Images in cardiovascular medicine
A malignant peripheral nerve sheath tumor presenting as an intrapericardial mass

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A 24-year old white man was admitted to our Department with a history of progressive shortness of breath and fatigue. The patient had been in good health until 1 month before admission.

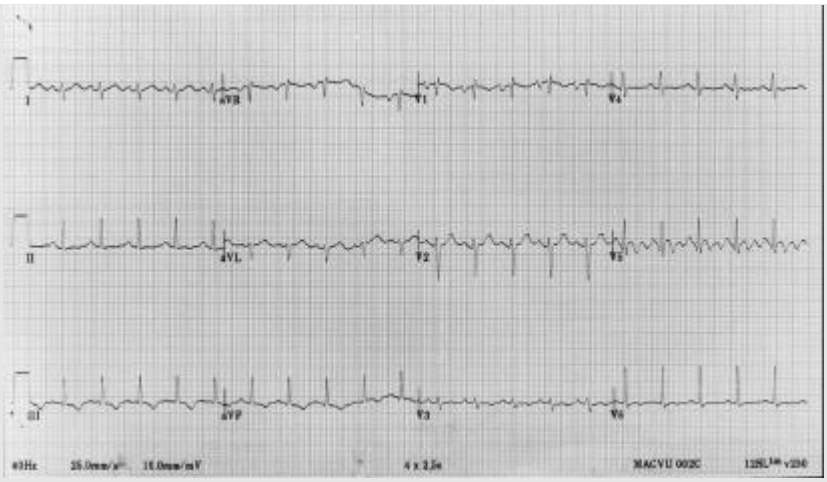
On admission, he had dyspnea and palpitations, a blood pressure of 130/80 mmHg and a pulse rate of 120 b/min. Complete blood count, automated serum chemistries and urinalysis were normal. The electrocardiogram revealed sinus tachycardia, incomplete right bundle branch block and inverted T wave in lead III-aVF, V_4-V_6 (Fig. 1A).

Echocardiography revealed an important pericardial effusion and a voluminous (10 u 7 cm), immobile mass that protruded from the epicardium of the diaphragmatic heart wall into the echo-free space of the pericardial effusion. The mass showed strands of tissue connecting it with the epicardium and the inferior vena cava wall at the inferior vena cava-right atrial junction. The mass (Figs. 1B and C) extended from the atrium to the apex and it appeared inhomogeneous with central echo-lucent areas. Left ventricular inferior wall was mildly hypokinetic, suggesting myocardial infiltration. A magnetic resonance imaging scan of the thorax showed a large (10.1 u 4.9 cm) and heterogeneous, oval shaped mass (Fig. 1D) and confirmed its adhesion to the surrounding cardiovascular structures, supporting the suspicion of the neoplastic nature of the mass. A total body computed tomographic scan excluded other neoplastic localizations.

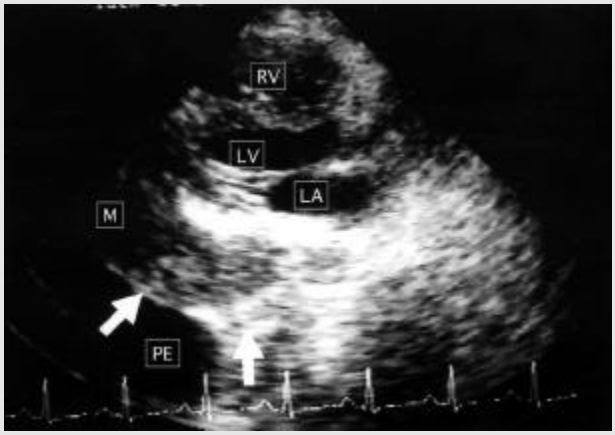
Due to recurrent effusion, several pericardiocenteses were performed. Cytological examinations of the serosanguineous pericardial fluid were negative for the presence of malignant cells. A surgical exploration permitted mass biopsy and the creation of a pleuro-pericardial window. The histological examination of the specimens revealed a rare spindle cell sarcoma: a malignant peripheral nerve sheath tumor[1]. To our knowledge this is the first reported case of pericardial localization of this tumor.

Reference

1. Pauwels P, Dal Cin P, Sciot R, et al. Primary malignant peripheral nerve sheath tumor of the heart. Histopathology 1999; 34: 56-9.



A



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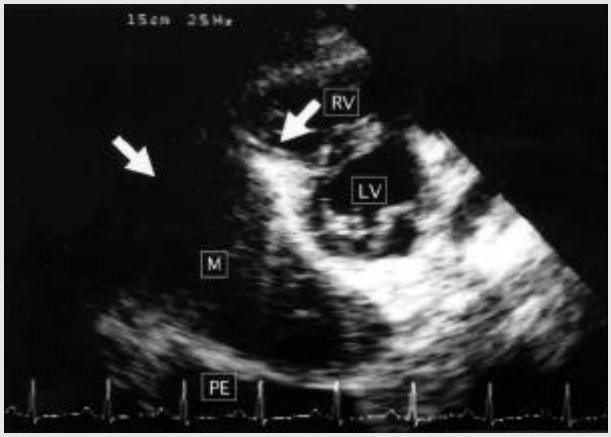

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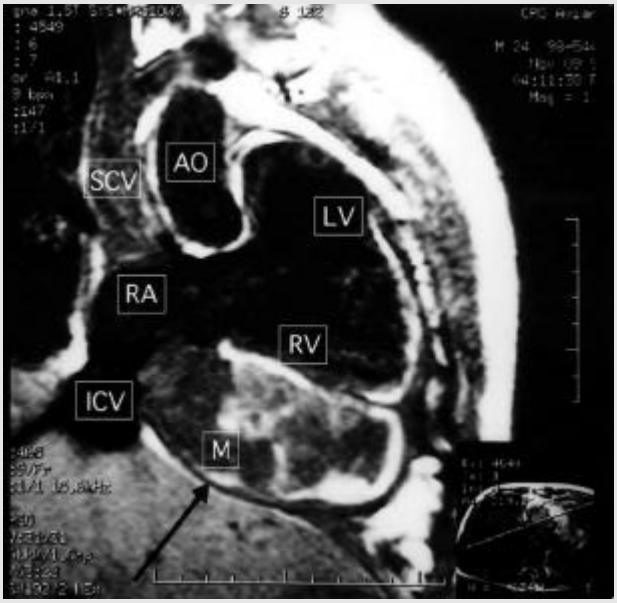

D

Figure 1. A: 12-lead electrocardiogram; B and C: echocardiographic parasternal long- and short-axis views; D: magnetic resonance imaging scan of the thorax. AO = aorta; ICV = inferior caval vein; LA = left atrium; LV = left ventricle; M = mass; PE = pericardial effusion; RA = right atrium; RV = right ventricle; SCV = superior caval vein.