Malignant vascular tumors are highly uncommon, with angiosarcomas as the most prevalent of these tumors, constituting <1% of all sarcomas. The presence of angiosarcoma in the lung is usually a consequence of metastasis from a primary site, such as the skin, subcutaneous tissue, or breast. Primary pulmonary artery sarcoma has been described in a limited number of reports. In 1988, Nonomura and colleagues [1] reviewed 110 cases of pulmonary artery sarcomas that included only four angiosarcomas. Since then, fewer than a dozen of cases have been reported in the English literature. Surgical resection remains the therapy of choice for pulmonary artery sarcoma and is believed to give the best chance for survival. However, even with surgical resection, the mean survival is approximately only 12 months [1]. Information on the benefit of adjuvant therapy is limited. Some believe that chemoradiation has no effect on long-term survival [2], whereas others have suggested that it can benefit patients and prolong survival.

A 39-year-old woman presented to an outside hospital with shortness of breath and atypical chest pain. Her initial differential diagnosis was pulmonary embolism versus acute coronary syndrome. Cardiac biomarkers were negative, but a computed tomographic (CT) scan of the chest revealed near occlusion of the main and left branch pulmonary arteries. She received thrombolytic therapy with no improvement in her symptoms. Subsequently, she was referred to our institution where re-evaluation of her CT scan showed occlusion with a lobulated mass (Fig 1) rather than a typical pulmonary embolus. Through a median sternotomy with hypothermic cardiopulmonary bypass and a brief period of circulatory arrest, a mass in the main pulmonary artery (PA) distal to the pulmonary valve resulted in near complete obstruction of the left PA and almost complete obstruction of the right PA. The mass was excised. The PA wall was resected en bloc with the tumor mass and was reconstructed with a bovine pericardial patch. Pathology revealed high-grade spindle-cell sarcoma. Six weeks later, the patient received chemoradiotherapy with two cycles of mitomycin, adriamycin, and cisplatin concurrently with 5,040 cGy radiotherapy in 28 fractions. This was followed 4 weeks later by four cycles of adriamycin and ifosfamide. On follow-up CT scan, 1 year later, a new left lower lobe lung nodule was noted, and the patient underwent video-assisted thoracoscopic wedge excision. Pathology confirmed metastatic PA intimal sarcoma. Five months later, a new pulmonary nodule was found in the superior segment of the left lower lobe. A left lower lobe superior segmentectomy was performed through a left posterolateral thoracotomy for a second recurrence of PA spindle-cell sarcoma. Nine months later, a 1-cm soft tissue mass in the inferior aspect of the left lung hilum presented on CT scan was worrisome for lymph node metastasis. An endobronchial biopsy was negative. Through a redo left thoracotomy, a station 9L mediastinal lymph node was removed. Pathology confirmed her third recurrence. Three months later, after experiencing increasing shortness of breath, a CT scan showed left main bronchial narrowing. On bronchoscopic evaluation, mucosal studding by a malignant appearing process in the left lower lobe was seen. Biopsies confirmed the fourth recurrence. The patient was evaluated for systemic recurrence with a positron emission tomographic scan in combination with a CT scan, with head magnetic resonance imaging, which all confirmed the disease was localized to the chest.

Through a fourth redo left thoracotomy, the tumor mass was present exterior to the left lower lobe and wrapped around the lobar bronchus, extending onto the left mainstem bronchus. There was insufficient margin to perform any form of reconstruction or sleeve reattachment of the left upper lobe. This was complicated with posterior extension into the descending thoracic aorta. To perform an en-bloc resection, we performed an intrapericardial pneumonectomy and a segmental aortic resection with reconstruction using a 20-mm Dacron tube graft (Hemashield woven; Meadox Medical, Wayne, NJ) on left heart bypass. A left serratus anterior muscle flap was used to cover the bronchial stump. Six months later, the CT scan showed a soft tissue mass immediately adjacent to the descending aorta, which had enlarged from the most recent prior study (Fig 2A), with high metabolic activity on positron emission tomographic scan (Fig 2B). Endoscopic transesophageal ultrasound-guided fine needle aspiration cytology confirmed the fifth recurrence of a high-grade spindle-cell sarcoma. At that time, she received 4,500 cGy in 25 fractions of radiation, which partially overlapped with her prior radiation fields. The head magnetic resonance image and positron emission tomographic scan in combination with a CT scan showed no evidence of systemic disease. Through a left thoracoabdominal approach we resected the mass, exenterating the lower portion of the pneumonectomy space and replacing the mid-descending thoracic aorta proximal to the prior graft with another 20-mm Dacron graft (Hemashield woven; Boston Scientific, Natick, MA) using cardiopulmonary bypass and profound hypothermia with circulatory arrest. The chest wall was reconstructed with a Gore-Tex patch (W.L. Gore & Assoc, Flagstaff, AZ). A persistent chylothorax required reoperation and ligation of the thoracic duct. The patient is currently alive without evidence of disease at 5 years from her initial diagnosis and resection, and 11 months from the last resection.
Comment

Primary sarcomas of the pulmonary artery are uncommon and remain a subject of case report. They can be either intimal or mural in origin. Intimal sarcomas arise from pluripotent intimal cells that have fibroblastic or myofibroblastic differentiation. Some cases can demonstrate distinct histologic features and can be further subclassified by immunohistochemical stains into rhabdomyosarcoma, osteogenic sarcoma, or angiosarcoma. In contrast with intimal sarcomas, leiomyosarcomas of the pulmonary artery represent mural sarcomas that originate from the media of the vessel [3]. A limited number of primary pulmonary artery sarcoma cases have been reported in the literature, with approximately only 200 histologically confirmed cases. The disease has been reported previously to be uniformly fatal, with the longest survival time in one series reported to be 3.5 years, despite surgical resection [4]. Surgical resection is believed to provide the best chance of prolonging survival. Generally, it is believed that pulmonary artery sarcoma occurs more frequently in middle-aged (range, 38 to 69 years) patients than in younger patients [5]. Intraluminal growth can lead to a progressive obstruction of the right ventricular outflow tract with a varying obstruction of pulmonary perfusion, thus clinically mimicking pulmonary thromboembolism in the majority of cases. These tumors are characterized by insidious growth, with the tumor usually showing extensive local invasion and hematogenous metastases by the time of presentation. Our patient did present with a lymph node recurrence, which is unusual for most sarcomas, but can be seen in angiosarcomas [6]. We did not perform a mediastinal lymph node dissection at the time of the initial two lung resections. The literature regarding lymph node metastases is very sparse for primary sarcomas of the lung, although thoracic lymph node metastases in metastatic sarcomas can be seen in as much as 20% of patients having pulmonary metastasectomy [7]. Most cases of pulmonary artery sarcoma are diagnosed late in the disease course or at the time of autopsy. Radiologic diagnosis can be difficult, despite technologic advances in imaging techniques [8]. The prognosis of these patients appears to be poor, with most patients dying within months of the initial presentation. To date, there have been no reports of an effective treatment for this condition. The main treatment of choice is radical surgical resection. As palliative treatment, endovascular stent grafting may be an alternative therapeutic option in inoperable cases. Extremity soft tissue sarcomas have been treated by adjuvant radiotherapy, and in the prospective randomized trial by Yang and colleagues [9], there was a statistically significant reduction in local recurrences in patients with either high-grade or low-grade extremity tumors. Overall survival and nonlocal recurrences were nearly identical for patients receiving or not receiving radiation. Neoadjuvant or adjuvant chemotherapy has been described in the literature with overall poor results. Prolonged tumor-free survival is rare and the role of chemoradiotherapy is still undefined. Aggressive and repeated surgical resection and the use of cardiopulmonary bypass to achieve complete resection can be effective in managing these cases. Several previous studies have demonstrated that angiosarcoma can be radiosensitive [10]. Prognosis in pulmonary artery sarcomas is extremely poor if complete resection of the tumor is not feasible, and the median survival without surgical resection is known to be approximately 1.5 months.
References


