Case Reports

Pulmonary Artery Sarcoma Mimicking Pulmonary Embolism

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Primary sarcomas that arise from major blood vessels are exceedingly rare, and some of the published cases have been autopsy reports. Most patients are adults. We report a case of pulmonary artery sarcoma in a 77-year-old man who presented with acute onset of dyspnea. Magnetic resonance imaging of the chest revealed a large mass within the pulmonary trunk and its main branches. Because massive pulmonary embolism was suspected, both anticoagulant and thrombolytic therapies were initiated. The patient responded poorly to these therapies, which then necessitated resection of both the mass and the pulmonary valve. A bioprosthetic porcine valve replaced the native valve, and we reconstructed the right ventricular outflow tract with a Dacron patch. Histopathologic examination revealed a high-grade sarcoma with focal myogenic and chondrogenic differentiation. The patient tolerated the procedure well and was discharged from the hospital on postoperative day 7. He was subsequently treated with chemotherapy and radiation and continued to show no evidence of disease.

The diagnosis of pulmonary artery sarcoma should be suspected in patients who present with manifestations of pulmonary embolism, especially when there is no evidence of deep venous thrombosis and poor response to anticoagulant therapy. Multimodal therapy can provide prolonged survival. ***(Tex Heart Inst J 2014;41(5):515-7)***

Pulmonary artery (PA) sarcoma is a rare tumor that often is mistaken for PA thrombus,[1,2] leading to underdiagnosis or delayed diagnosis. We describe a case in which PA sarcoma was found in a patient who presented with acute onset of dyspnea. The patient was misdiagnosed and was treated for pulmonary embolism.

Case Report

A 77-year-old man was transferred from another hospital for treatment of a saddle PA embolus. The patient had reported acute onset of shortness of breath. Computed tomographic (CT) angiography of the chest had revealed a large filling defect within the main PA, which extended into both right and left PAs (Fig. 1). He had been treated with thrombolytic and anticoagulant agents without clinical improvement. After the patient's transfer to our hospital for further evaluation and treatment, the decision was made to proceed with PA embolectomy. Physical examination revealed a heart rate of 95 beats/min, temperature of 36.6 °C, blood pressure of 122/60 mmHg, and respiratory rate of 18 breaths/min. Intraoperative transesophageal echocardiography showed a mass arising from the right ventricular outflow tract (RVOT) and extending into the main PA (Fig. 2), a left ventricular ejection fraction of 0.60 to 0.65, severe tricuspid regurgitation, and a right ventricular systolic pressure of 98 mmHg.

After initiating cardiopulmonary bypass, we incised the main PA and inspected the mass. The mass was fleshy and densely adherent to the leaflets of the pulmonary valve; it extended proximally into the RVOT and distally into both PAs. We then extended the arteriotomy into these areas. We divided the ascending aorta to provide better exposure of the right PA. En bloc resection of the mass and the pulmonary valve was achieved via a technique similar to that of PA endarterectomy. We replaced the valve with a bioprosthetic porcine valve and reconstructed the RVOT with a Dacron patch.

After an uneventful postoperative course, the patient was discharged from the hospital on postoperative day 7. The histopathologic report showed a high-grade sarcoma with focal myogenic and chondrogenic differentiation (Fig. 3). Two months later, baseline CT angiography of the chest showed a residual mass adjacent to the left PA

(Fig. 4A). After 2 cycles of single-agent chemotherapy with doxorubicin at 75 mg/m^2 as a 3-day infusion, the patient's tumor increased in size but showed development of central necrosis, which suggested a beneficial response. After 2 additional cycles, the tumor had decreased in size markedly, so chemotherapy was continued. At the end of 6 cycles, the patient had no definite evidence of residual tumor. However, there was still a residual hypodense thickening along the edge of the PA, so the patient underwent proton-beam therapy and received a total of 66 cobalt-60 Gy equivalents administered in 30 fractions to the areas of initial tumor involvement, including the mediastinal lymph nodes. The patient remained free of disease 2½ years later, as indicated by post-radiation changes on his CT scan (Fig. 4B).

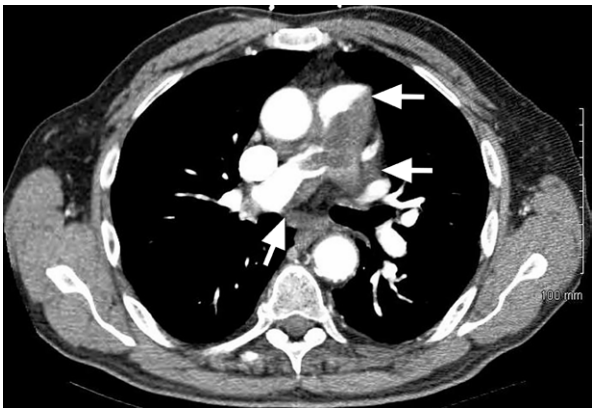

Fig. 1 *Computed tomogram of the chest shows the tumor mass within the pulmonary trunk and its extensions into the pulmonary artery main branches (arrows).*

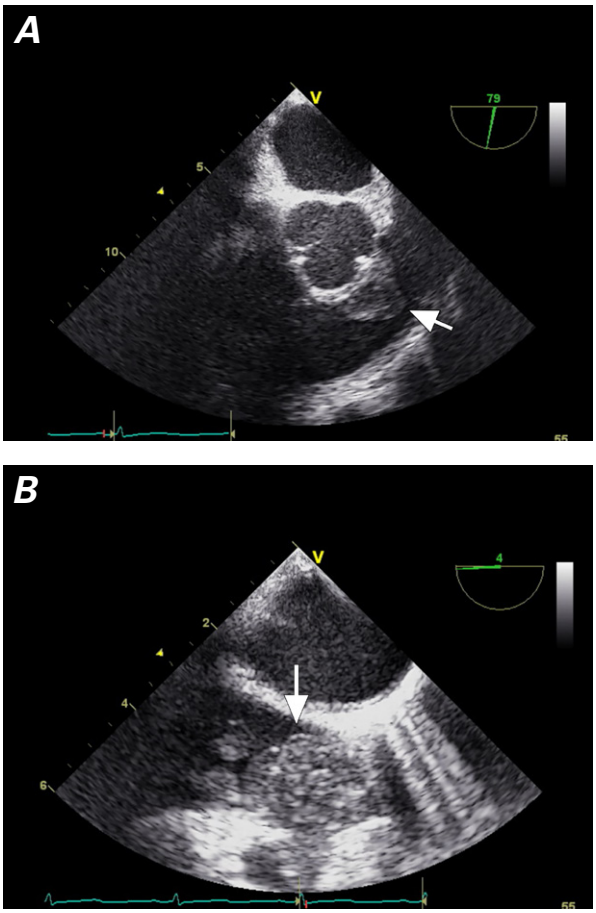

Fig. 2 *Transesophageal echocardiograms show an echodense structure **A**) at the root (arrow) of the pulmonary trunk and **B**) in the right ventricular outflow tract (arrow).*

Supplemental motion images are available for Figures 2A and 2B.

Discussion

Sarcoma of the large PAs is of 2 types: intimal and mural. Intimal sarcomas have a predominant intraluminal polypoid growth pattern, they often obstruct the lumen of the vessel of origin, and they usually show fibroblastic or myofibroblastic differentiation. Mural sarcomas are exceedingly rare and are distinct from intimal sarcomas: they are classified separately, according to histologic subtype, as soft-tissue sarcomas (leiomyosarcomas).[3] In our patient, the mass was of the intimal type, fleshy and displaying the characteristic polypoid pattern upon histologic examination. Moreover, the mass was extending into, and obstructing, the lumina of the main PA and its branches.

This tumor is highly malignant and carries a very poor prognosis. The usual survival period is 12 to 18 months after the onset of the symptoms.[4] This disease occurs mainly in adults and shows predominance in women, with a mean age at diagnosis of 48 years.[5] The

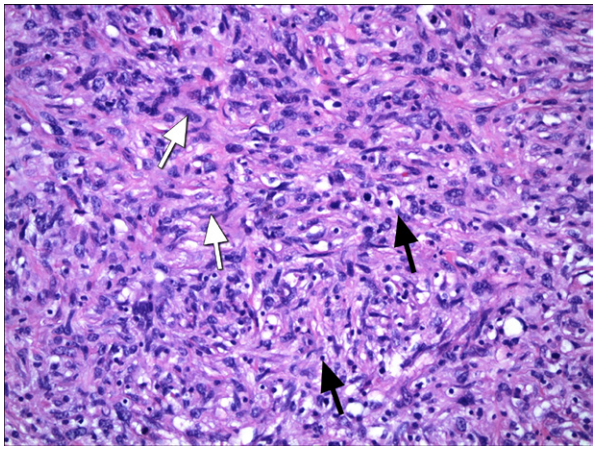

Fig. 3 *Photomicrograph of the resected specimen shows a proliferation of spindle cells in a vague storiform pattern (white arrows). The cells have pleomorphic nuclei with readily identifiable mitotic figures (including atypical forms), and an eosinophilic cytoplasm. Present as well in the collagenous stroma are scattered inflammatory cells, predominantly lymphocytes (black arrows) (H & E, orig. ×200).*

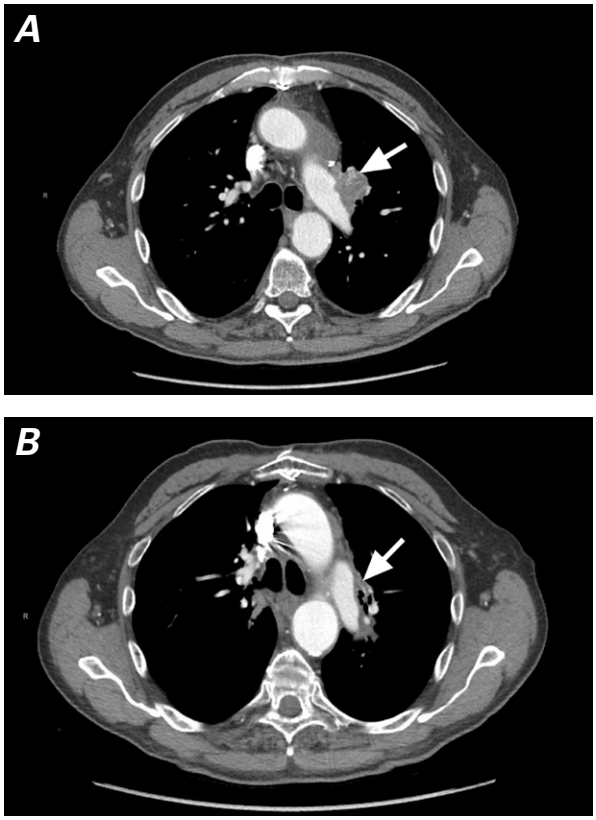

Fig. 4 *Computed tomograms of the chest show the tumor (arrows)* ***A****) before and* ***B****) after postoperative chemotherapy and radiation therapy.*

diagnosis is difficult and is often delayed by the gradual development of symptoms of PA obstruction and right-sided heart failure. Patients usually have a long asymptomatic course. At the time of the writing of this report, our patient had survived 36 months after surgery and was still doing well. This excellent response to chemotherapy and radiotherapy was evident both clinically and radiologically (Fig. 4B).

Although PA sarcoma is rare, its true prevalence is underestimated: many cases are misdiagnosed as pulmonary thromboembolism and are treated inappropriately. Both diseases appear as intraluminal filling defects in the PA system on contrast-enhanced CTs, and radiologic differentiation between PA sarcoma and PA thromboembolism can be difficult. However, the improvement of imaging techniques in recent decades has enabled distinction between these entities before surgery. Yi and colleagues[6] have reported that CT findings favor the diagnosis of PA sarcoma when they include a low-attenuation filling defect that occupies the entire luminal diameter of the proximal or main PA, usually accompanied by expansion of the involved arteries and extraluminal extension of the tumor. In addition, PA sarcoma is more likely to have a heterogeneous histologic appearance, characterized by areas of necrosis, hemorrhage, and ossification.[6] Pulmonary artery sarcoma often occurs unilaterally, in contrast with the more common bilateral involvement of PAs in thromboembolic disease.

This sarcoma is rare, but it should be suspected in patients who present with clinical manifestations of pulmonary thromboembolism, especially when there is no evidence of deep vein thrombosis and when the response to thrombolytic or anticoagulant therapy has been poor. Adjuvant chemotherapy and radiation therapy are recommended for the eradication of any residual tumor foci. We emphasize the value of multimodal treatment of PA sarcoma in our patient, who was still alive 36 months postoperatively.

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