A 46-year-old woman suffered effort intolerance for months. She had been healthy in the past, without systemic diseases. She had experienced near-syncope before arrival at our center. On admission, her physical examination was unremarkable. ECG exhibited normal sinus rhythm, P pulmonale, and first-degree atrioventricular (AV) block. A transthoracic echocardiogram demonstrated a huge right atrial (RA) mass, 7.98\,×\,5.70 cm in size, with 23 mm Hg of transvalvular pressure gradient. The global ventricular contractility was normal (Data Supplement Movies I and II). These findings were confirmed by 3-dimensional echocardiography (Figure 1; Data Supplement Movies III and IV). Cardiac magnetic resonance imaging showed a large, well-defined intracavity mass on T1-weighted image. After contrast injection, delayed images revealed a strong signal indicating hypervascularity (Figure 2 and online-only Data Supplement), highly suggestive of hemangioma, angiomylipoma, or angiosarcoma. Coronary angiography failed to locate the feeding artery, but RA and right ventricular angiography showed a large space occupied by the lesion (Figure 3).

The patient had an episode of syncope on her second day of stay. ECG showed complete AV block and junctional escape rhythm with a ventricular rate of 50/min (Figure 4). She received emergency surgical intervention for complete excision of the RA tumor (Figure 5). Cardiac function recovered promptly after surgery. A temporary epicardial pacemaker was used initially, but because of persistent complete AV block, a permanent pacemaker subsequently was implanted. The patient was discharged uneventfully 9 days after the operation. She has been followed up for 6 months without any echocardiographic evidence of recurrence.

On retrospective analysis of the patient’s data, we noticed that her ECG taken in September 2004 had a first-degree AV block with a PR interval of 0.24 second. In May 2007, the PR interval was found to be prolonged (0.32 second), and a 24-hour Holter depicted asymptomatic intermittent complete AV block. By June 2007, symptomatic bradycardia had developed. Thus, progression of AV block took at least 3 years.

Hemangiomas of the heart are benign by cell type but can have serious consequences. RA hemangioma on interatrial septum is typically silent, but the location and gross appearance of our case mimicked a mesothelioma of the AV node. Mesotheliomas of the AV node are well known to cause heart block and sudden death. Thus, the finding of complete heart block and syncope in our patient suggests that hemangiomas are not always clinically benign if located in areas of cardiac conduction systems. To the best of our knowledge, this is the first case of RA hemangioma to demonstrate complete AV block.

Disclosures

None.

References

Figure 1. Three-dimensional echocardiogram demonstrates significant dilatation of the RA with a huge, solid RA mass protruding into the right ventricle through the tricuspid valve. H indicates hemangioma; RV, right ventricle; and LV, left ventricle.

Figure 2. Cardiac magnetic resonance imaging documents the presence of a delayed high-signal mass (*) in the RA on T1-weighted image after gadolinium contrast injection. The cavernous type of cardiac hemangioma tends to not show a rapid signal enhancement with contrast administration because of the slow blood flow.

Figure 3. RA and right ventricular angiography show a large filling defect, which nearly obstructs the orifice of tricuspid valve and extends to right ventricular chamber, with decreased tricuspid inflow. RV indicates right ventricle.
Figure 4. ECG reveals complete AV block and junctional escape rhythm with a ventricular rate of 50/min.

Figure 5. The excised gross tumor. Arrow indicates a debulked part of the huge tumor. Upper left, Surgeon’s view of the open RA reveals a large, smooth-walled tan-red mass. The fixed part of the tumor arises from the interatrial septum close to tricuspid valve and is involved in endocardial cushion. Upper right, Histological section shows that the tumor is composed of endothelial-lined, blood-filled cavernous spaces separated by a fibromyxoid stroma (hematoxylin and eosin stain, ×100).
Malignant Presentation of Cardiac Hemangioma: A Rare Cause of Complete Atrioventricular Block
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