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CASE REPORT

PATHOLOGY/BIOLOGY

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Sudden Death Owing to Right Atrial Hemangioma*

ABSTRACT: Primary cardiac tumors are rare, with an autopsy incidence of 0.03%. Seventy-nine percent to 85% of primary cardiac tumors are benign, and of the benign tumors, cardiac hemangiomas account for 5–10% of cases. Most cardiac hemangiomas are asymptomatic and are discovered incidentally at autopsy, or by echocardiography, computerized tomography, or magnetic resonance imaging. We report a case of sudden death owing to cardiac hemangioma in a 22-year-old woman who collapsed while shopping and became unresponsive. The autopsy revealed a hemorrhagic mass on the surface of the right atrium which was infiltrating and replacing the wall of the right atrium; histopathological examination confirmed the tumor was a cavernous hemangioma. Fewer than 20 cases of right atrial cavernous hemangiomas have been reported in English literature, emphasizing the rarity of our case.

KEYWORDS: forensic science, tumor, hemangioma, sudden death

Primary cardiac tumors are rare, with an autopsy incidence of 0.03% (1). Seventy-nine percent to 85% of primary cardiac tumors are benign, and of the benign tumors, cardiac hemangiomas account for 5–10% of cases (2). Hemangiomas consist of benign proliferations of endothelial cells usually forming channels containing blood. Most cardiac hemangiomas are asymptomatic and are discovered incidentally at autopsy, or by echocardiography, computerized tomography (CT), or magnetic resonance imaging (MRI). We report a case of right atrial cavernous hemangioma found at autopsy in a 22-year-old woman.

Case

A 22-year-old lady collapsed while shopping and became unresponsive. In the accident and emergency (A&E) department, her Glasgow Coma Scale was 3/15, cardiopulmonary resuscitation was unsuccessful, and the patient was confirmed dead. The patient had presented to A&E a month earlier with shortness of breath and chest pain. She had no other past medical history but did have breast augmentation surgery 4 weeks prior to her death.

At autopsy, macroscopic examination revealed a hemorrhagic mass 15 × 10 mm on the surface of the right atrium which was infiltrating and replacing the wall of the right atrium (Fig. 1). This formed rounded blue-colored masses 20 × 15 mm (in total), in the subendocardium of the right atrium encircling the wall which was fragmented. It also involved the right atrial appendage. There was extensive hemorrhage within the mass. The mass infiltrated as far as the atrioventricular junction but did not appear to involve the right coronary artery or right ventricle. The sinoatrial node area could not be identified.

FIG. 1—*Mass 15 × 10 mm on the surface of the right atrium infiltrating and replacing the wall of the right atrium.*

Microscopic examination of the right ventricle showed a hemorrhagic mass consisting of dilated vascular channels lined by flat cells and other more solid areas with admixed red blood cells (Fig. 2*a*,*b*). The mass infiltrated and destroyed the right atrial wall, replacing it by tumor. The cells did not look malignant with no mitoses, but there was extensive hemorrhage. The sinoatrial and atrioventricular node could not be identified.

These findings were consistent with sudden cardiac death owing to an infiltrating hemangioma of the right atrium destroying the wall and leading to fatal arrhythmia.

Discussion

Cardiac hemangiomas are rare benign vascular tumors of the heart, with fewer than 50 surgically treated cases reported in the

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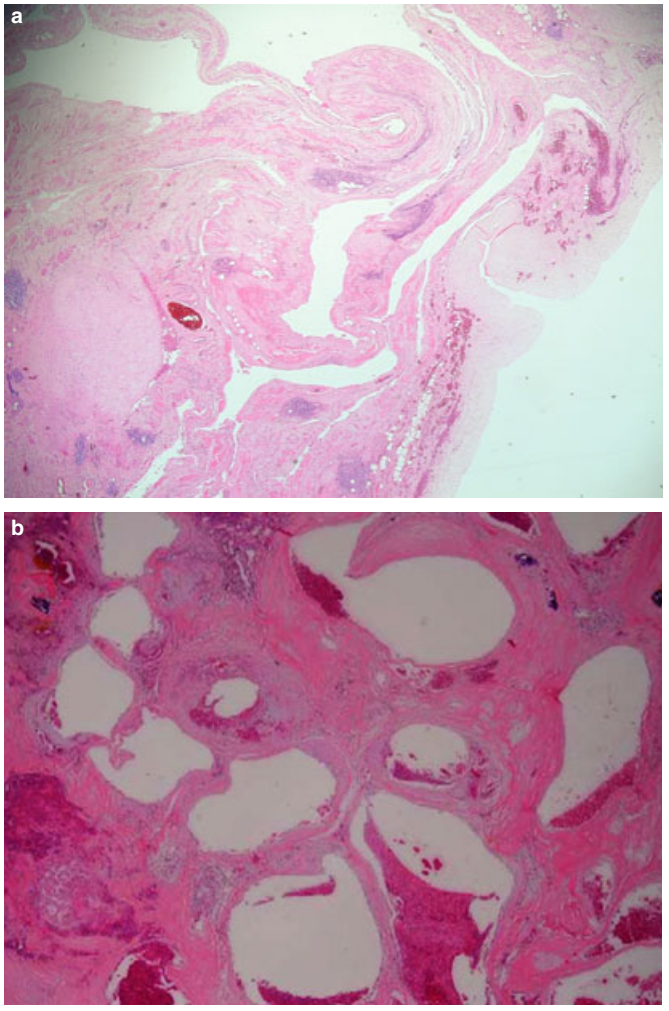

FIG. 2—*(a) Microscopy of the right atrium showing a hemorrhagic mass consisting of dilated vascular channels lined by flat cells and other more solid areas with admixed red blood cells (hematoxylin & eosin stain; magnification ×200). (b) Higher magnification showing irregular, relatively large endothelial-lined vascular structures filled with blood. Degenerative changes including organizing hemorrhage, inflammation, fibrosis, and calcification are also present (hematoxylin & eosin stain; magnification ×300).*

literature. They can occur in any age-group, with cases reported in infants and elderly (3). They can involve any part of the heart but show right ventricular predominance. A study by Burke et al. (2) looking at 10 cases of hemangioma found four cases occurring in the right ventricle, three in the left ventricle, two in the atrial septum, and one in the right atrium. Right atrial hemangiomas, like our case, account for 23.2% of all cardiac hemangiomas (3).

Most hemangiomas are clinically asymptomatic and are detected incidentally at autopsy, on chest X-ray, or on investigation of a murmur. Clinical presentation of symptomatic hemangiomas depends on their size and location. Patients may present with arrhythmias, pericardial effusion, chest pain, shortness of breath, or sudden death (2). Hemangiomas causing sudden death generally cause conduction disturbances in the heart and may be located in the region of the atrioventricular node or in the ventricles. In our case, the hemangioma infiltrated as far as the atrioventricular junction but did not involve the right coronary artery or right ventricle. A rare mechanism of sudden death in individuals with cardiac hemangioma is rupture of the tumor and pericardial tamponade (2). A study by Cina et al. (4) described 120 cases of sudden death attributed to primary cardiac tumors, of which six were attributable to hemangiomas, emphasizing the rarity of these tumors to present as sudden death.

Characteristically, hemangiomas appear red and hemorrhagic. They may be classified as: (i) cavernous hemangioma (widely dilated vascular channels lined by flattened endothelial cells with focal abundant connective tissue between the channels), as found in our case; (ii) capillary hemangioma (haphazardly arranged closely packed capillary structures lined by flattened endothelial cells with minimal stroma); and (iii) arteriovenous hemangioma or cirsoid aneurysm (dysplastic malformation of arteries and veins). Cardiac hemangiomas often have combined features of capillary, cavernous, and arteriovenous hemangiomas. The cavernous and capillary types are reportedly encountered more frequently. Fewer than 20 cases of right atrial cavernous hemangiomas have been reported in English literature, emphasizing the rarity of our case.

Diagnosis of cardiac hemangiomas can be established with two-dimensional transthoracic echocardiography (2DTTE), transesophageal echocardiography, CT scan, or MRI. Three-dimensional TTE (3DTTE) can be more definitive than 2DTTE in distinguishing hemangiomas from other cardiac masses with exhibited echolucencies, such as myxoma or thrombus (5). On a 3DTTE, the echolucencies in a hemangioma appear to involve the entire extent of the tumor, including the peripheral regions, in contrast to the regional occurrence seen in a myxoma or hematoma. CT scan and MRI provide better soft-tissue contrast than echocardiography and hence can provide useful information about the invasiveness of the tumor. Also, visualization of the entire mediastinum with these modalities helps in detection of the extracardiac extent of these tumors. However, in most instances, definitive diagnoses of cardiac hemangiomas are made after surgical excision and histological examination (5).

Regression of hemangiomas, either spontaneously or after steroid treatment, has been reported in the literature (2). However, surgical excision of the hemangiomas appears to be curative and is the treatment of choice. Complete surgical excision is possible in most cases, but excision of large hemangiomas can be complicated by incomplete excision or reconstructive surgery. There is a reported case (2) of cardiac angiosarcoma occurring in a patient 7 years after the surgical excision of a left atrial hemangioma. Thus, it seems reasonable to follow surgically treated patients with echocardiography to detect the recurrence of tumors after resection.

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