Leiomyosarcoma of the left atrium mimicking a left atrial myxoma

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Unlike benign neoplasms, primary cardiac malignancies are rarely intracavitary and, when located in the left atrium, may mimic myxoma on cardiac imaging. Cardiac surgeons should be alerted to ask for prompt surgical pathologic study in any case of intracavitary cardiac mass.

Clinical Summary

A 21-year-old woman was admitted to the hospital with pulmonary edema and shock. Transthoracic echocardiography showed a large (4 × 7-cm) mobile mass within the left atrium obstructing the mitral valve orifice during diastole (Figure 1). The diagnosis of left atrial myxoma was put forward, and an emergency operation was performed with the patient on cardiopulmonary bypass. The left atrium was approached via an extended transseptal incision. The left atrium was occupied by a whitish mass, and the tumor appeared entrapped in the mitral orifice without infiltration of the leaflets or the annulus. The mass originated from the roof of the left atrium, between the pulmonary veins. The tumor and the underlying endocardium were smoothly dissected from the myocardium of the atrial wall, which appeared soft and without evidence of invasion. The operation was performed without complications.

At gross examination, the mass appeared whitish and ovoid. It measured 7 × 3 × 3.5 cm and weighed 51 g. The surface was slightly irregular and wrinkled, hard to cut with a knife. Accessory clusters of small round masses were also present. Microscopic examination of histologic sections revealed a storiform proliferation with pleomorphic cells enmeshed in a myxoid extracellular matrix with a high mitotic index (Figure 2, A and B). Necrotic foci and lymphocytic-plasmacellular infiltrates were also observed. At immunohistochemistry, the neoplastic cells appeared vimentin, desmin, and α -smooth muscle actin positive (Figure 2, C and D) and myogenin negative, in keeping with a pleomorphic leiomyosarcoma.

The patient was treated with anthracycline-based chemotherapy (doxorubicin plus fosfamide plus cisplatin alternated with fosfamide plus cisplatin every 15 days for 2 months). Eleven months later, the patient underwent reoperation for suspected recurrence of the tumor on the anterior mediastinum, but the removed mass turned out to be a reactive hyperplasia of the thymus secondary to chemotherapy. At 24 months from surgical resection, the patient is well, without evidence of local recurrence or metastases on computed axial tomography.

Discussion

Primary leiomyosarcoma of the left atrium is extremely rare and has a poor prognosis. Symptoms of left atrial leiomyosarcoma are generally related to obstruction of the left atrial cavity, pulmonary veins, or mitral valve orifice.^{1,2} The extension of the tumor to the pulmonary veins often makes pneumonectomy necessary to obtain radical resection.³

In our case, the clinical presentation and the echocardiographic features strongly suggested a left atrial myxoma, but the rough surface and the unusual origin from the roof of the left atrium argued against a benign tumor. Thorough histologic and immunohis-

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Figure 1. Preoperative transthoracic echocardiography. A, A mass is visible occupying the left atrial cavity during systole. B, The mass is moving downward during diastole to occupy the mitral valve orifice.

Figure 2. Surgical pathologic investigation of the resected mass. A and B, At histologic examination, storiform proliferation of pleomorphic cells with frequent mitoses was seen (stain, hematoxylin and eosin; original magnification, 15 $\times$ and 160 $\times$, respectively). C and D, Positive immunohistochemistry findings for α -smooth muscle cell and desmin (original magnification, 32 $\times$).

tochemical investigations allowed to disclose the malignant features of leiomyosarcoma. As in the case reported by Evans and Haw,³ the tumor was attached to the endocardial surface without infiltration of the atrial wall, thus making its complete removal feasible. Although we believed that the tumor was removed completely and, in the early postoperative period, no metastases could be demonstrated, we decided to treat the patient with anthracycline-based chemotherapy to destroy or prevent repetitions. As reported by Pessotto and associates,⁴ our case supports the hypothesis that multimodal treatment of cardiac leiomyosarcomas may prolong survival with a good quality of life.¹

From this experience, we have learned that it may be difficult to distinguish a noninfiltrating intracavitary malignancy from a left atrial myxoma by using echocardiography. Skilled histopathologic examination is mandatory in any apparently benign cardiac mass to exclude malignancy⁵ and, if so, to allow

early chemotherapeutic therapy, which can be successful, as in this case.

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Ventricular lateral wall rupture after myocardial infarction detected by means of multislice computed tomography

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Ventricular septum ruptures are known and severe complications after myocardial infarction with a corresponding high mortality. Ventricular free wall ruptures are less common because these patients often die before sufficient diagnostics can be performed.¹ If an examination is possible, diagnosis is most often performed by means of Doppler echocardiography.² However, detection of a ventricular free wall rupture by means of multislice computed tomography (MSCT) has not yet been published and is presented here.

Clinical Summary

A 51-year-old patient with hemodynamic deterioration after acute myocardial infarction was referred to our department in June 2005. The case history revealed aortic valve replacement with a Lillehei Custer valve in 1972, as well as aortocoronary bypass graftings in

1992 and 2001. The patient was admitted first to a cardiology unit with the diagnosis of acute myocardial infarction. A coronarangiography showed a stenosis of the right coronary artery (RCA), as well as occlusions of the left anterior descending coronary artery (LAD) and the circumflex artery. Also occluded were all bypass grafts. Stent implantation was then performed successfully in the LAD and RCA. However, hemodynamics deteriorated a day after the intervention, and the patient was referred to our department. Transthoracic echocardiography showed severely reduced cardiac function and an approximately 1.5-cm pericardial effusion. Repeated coronarangiography and ventriculography revealed open stents in the LAD and RCA and the suspicion of a ventricular wall perforation. Thus MSCT was performed. In this computed tomographic scan, 2 large ruptures (8×8 mm and 7×6 mm) in the lateral wall of the left ventricle were found (Figure 1). The patient probably survived up to that point because of pericardial adhesions, which might have prevented further massive perforation into the thoracic cavity. However, contrast enhancement in the pericardial wall and contrast flow in the pericardial cavity partly suggested perforation. Thus an emergency operation with the use of extracorporeal circulation was performed. Intraoperatively, we found 2 ruptures in the lateral myocardial wall, as well as blood in the pericardial cavity. Because of the freshly infarcted myocardial tissue, a direct closure of the ventricular defects was not possible. Therefore a polytetrafluoroethylene patch* was adapted to the lateral wall and secured with surgical glue. Because of the preop-

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