

Paraneoplastic thrombus or relapse of a pulmonary artery sarcoma?

A 34-year-old man with a previous history of left pneumectomy with curative intent after diagnosis of intimal sarcoma arising from the left pulmonary artery was referred for increasing dyspnoea, and pain at the right shoulder. The patient was free of tumour relapse at nearly 5 years follow-up.

A contrast-enhanced multidetector CT (MDCT) shows a filling defect within the main pulmonary artery (figure 1A). Axial fusion image of integrated [^{18}F]fluorodeoxyglucose positron emission tomography (^{18}F FDG PET)/CT confirmed an increased uptake at the same level and additionally at the right scapula, highly suggestive of a metastasis (figure 1B). To relieve the patient's symptoms, and improve his quality of life and his short-term prognosis, a palliative operation was carried out. The main and the right pulmonary arteries were completely resected and successfully reconstructed using a polytetrafluoroethylene (PTFE) vascular prosthesis (Impra Flex, 19 mm) with

supravalvular anastomosis, and radiotherapy of the osteolytic lesion at the right scapula was performed.

Intraoperatively, a solid whitish-grey intravascular mass with a maximum diameter of 3.0 cm, was identified (figure 1C) and histology confirmed a relapse of a pulmonary artery sarcoma showing abundant malignant spindle cells with high cellularity, frequent mitoses and nuclear pleomorphism (figure 1D).

DISCUSSION

Pulmonary artery sarcoma is a rare malignancy with a poor prognosis, arising from the mesenchymal cells of the intima of the pulmonary artery.

Invasion of the pulmonary artery by angiosarcoma or other tumour is part of the differential diagnosis of thromboembolic disease.^{1 2}

MDCT findings can be specific in patients with advanced disease, particularly in the case of extraluminal tumour extension; other findings favouring the diagnosis of pulmonary artery sarcoma include a low-attenuation filling defect occupying the entire luminal diameter of the proximal or main pulmonary artery and expansion of the involved arteries.³ However, these

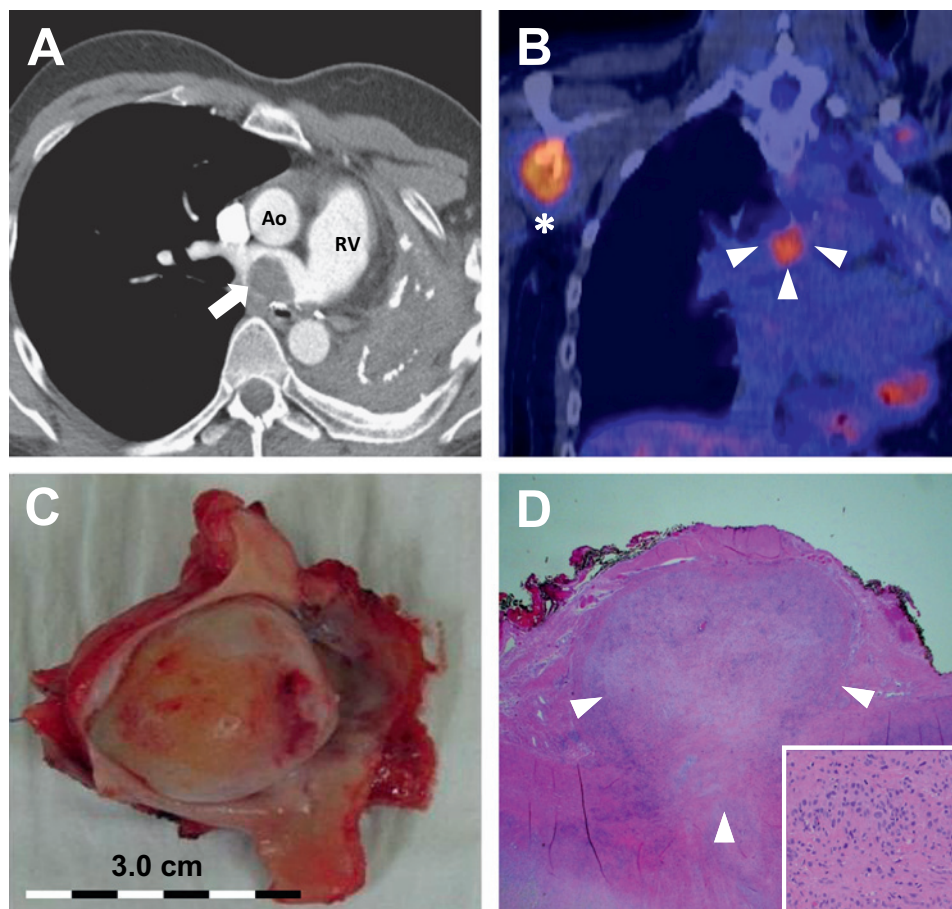


Figure 1 (A) A chest contrast-enhanced multidetector CT image shows an intraluminal filling defect in the main pulmonary artery (arrow) suspicious of a relapse of pulmonary artery sarcoma (maximum diameter of 3.0 cm). The right ventricle is moderately dilated (4.1 cm). Ao, aorta; RV, right ventricle. (B) Axial fusion image of integrated [^{18}F]fluorodeoxyglucose positron emission tomography (^{18}F FDG PET)/CT shows increased uptake (standardised uptake value (SUV) of 7.0) in the main pulmonary artery (arrowheads). Note the presence of an osteolytic lesion with high uptake of ^{18}F FDG at the level (diameter 3.0 cm) of the right shoulder (asterisk) with an SUV measured at 8.4 compatible with a metastasis. (C) Photograph of specimen showing a whitish-grey, intravascular mass (maximal diameter of 3.0 cm) within the main pulmonary artery. (D) Photomicrograph of the intravascular mass showing abundant spindle cells with high cellularity, frequent mitoses, nuclear pleomorphism and a high nuclear/cytoplasmic ratio; these findings are suggestive of moderately differentiated sarcoma. The tumour cells were positive for CD31 and vimentin, and negative for pan-cytokeratin, CD34 and S100.

MDCT findings can be non-specific in cases of extensive pulmonary thromboembolism. On the other hand, the application of ¹⁸F-FDG PET shows negative FDG uptake in the case of blood thrombi, whereas a malignant tumour shows clearly positive FDG uptake.^{4 5} Therefore, for early and accurate differential diagnosis,⁵ a combined PET/CT scan is found to be useful and should be considered mandatory, since the two conditions need a different therapeutic approach.

LEARNING POINTS

- ▶ The differential diagnosis of obstruction of the pulmonary artery is relatively difficult; invasion of the pulmonary artery by angiosarcoma or other tumour should be considered.^{1 2}
- ▶ MDCT findings can be specific for angiosarcoma in patients with advanced disease, particularly in the case of extraluminal tumour extension.³
- ▶ The finding of a low-attenuation filling defect at MDCT occupying the entire luminal diameter of the proximal or main pulmonary artery can be non-specific in cases of extensive pulmonary thromboembolism.
- ▶ A combined CT/PET should be considered mandatory, since the two conditions need a different therapeutic approach. ¹⁸F-FDG PET shows negative FDG uptake in the

case of blood thrombi, whereas a malignant tumour shows clearly positive FDG uptake.^{4 5}

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