Pulmonary artery obstruction caused by a rare type of primary cardiac synovial sarcoma

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Video clip is available online.

A 17-year-old male presented to our hospital for recurrent episodes of syncope. A preoperative transthoracic echocardiogram showed a giant mass in the main pulmonary trunk causing obstruction of the pulmonary artery while protruding into both the right and left pulmonary arteries (Figure 1, A). Cardiac magnetic resonance imaging (MRI) confirmed a clear-border giant mass located in the main pulmonary trunk and the 2 pulmonary arteries. The MRI signal was isointense to myocardium on T1-weighted imaging and hyperintense on both T2-weighted and fat-suppression images (Figure 1, B and C). Diffuse, mild enhancement of the tumor

as well as dynamic pulmonary obstruction was noted in MRI contrast-enhanced images (Figure 1, D and E, and Video 1). This patient underwent an operation and the tumor was resected from the pulmonary artery (Figure 1, F). Pathologic examination confirmed a primary cardiac synovial sarcoma with SS18 gene translocation.

Primary cardiac tumors are uncommon conditions (incidence in autopsy series ranges only from 0.001-0.03%), with the occurrence of a malignant neoplasm <25%.¹ Cardiac sarcomas account for the majority of this primary malignancy, which is usually found in the atrium and ventricle.² A primary cardiac synovial sarcoma located in the pulmonary artery is extremely rare. This report highlights the importance of cardiac MRI in confirming the presence of a space-occupying lesion, localizing it, assessing the extent of its involvement, evaluating the functional impact of the lesion, and helping to determine tissue characterization.³

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FIGURE 1. Image of primary cardiac synovial sarcoma (white arrow). A, The echocardiogram shows a giant mass in the main pulmonary trunk that protrudes into both the right and left pulmonary arteries. B and C, T2-weighted and fat suppression sagittal images show a giant mass with clear borders located in the main pulmonary trunk. D and E, Contrast-enhanced magnetic resonance imaging (both sagittal and axial views) demonstrated a diffuse, enhanced tumor located in the main pulmonary trunk and 2 bifurcating pulmonary arteries causing obstruction of the pulmonary artery. F, Photograph of the cardiac synovial sarcoma. RPA, Right pulmonary artery; LPA, left pulmonary artery.