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## APPENDIX. CONFERENCE DISCUSSION

**Dr P. Ciriaco (Milan, Italy):** I understand that preoperative diagnosis is an important issue in pulmonary artery sarcoma. So my first question deals with the preoperative work-up. We have found that endobronchial ultrasound is a very useful tool for TBNA, especially to locate pulmonary vessels, and specifically the pulmonary artery. Do you think that this procedure could have a role in the differential diagnosis between pulmonary artery sarcoma and thromboembolic disease?

My second question deals with fibrosis that can be induced by chemotherapy in patients who had neoadjuvant chemotherapy. Did you find any technical differences in patients who underwent neoadjuvant chemotherapy?

**Dr Mussot:** First, I don't think that EBUS or another ultrasonography technique for adults could be of very much help, but of course if you see an invasion outside of the artery, it is a good clue to imagine it is not CTEPH. Of course, you shouldn't do a biopsy. We had a patient referred after a mediastinoscopy who had proven sarcoma because in fact it was the artery that had been biopsied. EBUS is a new technique we haven't used up to now, but we'll try it in the future.

Concerning neoadjuvant chemotherapy, we haven't noticed any difference in the difficulty of the surgery. It's always a difficult surgery, and this adds no extra major difficulty.

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## EDITORIAL COMMENT

# Pulmonary artery sarcoma

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Angiosarcomas carry a poor prognosis, and those of the pulmonary artery are no exception. Pulmonary artery sarcomas generally derive from the endothelial cells of the pulmonary artery, and tend to spread within the pulmonary vasculature. Rarely, the pulmonary parenchyma itself is invaded (usually only in the later stages), and, unlike angiosarcomas elsewhere, pulmonary artery sarcomas do not have a high rate of lymph node and systemic metastases. This is perhaps because of the particular anatomy of the lungs—the pulmonary artery being a low-pressure system carrying venous blood and the pulmonary parenchyma having a dual blood supply (the bronchial arteries).

Pulmonary artery sarcomas generally remain asymptomatic until a pulmonary vascular obstruction occurs, and, unless routine screening tests such as X-rays show an abnormality, it is generally the patient presenting with pulmonary hypertension that signals the presence of the intravascular tumour.

Treatment is rarely curative. Since the tumour arises from the endothelium, surgical resection is by endarterectomy and concentrates on restoring the blood flow to the affected areas of

the lung to relieve pulmonary hypertension and to restore oxygenation. To date, neither chemotherapy nor radiotherapy has provided significant palliation. A surgical cure of this bad malignancy occurs therefore largely by chance, in a very unusual case when an early diagnosis of unilateral disease allows a pneumonectomy, since pulmonary endarterectomy rarely removes all microscopic cells.

In this paper by Mussot *et al.* [1], 31 patients underwent operations for pulmonary artery sarcomas over a 14-year period by a surgical team experienced in pulmonary endarterectomy and lung transplantation. Such a concentration of cases of this rare disease is unusual, and results from their referral base in patients with thromboembolic disease of the lungs. Apart from the autopsy series, the only others reporting such a series have also had pulmonary endarterectomy programmes; there have for example been previous reports from Mayer's group [2], the group in Cambridge [3] and our group in San Diego [4]. Mayer's report, as mentioned in this paper, contained a surprisingly small number of sarcomas in their otherwise large practice; perhaps this represents a difference

in referral patterns. However, we have now encountered 45 pulmonary artery sarcomas in a series of close to 3000 pulmonary endarterectomy operations. As in the paper presented here, in our series, the diagnosis was generally known or suspected before the operation. Pulmonary artery sarcomas usually present with a different radiographic appearance from thromboembolic disease on angiography, generally being more central and bulky in character, and often apparently unilateral.

As in the experience presented here, in our series, the operation was commonly performed for palliative reasons, and life expectancy considerably extended; the operation was rarely curative. This is because the disease is always bilateral if the patient presents with pulmonary hypertension, and even in the absence of pulmonary hypertension in an apparently unilateral case, bilateral seeding has probably occurred by the time the patient presents. For that reason, we have not performed a pneumonectomy—one would have to be sure that the tumour is confined to one lung to favour this approach, and this is almost never the case.

The review presented here is retrospective, and therefore has the disadvantages of all retrospective studies. It is not clear why patients with lung parenchymal sarcomas were excluded, as certainly the tumours in these cases originated in the pulmonary vasculature. Similarly, it is unclear why patients with 'inoperable primary pulmonary artery sarcomas in a fully palliative situation' were excluded, since obviously these patients are part of a spectrum of the generally palliative nature of surgical treatment for this disease. It would have been interesting to know the subsequent survival of these patients and why they were considered inoperable.

However, the paper is a helpful summary of the way these patients generally present, and the surgical treatment by removing all visible tumour by endarterectomy. As stated above,

what is perhaps unusual is the treatment of patients by pneumonectomy; usually this is avoided as the disease is almost always bilateral at the time of diagnosis, even if microscopically so. It would have been useful to know the specific follow-up in these patients. It must be emphasized that treatment of this disease by pneumonectomy is not generally recommended. Adjuvant therapy with chemotherapy or radiotherapy is advisable in nearly every case, to extend palliation, though to date this has not been curative. It appears that not all the patients in this series underwent such treatment. Five-year survival was dismal at 22%. This conforms to the experience of others.

Newer and specific methods of tumour treatment after endarterectomy may, in the future, improve the prognosis of this bad disease. This includes DNA analysis of the specific tumour for genetic mutations that may direct chemotherapy, and growing the tumour in immunodeficient mice, which then allows targeted testing of the patient's own tumour with newer specific drugs. There are several of these (including vascular disrupting agents), which are now showing promise for angiosarcomas.

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