Cutaneous Pigmentation and Palpitations: A Sign of Familial Atrial Myxoma?

Sir,

Cardiac myxomas are rare tumours that constitute half of the primary tumours of the heart. About 7-10% of myxomas are familial and transmitted as an autosomal dominant trait. In contrast to the sporadic types of cardiac myxomas, 75% of which occur in the left atrium with a mean age of 56 years, familial myxomas occur in younger patients (mean age of 25 years) and can be accompanied by spotty skin pigmentation.

A 30-year-old man with a one-year history of cutaneous maculopapular lesions (Figure 1), evident on his foot and back, was referred by his dermatologist in May 2005 to a cardiologist because of palpitations. His vital signs were stable except for pulse rate of 110 beats per minute. Blood pressure was 120/70 mmHg. Hemoglobin level and hematocrit were normal and ESR was 50 mm after one hour. Cardiac auscultation revealed opening snap and a diastolic rumble. Trans-thoracic echocardiography (TTE) revealed a large pedunculated irregular mass (8x3.5 cm) with attachment to the interatrial septum with a narrow base and protruding to the left ventricle through the mitral valve and producing functional stenosis (Figure 2).

The patient underwent cardiac surgery. Median sternotomy was performed and after cardiopulmonary bypass and cannulation of the inferior vena cava and superior vena cava and exploration of left atrium, a large mass (8 x 3.6 cm) with a pedicle of about 5 mm, attached to the interatrial septum, was completely excised along with a surrounding border of healthy tissue. Other cardiac chambers were normal. Pathologic examination was compatible with myxoma. The patient was discharged 3 days later. Postoperative TTE, 2 weeks later, showed no residual mass. Periodic TTE was performed every 2 months. No recurrence has been detected to-date and skin lesions had almost completely subsided.

In 1985, Carney described for the first time, the complex of cardiac myxoma, spotty skin pigmentation and endocrine overactivity. Cardiac myxomas comprise approximately 50% of the tumours of the heart; two-third of patients are females. These tumours are usually in the atrium, with the site of attachment in the region of the limbus of the fossa ovalis. It may present with manifestations relating to the obstruction of the outlet of cardiac valves and symptoms of embolism. Extra-cardiac manifestations are rare. Failure to recognize extra-cardiac manifestations, as in this patient, delays treatment, which may lead to tumour enlargement.

REFERENCES