with a bronchospastic component. He experienced suppression of recalcitrant inducible ventricular tachycardia during electrophysiologic testing and clinical suppression of recurrent syncope after initiation of sotalol therapy. However, initiation of the racemic mixture of dl-sotalol was associated with worsening of his asthmatic condition. After switching from dl-sotalol to the d-isomer alone, there was improved pulmonary function and persistent clinical improvement with respect to ventricular arrhythmias and the absence of recurrent syncope.

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REFERENCES

We report a case of angiosarcoma of the heart, manifested as a continuous murmur. Right coronary arteriography disclosed a paracardiac mass with fistulas from the coronary vessel to the right atrium. Histologic study revealed this to be an angiosarcoma with sinusoidal pattern. To our knowledge, this is the first case of this kind of cardiac tumor presenting as a fistula from a coronary artery to the right atrium.

(Chest 1992; 102:1629-30)

Primary malignant tumors of the heart are rare, and angiosarcomas are the most commonly reported histologic type.1 Until 1986, at least 139 cases of cardiac angiosarcoma were reported, with a very low percentage of early diagnosis.2

We report the case of a patient with right atrial angiosarcoma involving the right coronary artery that produced signs and symptoms of left-to-right heart fistula, a finding not previously reported in the literature.

CASE REPORT

A 45-year-old woman began having chest pain one month prior to hospital admission. At that time the patient presented with atrial flutter and mild pericardial effusion in the cross-sectional echocardiogram. With the initial diagnosis of myopericarditis, the patient was treated at home with salicylic acid and angiotensin.

One month later, the patient reappeared with pleuritic chest pain, and a continuous murmur was heard over the right sternal border. Chest roentgenogram disclosed an alveolar infiltrate in the right middle lobe. A dynamic axial computed tomographic scan revealed a mediastinal mass involving the right atrium and middle lobe of the right lung, as well as left subpleural metastases (Fig 1, arrow). One week later, the patient developed increasing dyspnea and chest pain, peripheral edema, and petechiae in lower extremities and was referred to our hospital. On hospital admission, the physical examination revealed tachypnea (30 breaths/min), normal systemic arterial pressure (130/80 mm Hg), and normal jugular venous pressure. There was an intense continuous murmur, with increased systolic component, over the right sternal border. Blood analysis disclosed moderate normocytic anemia and thrombocytopenia. Chest roentgenogram revealed right pleural effusion that was hematic by thoracocentesis. Cross-sectional echocardiography revealed a dilated right atrium with increased thickness in close contact with a mass with a partially organized fluid collection. Doppler analysis showed increased flow velocity within the right atrium and turbulence within this collection. Right heart catheterization revealed a large paracardiac mass invading the right pulmonary pedicle and a metastasis in the right lung (arrow).

FIGURE 1. Computed tomography of the heart showing the right paracardiac mass invading right pulmonary pedicle and a metastasis in the right lung (arrow).

Right Coronary Artery-Right Atrium Fistula in Primary Angiosarcoma of the Heart*

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Angiosarcoma

There was a step-up in oxygen saturation between the blood samples obtained at superior vena cava ($O_2$ saturation = 62 percent) and right atrial level ($O_2$ saturation = 73 percent) consistent with a small left to right shunt ($Qp/Qs = 1.5:1$).

Right coronary arteriography revealed an ectatic vessel, with a total obstruction in its medial segment, where the contrast flushed into a paracardiac mass, lung fissure, and into the right atrium (Fig 2). The angiographic findings were interpreted as a fistula between right coronary artery and right atrium. There was neovascularization of the mass, coming from an enlarged conal artery and septal branches of left anterior descending coronary artery (Fig 3).

The patient was then diagnosed as having a malignant paracardiac mass, probably vascular; therefore, she underwent surgery. In the operating room the mass was partially resected, and found to be very friable, cavernous in appearance, with invasion of the atrioventricular groove, right coronary artery, right atrial wall, and pericardium, with continuity to the right pulmonary pedicle. After resection of the mass, the atrial wall was substituted by Goretexpatch and a saphenous vein bypass graft was implanted to relieve the right coronary artery obstruction. Pathologic study of the surgical specimen disclosed normal heart muscle fibers wholly infiltrated by a neoplastic proliferation formed by irregular vascular channels in a sinusoidal pattern. Histologic features of the cellular component were diagnostic of angiosarcoma (nuclear pleomorphism, cell hyperplasia with piling and papillae formation). The patient died one month later, and necropsy was not performed.

FIGURE 2. Angiogram of right coronary artery, left anterior oblique projection, showing contrast flushing into the paracardiac mass. See explanation in text.

FIGURE 3. Left coronary artery and its branches, lateral view. See explanation in text.

DISCUSSION

The clinical spectrum of the case we show herein is not different from those previously reported in the literature, but it has a unique feature: the presence of a continuous murmur suggesting the existence of an arteriovenous fistula.

Neovascularization has been reported widely in a variety of cardiac neoplasms, with the appearance of either small or great lacunar dilatations over terminal branches in atrial and ventricular myocardial vessel. Only in one report, Ugarte et al. suggested a left-to-right shunt according to angiographic data of vast neovascularization and oximetric parameters. Nevertheless, to our knowledge, there is no previous report of angiosarcoma of the heart with the whole clinical and angiographic spectrum of a fistula from a coronary artery to the right atrium, as we report herein.

These features oriented the diagnosis in our case to a vascular malignancy, although only the histologic study revealed the definite diagnosis.

We think that the clinical and angiographic findings we describe must be included into the wide spectrum of presentation of angiosarcomas of the heart.

REFERENCES

3 Francisos RA, Gay RH, Ah-Tye P. Vascular hamartoma of the heart in a child. Am Heart J 1970; 76:676-82