A cryptogenic pulmonary embolism: Left atrial myxoma occluding an atrial septal defect

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Acute pulmonary embolism represents a severe condition associated with significant morbidity and mortality.¹ The main source of pulmonary embolism is represented by deep venous thrombosis. Less frequent causes are right-sided heart chamber clots, neoplastic emboli into the systemic veins, or the association of left atrial (LA) thrombi and atrial septal defects (ASDs). We describe a unique case of acute pulmonary embolism related to the contemporary presence of LA myxoma and ASD, with the mass behaving as an ASD occluder.

Clinical Summary

In November 2005, a 47-year-old man was admitted to our emergency department for acute onset of dyspnea and chest pain. The patient had no history of cardiovascular disease. An acute pulmonary embolism was suspected. The patient underwent a thoracoabdominal contrast-enhanced computed tomography AQ scan evaluation. This showed a thromboembolic fragment partially occluding the right pulmonary artery (Figure 1, A). A huge LA mass (7.5 long $\times$ 4.5 cm wide) was also demonstrated (Figure 1, B). The lower limb venous Doppler examination was negative. A transthoracic echocardiogram (TTE) confirmed a solid mass in the LA, partially projecting into the mitral valve orifice, consistent with the features of an LA myxoma. However, no ASD was detected, even after contrast media injection. The patient was therefore scheduled for urgent operation. The procedure was performed on cardiopulmonary bypass and bicaval cannulation. The technique we use for atrial myxoma is the transeptal approach first described by Piwnica and colleagues,² which allows resection of the tumoral mass with its attachment to the septum. After the right atrium was opened, an ostium secundum ASD, approximately 2.5 cm wide, was easily diagnosed, completely occluded by the tumoral mass (Figure 2). The neoplasm, whose dimensions were those of a chicken egg, was attached to the region of the superior limbus. The LA incision was then completed, and the LA myxoma was extracted in the usual manner. Afterward, a second incision was performed starting from the main pulmonary artery and extending toward the right pulmonary branch. A mass of tissue similar to the myxoma was found in the right pulmonary artery and easily withdrawn. The pulmonary arteriotomy was closed by a simple Prolene 5-0 (Ethicon Inc, Somerville, NJ) running suture, and an autologous pericardial patch was used to repair the ASD. Pathologic evaluation confirmed the myxomatous nature of both atrial and pulmonary masses. The postoperative course was uneventful. The patient was discharged on postoperative day 8. PredischARGE TTE showed no residual lesions or shunts.

Discussion

Pulmonary embolism generally has a right-sided origin; however, a few cases of left-sided sources have been described.³ Obviously, these conditions are always associated with some form of cross-circulation allowing the embolic mass to pass from the left to the right side of the heart. The most frequent left-sided sources of pulmonary embolism reported in the literature are LA thrombi, often the result of mitral valve stenosis or atrial fibrillation. A left-sided origin of an acute pulmonary embolism should always be suspected when TTE

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Figure 1. Computed tomography images showing (A) the thromboembolic mass (arrow) partially occluding the right pulmonary artery branch and (B) the LA mass (arrow), suspected to be a myxoma. No sign of atrial septal defect may be detected.

demonstrates an ASD in the workup of a patient. In these cases, transesophageal echocardiography confirms this finding and often allows visualization of the embolic source in the LA.³

Atrial myxoma represents the most frequent neoplasm of the heart.⁴ The majority of these lesions are located in the LA. Right atrial myxomas are rare; embolism occurs in 10% of the cases, often leading to massive pulmonary artery obstruction.⁵

Figure 2. After the right atrium was opened, a 2.5-cm-wide atrial septal defect was clearly evident. The myxoma is visible underneath.

The singularity of this case consists in a few points. First, to our knowledge this is the first case of pulmonary embolism produced by an LA myxoma. This may be explained by the fact that a right-to-left embolism necessitates an ASD. Because atrial myxomas frequently originate from the interatrial septum, it is easily realized that both conditions may seldom coexist. Another interesting feature was the ineffectiveness of echocardiography assessment in preoperatively demonstrating the presence of the ASD. In fact, the tumor mass acted as a perfectly shaped occluder. The pressure gradient between the 2 chambers allowed the myxoma to stay firmly stacked in the septum orifice. This case demonstrates, once again, that in pulmonary embolism the quest for the embolic source is sometimes a difficult task. The intraoperative findings allowed a precise reconstruction of the pathologic events.

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