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IMAGES IN CARDIOLOGY

Echocardiographic Findings in Carcinoid Syndrome

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A 57-year-old Caucasian female was referred to our clinic due to hypertension, flushing, and diarrhea. She had a medical history of hypertension and paroxysmal atrial fibrillation. On physical examination, the patient had a heart rate of 70 bpm and a respiratory rate of 12 breaths/min. Her temperature was 37°C and her blood pressure was 120/80 mmHg. Cardiac examination revealed a left parasternal holosystolic murmur, and a palpable right ventricular heave. Lung auscultation was unremarkable. From the initial biochemical exam she had no specific abnormalities. The ECG showed sinus rhythm, negative T-waves in leads III, V_{1-5}. The transthoracic echocardiography study revealed a left ventricle with normal size and normal systolic function and dilatation of the left atrium, whereas the right cardiac chambers were dilated with thickened, immobile leaflets of the tricuspid and pulmonic valve, leading to malcoaptation and severe tricuspid and pulmonic regurgitation (Fig. 1-5). The clinical and echocardiographic findings raised the suspicion of carcinoid heart disease. Abdominal computed tomography (CT) demonstrated hepatic metastases and the patient was treated with chemotherapy and with the somatostatin analog octreotide.

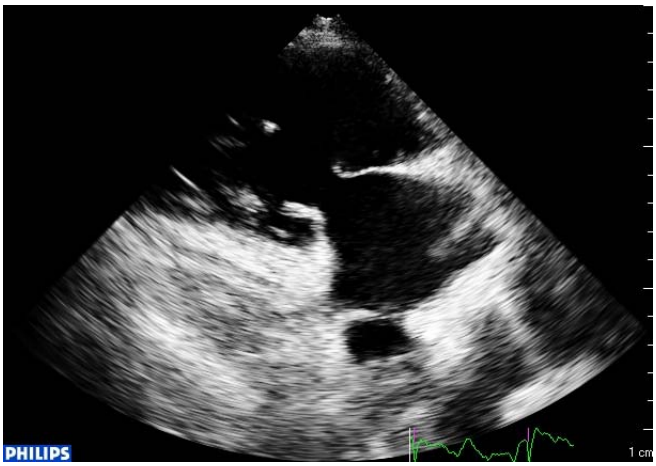

Figure 1. Right ventricular inflow view in systole showing thickened, immobile and retracted anterior and septal leaflets of tricuspid valve**.**

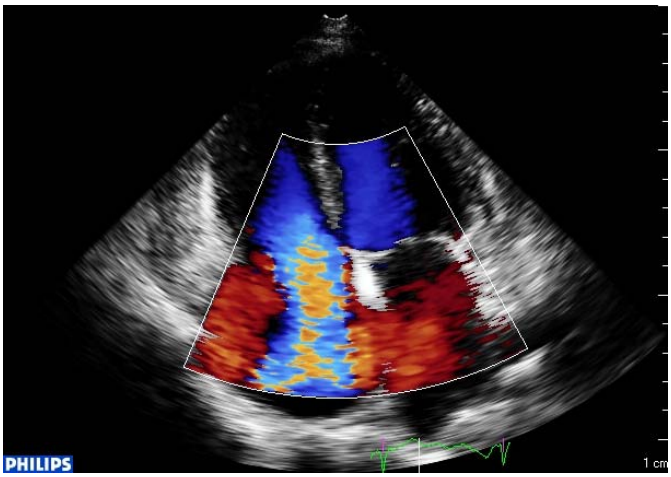

Figure 2. Apical four-chamber view: color Doppler demonstrates severe tricuspid valve regurgitation.

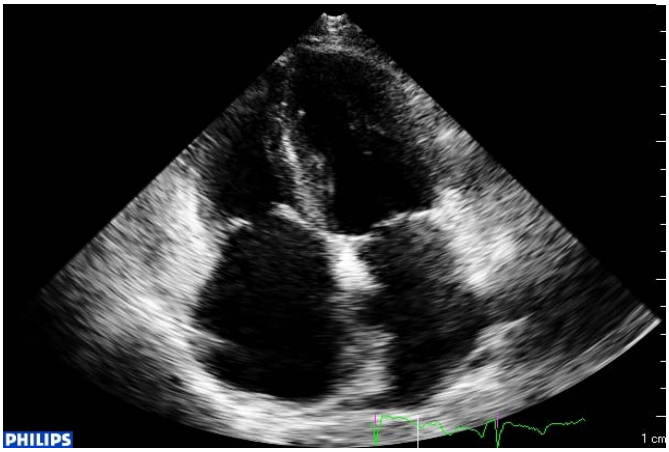

Figure 3. Apical 4-chamber view in systole: opened and retracted tricuspid valve (left); mitral valve is closed (right)

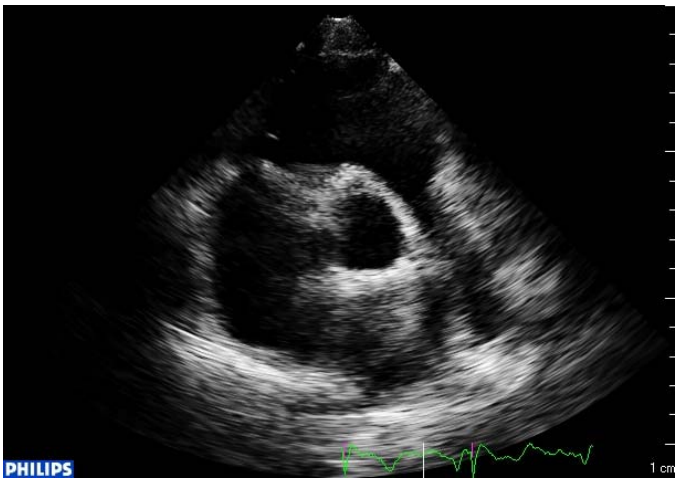

Figure 4. Short axis view in diastole depicting a fixed and immobile pulmonic valve**.**

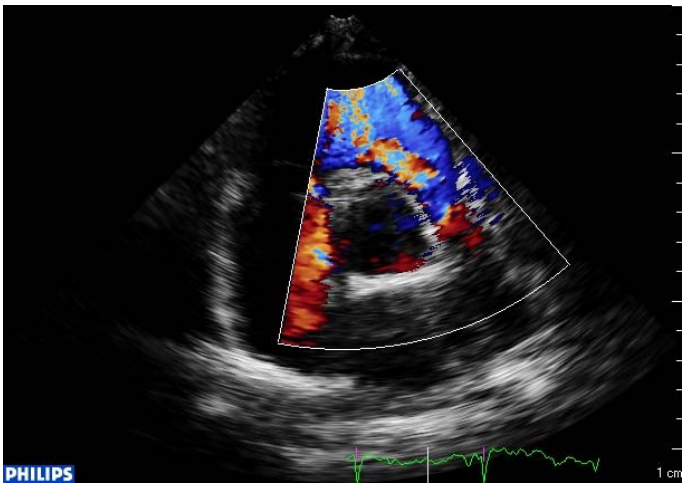

Figure 5. Short axis view in diastole. Color Doppler demonstrates severe pulmonic regurgitation.

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Carcinoid tumors are uncommon malignancies that arise from enterochromaffin cells typically located in the gastrointestinal tract or lungs.[1] These tumors may secrete large amounts of vasoactive substances, including 5-hydroxytryptamine and prostaglandins, which in turn cause various clinical manifestations such as flushing, diarrhea, and bronchospasm. Carcinoid syndrome occurs in <10% of patients and is caused by tumor secretion of hormonal mediators (serotonin, somatostatin, gastrin). Its manifestations include facial flushing, edema of the head and neck, abdominal cramps and diarrhea, bronchopspasm, cardiac lesions. Its treatment is resection of hepatic metastases, the somatostatin analog octreotide and chemotherapy.[2] Cardiac lesions are found in 50-60% of patients with carcinoid syndrome, usually between 18-24 months after diagnosis is established.[3] Carcinoid heart disease results from the layering of plaque-like material over the tricuspid valve, right ventricular endocardium and pulmonic valve and presents with right heart failure and tricuspid and pulmonic regurgitation. Carcinoid involvement of the heart is a late manifestation of metastatic disease. Cardiac surgery for valve replacement remains the mainstay therapy for valvular heart disease caused by carcinoid tumors.[4,5]

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