Venous Hemangioma in the Right Atrium Possibly Related to Radiofrequency Catheter Ablation

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Venous hemangioma is a rare, benign cardiac tumor, usually in young or adult males. A 61-year-old woman with a clinical history of radiofrequency catheter ablation had venous hemangioma of the right atrium diagnosed after an episode of acute pain in the precordial region. The incidental discovery of the mass, clinical evaluation, operative procedure, pathologic findings and follow-up are reported, as well as the epidemiology, natural history, and diagnostic and therapeutic approaches. Etiology may be related to damage to endothelial cells of the interatrial septum or to endocardial cells. (Circ J 2008; 72: 1712–1714)

Key Words: Catheter ablation; Radiofrequency surgery; Right atrium; Venous hemangioma

With an incidence varying from 0.002 to 0.3% at autopsy, primary cardiac tumors are very uncommon.1,2 Cardiac hemangioma, which is predominantly composed of blood vessels, has an incidence of less than 5% of all benign primary cardiac tumors.3,4 It occurs in patients of all ages, with a male predominance and a mean age at clinical presentation of 43 years. We present here a 61-year-old woman with a clinical history of radiofrequency catheter ablation whose symptoms were initially precordial chest pain.

Case Report

A 61-year-old woman with a clinical history of radiofrequency catheter ablation for paroxysmal supraventricular tachycardia in another hospital 18 months previously was referred for an episode of acute precordial chest pain. She had been diagnosed as having atrioventricular nodal reentrant tachycardia and slow atrioventricular nodal pathway ablation had been performed, but no further details of the procedure could be obtained. Post-ablation, she did not have any discomfort in the precordial area.

On presentation, the patient did not have signs of fever, general illness, joint pain, dyspnea, or cough. There was no clinical history of cutaneous or visceral hemangioma. Auscultation revealed a faint systolic murmur and she underwent transthoracic echocardiography, which demonstrated a weak-echo right atrial mass with a non-homogeneous pattern, apparently capsulated, ovoid in shape ($\approx 35 \times 18$ mm) and labile throughout the cardiac cycle. It was approximately 35x18 mm in diameter and originated in the middle of the cranial surface of the interatrial septum (Fig 1). No cardiac tumor had been detected in transthoracic echocardiography performed before radiofrequency catheter ablation. Her chest X-ray was normal, and electrocardiogram showed only non-specific alterations of the T-wave and S-T complex (Fig 2). The patient was also underwent coronary angiography, which revealed normal myocardial circulation, but contrast injections from both the right and left coronary arteries showed abnormal circulation in the region of the atria (Fig 3). The patient was classified as New York Heart Association II and surgical treatment was proposed. Access was gained through a right muscle-sparing thoracotomy and a standard lateral right atriotomy was performed. The mass, which originated from the right atrial septum, was completely resected and the interatrial septum was rebuilt with an autologous pericardial patch. The procedure required...
Fig 2. Electrocardiography reveals only non-specific alterations of the T-wave and S-T complex.

Fig 3. Neovascularization to the mass originating from (a) the left circumflex and (b) the right coronary artery.

Fig 4. Histopathological evaluation of a venous hemangioma. (a) Large vascular spaces are separated by fibrous septa and partly filled with blood. Focal hemorrhagic areas and hemosiderin pigment can be seen, but cell atypia and necrosis are absent (×200). (b) Thin- and thick-walled vascular cavities partly filled with blood and lined with flattened endothelium (×60).
cardiopulmonary bypass and mild hypothermia, but was uneventful. The patient had a smooth recovery and was discharged 7 days after operation.

Macroscopically, the mass was 20×25×35 mm, purple, moruloid and well capsulated. The cut surface was bleeding. Histopathology evaluation revealed a venous hemangioma with large cavernous vascular spaces separated by fibrous septa and partly filled with blood (Fig 4). Focal hemorrhagic areas and hemosiderin pigment were found. Cellular atypia and necrosis were absent.

Discussion

Cardiac hemangioma is a very rare benign tumor; no more than 50 cases have been reported in the literature to date, and cardiac venous hemangioma is the most uncommon. The tumor originates from vascular cells and has a varied histological appearance: capillary, arteriovenous, and racemose and cavernous. Cardiac hemangioma is usually solitary, but has been associated with extracardiac hemangioma, including cutaneous or visceral sites. To date its etiology is unknown. In the present patient, a cardiac venous hemangioma originated less than 18 months after radiofrequency catheter ablation, without a clinical history of cutaneous or visceral hemangioma. Therefore, we infer that damage to the endothelial cells of the interatrial septum or to endocardial cells may have induced development of the tumor, but we have no evidence.

Anatomically, cardiac hemangioma can be pericardial, intramyocardial or subendocardial. Although any of the cardiac chambers can be involved, it is more frequent in the right atrium and usually arises in the posteroseptal aspect, close to the superior vena cava and compressing it without sign of invasion. Age at presentation can range from infancy to late adulthood. Clinical presentation is highly variable according to the location, size and extension of the tumor. The most common presentation is exertional dyspnea, but arrhythmias, pseudoangina, and signs of right heart failure are not uncommon presentations. Rarely, the initial presentation includes pericardial effusion, pericarditis, or failure to thrive. A small percentage of cases are asymptomatic. Auscultation often reveals a systolic murmur similar to that associated with pulmonary stenosis. In the present case, the cardiac hemangioma originated from the right upper interatrial septum, and the clinical presentation included precordial chest pain and a faint systolic murmur.

The differential diagnosis is obtained with echocardiography, computed tomography or magnetic resonance imaging and confirmed by histological examination. Preoperative diagnosis occurs in a minority of cases. Echocardiography has an accuracy rate of 81% for detecting cardiac tumors. The differential echocardiography diagnosis is usually cardiac myxoma arising from the fossa ovalis of the interatrial septum, but unlike hemangioma, in most cases myxoma is located in the left atrium, pedunculated and mobile, without any sign of atrial wall invasion. Cardiac catheterization studies can help to diagnose cardiac tumors in 40% of cases by revealing an intracavitary filling defect. Coronary angiography will often reveal characteristic vascularity? In this case, echocardiography identified a cardiac mass, coronary arteriography showed abnormal atrial circulation, and venous hemangioma was finally confirmed by histological examination.

The natural history of cardiac hemangioma is unpredictable but once found, the patient should be treated quickly, usually requiring surgical resection if technically feasible. The surgical outcome is generally favorable, and surgical resection is considered to be curative in most cases, although recurrence has been reported in a few cases. Patients typically do well postoperatively, but the risk of developing hemangioma elsewhere in the body is not known. In 1 case, the patient developed an intracardiac angiosarcoma after excision of a left atrial hemangioma. The current patient is doing well at 6-month follow-up.

Conclusion

Cardiac hemangioma is a rare cardiac tumor with an unknown etiology, but its development may be related to damage of the endothelial cells of the interatrial septum or to endocardial cells. The diagnosis is obtained from imaging studies, cardiac catheterization and arteriography, and confirmed by histological examination. Successful treatment usually requires timely surgery and the postoperative outcome is generally favorable, although periodic examinations and echocardiography are recommended.

References