CASE REPORT

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Sudden Death Due to Tricuspid Valve Myxoma with Massive Pulmonary Embolism in a 15-Month Old Male

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ABSTRACT: Myxomas of the tricuspid valve are extremely rare, with only 13 cases reported in the world literature (1). We report the case of a 15-month old male with tricuspid valve myxoma and massive myxomatous pulmonary emboli discovered at autopsy.

KEYWORDS: forensic science, forensic pathology, myxoma, tricuspid valve, death

A previously healthy 15-month old male was admitted to Athens-Limestone Hospital, Athens, Alabama, for evaluation of acute onset of respiratory distress, non-productive cough and hepatomegaly. Shortly after admission and before any work-up could be completed, he suffered a respiratory arrest. Resuscitative measures were initiated immediately, with profound bradycardia deteriorating to agonal rhythm on cardiac monitoring. The patient expired in spite of aggressive and prolonged attempts at resuscitation. A postmortem examination was performed the following day at the Alabama Department of Forensic Sciences in Birmingham, Alabama.

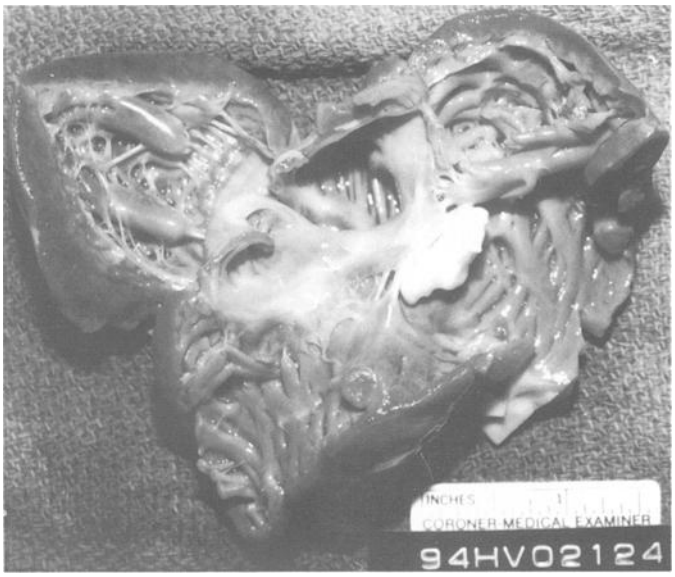

FIG. 1—*Gross photograph of opened heart showing bosselated, pale yellow mass attached to the septal cusp of the tricuspid valve.*

Autopsy Findings

Significant findings at autopsy were in the cardiovascular and pulmonary systems. Examination of the heart revealed mild dilatation of the right atrium and right ventricle. A bosselated, pale yellow, partially calcified mass was attached to the septal cusp of the tricuspid valve (Fig. 1). The mass measured 2 by 1 by 1 cm in greatest dimensions, and had a smooth and glistening external surface. The pulmonary, aortic, and mitral valve leaflets were delicate and pliable. The atrial appendages contained no thrombi and no additional masses were seen within the heart.

Histologically, the mass was composed of stellate and globular myxoma cells and mononuclear leukocytes embedded within an amorphous, vascularized, myxoid stroma (Fig. 2). Areas of fibrosis

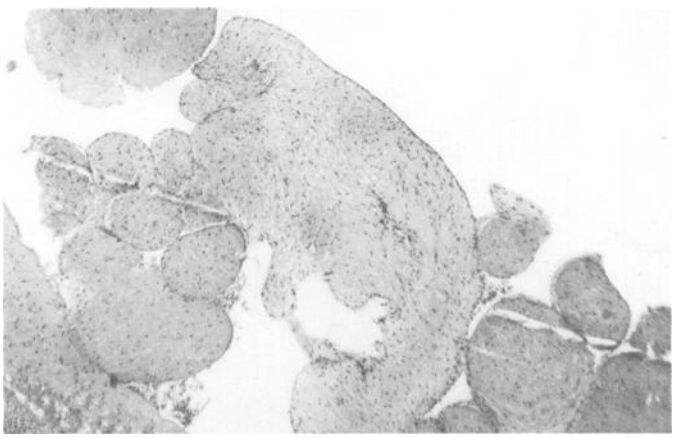

FIG. 2—*Photomicrograph of tricuspid valve (TV) mass showing stellate cells embedded within an amorphous, vascularized, myxoid stroma. (Hematoxylin and Eosin; original magnification ×40).*

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and calcification were present (Fig. 3). No iron pigment nor gamnabodies were seen.

The lungs were congested (combined weight 280 g) with variegated thromboemboli completely occluding both main pulmonary arteries and extending into several smaller branches (Fig. 4). Microscopic examination of the large emboli showed myxomatous material identical to that in the tricuspid valve mass (Fig. 5). Multiple additional myxomatous emboli were seen throughout the pulmonary parenchyma.

No additional findings that could be related to the patient's death were detected at autopsy. An incidental finding was hypoplasia and fibrosis of the left lobe of the liver.

Discussion

Cardiac myxomas are rare neoplasms occurring in 0.001 to 0.03% of all autopsies (2–4). Although myxomas have been described in all four chambers of the heart, most occur as sporadic lesions arising within the left atrium (5). Myxomas are more common in females with an estimated sex ratio of 3:1 (3). These uncommon neoplasms have been reported in patients from 3 months to 95 years of age (3), but are usually seen in young to middle-aged adults (3–6).

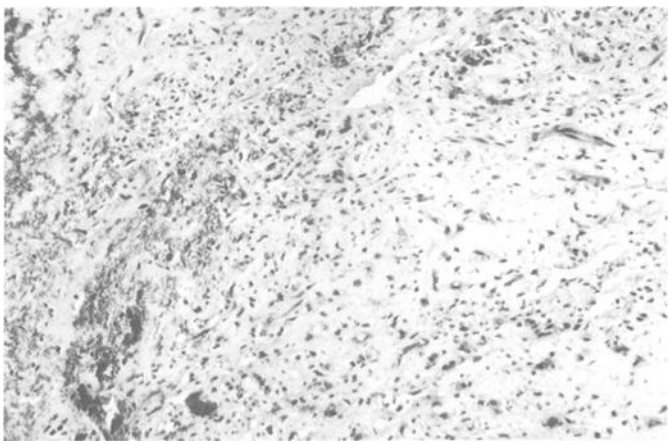

FIG. 3—*Photomicrograph of TV mass showing fibrosis and granular calcification. (Hematoxylin and Eosin; original magnification ×100).*

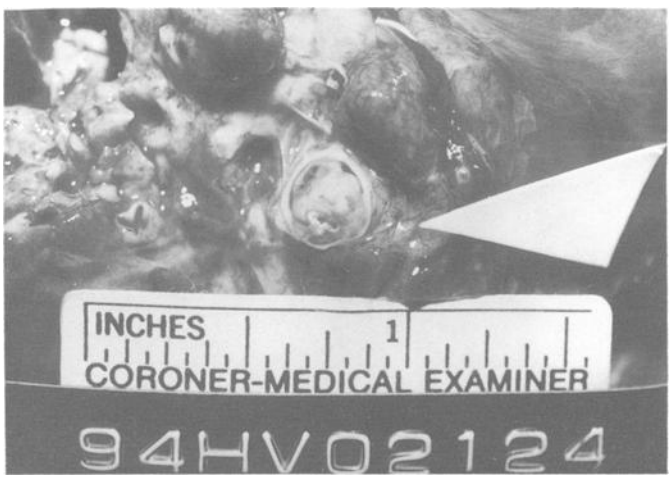

FIG. 4—*Gross photograph of thromboembolus occluding pulmonary artery (left of marker).*

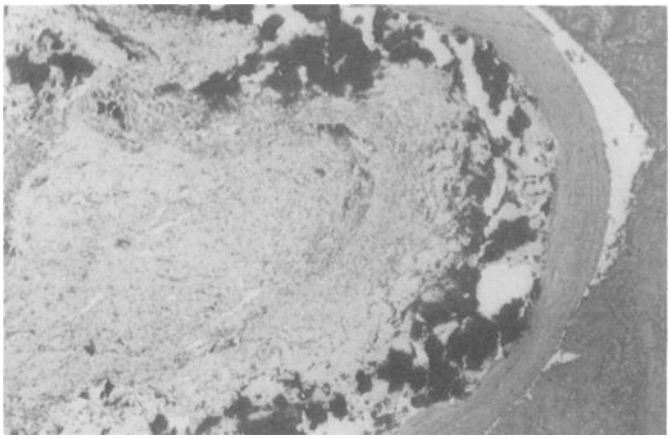

FIG. 5—*Photomicrograph of pulmonary embolus, which is composed of calcified, myxoid material identical to that in the TV mass. (Hematoxylin and Eosin; original magnification ×40).*

Unlike myxomas arising from the atria, those arising from valvular tissue are more often found on the right side of the heart (7). Myxomas arising from the tricuspid valve are extremely rare, with only 13 cases reported in the world literature (1). To our knowledge, the subject of the current case report is the youngest patient thus far described with a myxoma attached solely to the tricuspid valve (TV).

Although TV myxomas give no distinct clinical presentation, most patients have symptoms related to obstruction of blood flow (2) and present with acute right-sided heart failure, as in the current case. Other presenting symptoms include syncope, fever of unknown origin, dyspnea on exertion, productive cough, palpitations or rarely, sudden death (1,2,7).

A review of ten published reports of TV myxomas by Cole et al. revealed four cases with tumor attachment on the atrial side of the valve, as did our case, and six with attachment on the ventricular side (7). Among those reports that include a description of the involved leaflet(s), five cases, including the current one had tumor attached to the septal leaflet of the valve (1,7–9).

Echocardiography is considered the diagnostic method of choice for cardiac myxomas (6,10), and although this modality was used in only 5 of the 13 reported cases of TV myxoma, an accurate diagnosis was made in all five cases (1,7,8).

Echocardiographic findings that suggest valvular origin include the presence of a right atrial mass with complete prolapse into the right ventricle during systole (7).

The histologic appearance of TV myxomas is similar to myxomas arising elsewhere in the heart (1,2,7,8). Worthington et al. reported a TV myxoma infected with a gram-negative rod of the dysgonic-fermenter group 2 (8).

Familial and syndromic forms of cardiac myxoma have been described (3,5,6,9,11–14). Fewer than 5% of all myxomas are seen as part of the syndromic form, first described by Carney, and also known as Carney's Complex (5,6,11). This syndrome is characterized by multiple, recurrent myxomas, pigmented skin lesions and endocrine abnormalities (6,11). In a review of the world literature by van Gelder et al., 17 families with myxoma were identified (5). Although the mode of inheritance has not been definitively established, autosomal dominant inheritance has been indicated (15). Siltanen et al. described minor malformations of other organs (double ureter, spondylolisthesis, and cystic fibromas of the tibia and femur) in three of four patients with familial atrial myxoma

(3). A minor malformation found in our case was hypoplasia and fibrosis of the left lobe of the liver.

Histologically, familial myxomas are similar to other myxomas but have been described as extensively myxomatous lesions (6) with calcification (5) and lacking gamna-bodies (6). In our case, there was multifocal calcification without hemosiderin deposition or gamna bodies.

Patients with both the syndromic and familial forms tend to present at an earlier age, have multicentric tumors, tumors in unusual locations, and a much higher rate of recurrent lesions (5,6,10,16). In those cases in which there is clinical suspicion of a familial or syndromic form of myxoma, screening of family members with periodic echocardiographic examination is recommended to detect subclinical or recurrent lesions (5,9,13).

Total excision of the primary tumor with a rim of uninvolved cardiac tissue is the treatment of choice (1). This may result in tricuspid valve insufficiency, which may require replacement of the valve (1).

The young age of the patient and unusual location of the tumor in this case are suggestive of the familial form of cardiac myxoma. The family members of the subject are reportedly in good health and have not, as yet, undergone echocardiographic examination. The presence of subclinical myxomas in the siblings or parents of this patient would support the diagnosis of familial myxoma.

In summary, TV myxomas are exceedingly rare neoplasms which may present with signs or symptoms of right-sided heart failure or sudden death. Echocardiography is the diagnostic method of choice. Surgical excision with a margin of uninvolved cardiac tissue is the preferred treatment. An atypical site of tumor attachment, occurrence in a child, or the presence of multiple or recurrent tumors, should raise clinical suspicion of a familial or syndromic form of myxoma, and prompt echocardiographic screening of family members.

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