An Unusual Case of Cardiomegaly

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A 64-year-old woman presented to the emergency department because of progressive fatigue and weight loss for the previous 3 months. She had a 10 pack-year history of smoking with no other significant medical history. Physical examination was unremarkable with normal cardiac and pulmonary auscultation. Her electrocardiogram showed sinus rhythm with very low voltages in all leads (Figure 1A). A chest x-ray film showed enlargement of the cardiac silhouette with normal pulmonary vascularization (Figure 1B). A trans-thoracic echocardiography was performed that revealed an homogeneous mass next to the right side of the heart without clear infiltration and moderate pericardial effusion (Figure 1C; see Movies I through III in the online-only Data Supplement). A contrast-enhanced full-body computed tomography showed a 12 × 8 cm soft-tissue density mass with some necrotic areas in the anterior and medium mediastinum (Figure 1D). No clear pericardial infiltration or involvement was noted, but small pleural and pericardial effusion was present. To better characterize the tumor, a cardiac magnetic resonance was performed, showing an 11 × 8.5 cm intrapericardial mass, with well-defined borders and no signs of pericardial or myocardial infiltration. Partial right atrium and superior vena cava compression was noted, without significant hemodynamic disturbance. T1-weighted fast spin-echo sequence showed multiple hyperintense subcentimetric areas that were hypointense in T2-weighted sequences. There were likely fibrous hypointense septa in both T1- and T2-weighted images (Figure 1E through 1G; see Movies IV and V in the online-only Data Supplement). Initial differential diagnosis was lymphoma, teratomatoma, or thymoma. A computed tomography–guided biopsy was made, and the histological examination confirmed the diagnosis of a cortical subtype (World Health Organization type B1) thymoma (Figure 1H). A combined chemotherapy treatment (cisplatin, doxorubicin, and cyclophosphamide) was initiated with no significant response. The patient developed recurrent pericardial effusion and pericardial tamponade, so a surgical approach was undertaken. Surgery was consistent with diagnosis of intrapericardial tumor with no pericardial or myocardial infiltration observed (Figure 2A). Partial resection of the mass was performed. A control cardiac magnetic resonance 2 months after surgery showed a 48 × 32 mm tumoral mass posterior to the main pulmonary artery, consistent with a nonexcised tumor. No other tumoral component was depicted (Figure 2B through 2F). The patient remains asymptomatic after 4 months of follow-up.

Thymoma and thymic carcinomas are rare epithelial tumors, mainly detected in anterior or anterosuperior mediastinum. Clinical findings vary from asymptomatic patients (50% of the cases) to compression of adjacent structures and paraneoplastic syndromes, such as myastenia gravis, pure red cell aplasia, or immunodeficiency.1 Heart and great vessels are occasionally affected by orthotopic thymomas when they extend from the thymic cell in the anterior mediastinum. They are diagnosed very often after an incidental finding on chest radiography, but many cases require a multimodal approach. In our case, echocardiography made the initial diagnosis of the mass, confirmed on computed tomography, but only cardiac magnetic resonance was able to clearly delineate intrapericardial location and rule out pericardial or myocardial infiltration. As a rule, echocardiographic examination and especially cardiac magnetic resonance are essential when cardiac involvement is present as illustrated in the present patient. Although very rare, primary intrapericardial thymomas and thymic carcinomas have been described, originating from thymic cells that migrated to the pericardium during the embryonic development. There have been 6 other cases reported in the literature. Other reported ectopic sites for thymic cell tumors have been cervical, middle, or inferior mediastinum, diaphragm, or pleura.2 The prognosis varies widely depending on the tumor histology (classified by the World Health Organization) and the disease stage, which is determined by clinical and histopathologic findings.

Disclosures
None.

References
Figure 1. A, Electrocardiogram showing low voltage in all leads. B, Posteroanterior chest radiograph showing cardiomegaly. C, Two-dimensional echocardiogram obtained in parasternal long-axis view showing an homogeneous mass next to the right side of the heart. D, Full-body computed tomography in a sagittal view showing a soft-tissue density mass in the anterior and medium mediastinum with no clear pericardial visualization. E, T1-weighted fast spin echo in the axial plane showing an homogeneous 11 × 8.5 cm iso/hypointense mass anterior to the right ventricular outflow tract and right ventricle. Fat-plane separation is seen without evidence of pericardial or myocardial infiltration. F, Spectral presaturation with inversion recovery sequence showing hyperintense mass without fat component. G, T1-weighted fast spin echo post-Gadolinium in which light enhancement is seen. H, The mediastinal biopsy showed a tumor with thymic oval cells with low-grade cellular atypia and plenty of lymphocytic T cells (×20 magnification). The stain with cytokeratin AE1/AE3 highlight the thymic epithelial cells.
Figure 2. A, Surgical resection of the intrapericardial thymoma (view from a standard median sternotomy). * shows the intrapericardial tumor, and ** shows the pericardial edges. B, T1-weighted fast spin echo in the axial plane after surgical resection showing tumor absence next to the right heart chambers. C and D, T1-weighted fast spin echo in axial plane at the level of great vessels showing the remaining tumoral mass posterior to the main pulmonary artery. E, Cine cardiac magnetic resonance image (gradient echo sequence) in a 4-chamber view showing small pericardial effusion with no compression of the right cardiac chambers. F, Cine cardiac magnetic resonance image (gradient echo sequence) in a 2-chamber view showing the postsurgical remaining tumoral mass inferior to the left pulmonary artery.
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Movie Legend

Movie 1. Two dimensional echocardiogram obtained in subcostal view showing an homogeneous mass next to the right side of the heart and small pericardial effusion. Video can be viewed with the following applications: Windows Media Player, VLC Media Player.

Movie 2. Two dimensional echocardiogram obtained in apical four-chamber view. The tumor is located lateral to the right atrium that shows diastolic compression without hemodynamic compromise. Anterior and lateral moderate pericardial effusion is also noted. Video can be viewed with the following applications: Windows Media Player, VLC Media Player.

Movie 3. Two dimensional echocardiogram obtained in parasternal long-axis view. A homogeneous anterior mass with low echogenicity is seen in close relation with aortic root. Right ventricular wall moves normally and extrinsic compression is not seen. Video can be viewed with the following applications: Windows Media Player, VLC Media Player.

Movie 4. Cine Cardiac Magnetic Resonance image (gradient echo sequence) in a four-chamber view showing the heterogeneous tumor within the pericardial layers along with pericardial effusion. Partial right atrium compression is seen. Epicardial fat is depicted without evidence of pericardial or myocardial infiltration. Video can be viewed with the following applications: Windows Media Player, VLC Media Player.
**Movie 5.** Cine Cardiac Magnetic Resonance (gradient echo sequence) in a short axis view showing the anterior tumor location within the pericardial sac. Video can be viewed with the following applications: Windows Media Player, VLC Media Player.