.

A precise preoperative diagnosis enables the best surgical approach and technique to be chosen, and allows time for ordering of prostheses or homografts which are not always readily available.

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