Pulmonary artery sarcoma mimicking a pulmonary embolism

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An 82-year-old woman with no previous history of heart or lung diseases presented with exertional dyspnoea and episodes of an acute shortness of breath over the preceding month. Contrast-enhanced CT, performed for the assessment of suspected pulmonary embolism, revealed a large filling defect in the main pulmonary artery (Panels A and B). A more detailed anatomic study by T1-weighted double inversion recovery fast spin echo cardiac-MRI sequences confirmed the existence of a large mass in the pulmonary trunk (Panel C). Further tissue characterization assessment by T1-weighted images acquired after the administration of gadolinium contrast demonstrated the absence of delayed hyperenhancement (Panel D), suggesting the presence of an avascular structure (i.e., thrombus). Based on the cardiac-MRI findings and despite the rare location and the absence of distal embolization, which are atypical findings for pulmonary embolism, the patient was initially anticoagulated for presumed pulmonary thromboembolism. Because of sudden and complete pulmonary outflow tract occlusion (Panels E and F; Supplementary data online, Videos), the patient suffered haemodynamic deterioration and died from cardiogenic shock. Macroscopic (Panels G and H) and microscopic (Panel I) examination revealed the presence of an intimal sarcoma of the pulmonary artery.

Intrapulmonary sarcoma is a rare neoplasm that can appear morphologically similar to pulmonary thromboembolism. Although some radiological findings such as occupation of the entire luminal diameter of the main pulmonary artery or an extraluminal expansion of the defect beyond the artery wall may help in the diagnosis of malignancy rather than pulmonary embolism, tissue characterization images by cardiac-MRI may be limited. While the absence of contrast-enhancement has been shown to be useful for the diagnosis of intracardiac thrombus, poorly vascularized neoplasms can manifest minimal contrast uptake and occasionally be difficult to distinguish from thrombus based on first-pass or conventional delayed-enhancement tissue characterization methods. To obtain a more accurate tissue characterization, probably, additional delayed enhancement cardiac magnetic resonance imaging by increasing the inversion time (i.e., 600 ms) might have been useful, showing non-vascularized structures (thrombus) in black, in contrast to those vascularized (grey appearance).

Supplementary data are available at European Heart Journal – Cardiovascular Imaging online.