

Intimal Sarcoma of the Pulmonary Artery as an Embolic Cause of Sudden Death

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A 70-year-old woman was transferred after presenting to another hospital with exertional dyspnea that had persisted for 3 months. Enhanced computed tomography (CT) revealed thrombus-like protuberant structures in the main pulmonary artery (PA) and scattered distal obstructions in the bilateral PA (Figure A). Perfusion lung scintigraphy demonstrated multiple defects in both lung fields (Figure B), and echography showed marked pulmonary hypertension with a transtricuspid pressure gradient of 80 mmHg, with no deep vein thrombosis. Transesophageal echocardiography revealed mobile structures with a “halo”-like sign (i.e., hyperechoic core surrounded by hypoechoic structures) in the main PA (Figure C, Supplementary Movie), indicative of thrombi or tumors. The laboratory workup revealed slightly elevated D-dimer ($1.6\mu\text{g/mL}$) and BNP (179pg/mL). An integrated FDG-PET/CT scan demonstrated a “hot” spot in the PA (Figure D). A 2-week course of rivaroxaban did not alter the size of the structures or lower the pulmonary hypertension, thus providing further indications that the mobile structures were tumors, not thrombi. An operation for tumor excision was consequently planned; however, 32 days after her admission the patient

suddenly developed fatal cardiopulmonary arrest. Macroscopic autopsy confirmed the presence of jelly-like whitish emboli occluding both PA (Figure E), suggesting the tumors in the main PA were an embolic source. Histopathology revealed that the structures consisted of CD31- and αSMA -negative atypical spindle cells, providing the diagnosis of intimal sarcoma (Figure F).

PA intimal sarcoma is a poorly differentiated mesenchymal malignant tumor that can mimic pulmonary thromboembolism. Delayed diagnosis and treatment may be fatal; however, distinguishing tumors from thrombi is challenging using only images. Thus, when the tumor-indicating halo-like sign is detected by echocardiography or pulmonary angiography, an aspiration biopsy or open chest surgery should be performed as early as possible. Interventional treatments, including the insertion of extracorporeal membrane oxygenation, should be considered.

Supplementary Files

Supplementary Movie. The images of transesophageal echocardiography.

Please find supplementary file(s);
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