Pulmonary Artery Sarcoma

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Figure 1. Chest radiograph revealed a right hilar mass (A), which on contrast-enhanced computed tomography was found to be a large intravascular tumor occluding the right main pulmonary artery with extension into the lumen of the main and left pulmonary arteries (B). A dynamic magnetic resonance image demonstrated the tumor within the main pulmonary artery but without mural invasion (C). Tumor within the main pulmonary artery as visualized intraoperatively after pulmonary arteriotomy (D). Gross histology demonstrated involvement of superior, middle, and inferior lobes (E) with tumor completely occluding the right pulmonary artery lumen (F).

A 34-year-old man with previously treated acute lymphoblastic leukemia presented with chronic cough and hemoptysis. Chest radiograph (Figure 1A) showed a right hilar mass that, on cross-sectional imaging, occluded the right pulmonary artery (PA) and extended into the main and left PA (Figures 1B and 1C). Distal invasion of lung parenchyma suggested malignancy rather than thromboembolic disease (1). Percutaneous lung biopsy confirmed a sarcoma. Cine-magnetic resonance imaging demonstrated that although the right PA was inseparable from the mass, the tumor did not invade the main and left PA (Video 1). Intrapericardial right pneumonectomy was performed using cardiopulmonary bypass via median sternotomy (Video 2). The tumor was extracted from the main and left PA and resected en bloc with the right lung (Figure 1D). Histology revealed a myxofibrosarcoma arising from the right pulmonary artery with diffuse intravascular extension invading all lobes (Figures 1E and 1F). This case highlights the improved accuracy of dynamic imaging over static techniques, particularly when accurate anatomic evaluation plays a critical role in clinical decision-making. Although computed tomography showed bilateral PA involvement precluding surgery, the dynamic magnetic resonance imaging clearly demonstrated the feasibility of curative resection. Surgical resection improves survival in PA sarcoma compared with palliative procedures, with reported median survival of 26.8–36.5 months versus 6.6–11 months, respectively (2–4). Several studies have suggested that adjuvant chemotherapy and/or radiotherapy improves survival (2, 5). This patient received adjuvant therapy and remains recurrence-free at 8 months. ■

The uncompressed videos are accessible from this article's supplementary material page.

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Video 1. Contrast-enhanced cine-magnetic resonance imaging demonstrated tumor extending into the lumen of the main and left pulmonary arteries without invasion. The *arrow* indicates the region where the tumor is free from the arterial wall.

Video 2. Intraoperative video. Pulmonary arteriotomy being performed on cardiopulmonary bypass. Intraoperative visualization of the pulmonary artery sarcoma extending into the main and left pulmonary artery. Not shown owing to length constraints are median sternotomy and institution of cardiopulmonary bypass. As the video progresses, the *arrow* points to the tumor once exposed.

Author disclosures are available with the text of this article at www.atsjournals.org.

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