Cardiac Rhabdomyoma in an Adult Patient Presenting With Ventricular Arrhythmia*

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Cardiac rhabdomyomas are extremely uncommon in the adult patient. We describe a previously healthy man who presented with ventricular arrhythmias resulting from a right ventricular, cardiac rhabdomyoma. Echocardiography, CT scanning, and MRI are recognized as useful diagnostic modalities for intracardiac lesions. Cardiac catheterization in our patient demonstrated the presence of a tumor blush. This has not previously been reported with cardiac rhabdomyomas. Although lesions may spontaneously regress, surgery is often necessary and frequently resolves the underlying arrhythmia.

Key words: adult; arrhythmia; blush; cardiac; coronary; rhabdomyoma; spider

Primary cardiac tumors are uncommon, particularly in the adult population. It is even more unusual when they are associated with arrhythmic events that are reversible with resection of the tumor. We present such a case and focus our discussion on three unique characteristics this case demonstrates.

CASE REPORT

The patient is a 35-year-old man with no significant medical history who presented to his local emergency department complaining of palpitations. He had previously been in excellent health and jogged at least two miles at least three times weekly without any symptoms. Two months prior to presentation, he began noting episodes of palpitations. These usually occurred as he was sitting at his desk at work, lasted a few seconds to several minutes, and were not associated with chest pain, shortness of breath, nausea, or presyncope. He initially attributed these to his stressful lifestyle, and did not seek medical attention until the morning of presentation, when he had a prolonged episode that was associated with severe lightheadedness. He had no history of hypertension, diabetes mellitus, or hyperlipidemia. His father required an angioplasty at age 50 for crescendo angina; the patient admitted to smoking 1 pack/d for 20 years.

The patient was taken to the local emergency department by his coworkers, and on arrival again began feeling lightheaded. When seen in triage, his pulse was noted to be rapid and he was placed on a cardiac monitor. He was noted to have frequent premature ventricular contractions and several short runs of non-sustained ventricular tachycardia (Fig 1). He was given an aspirin, IV metoprolol, and started on a lidocaine drip. A chest radiograph demonstrated clear lung fields and no cardiomegaly. Myocardial infarction was ruled out via serial ECGs and creatine kinase measurements. An echocardiogram was subsequently performed to assess his ventricular function. This showed evidence of mild left ventricular dysfunction, but was more notable for the presence of a 2 × 2-cm right ventricular mass attached to the interventricular septum (Fig 2). After transfer to Duke University Medical Center, the patient underwent a metastatic workup, including a CT scan of his chest and abdomen, which was notable only for the right ventricular mass (Fig 3); a MRI scan, which was aborted secondary to his erratic cardiac rhythm; and a total body positron emission tomography scan, which showed no evidence of metastatic disease.

A cardiac catheterization was performed to assess the coronary anatomy and to determine whether a tumor blush was present. The left ventriculogram demonstrated mild diffuse hypokinesis with an estimated ejection fraction of 50%. The coronary arteries showed no evidence of atherosclerotic plaque, but were notable for a tumor blush in the right ventricle originating from the septal perforating vessels of the left anterior descending coronary artery (Fig 4). He was evaluated by the thoracic surgical team and taken

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to the operating room for resection of the presumed malignancy. A single lesion was noted in the right ventricle with septal attachment. The lesion was resected (Fig 5); intraoperative frozen section evaluation revealed a single hypercellular neoplasm with areas of marked nuclear atypia worrisome for malignancy. The margins were clean, and the septum was patched with pericardium.

The final pathology was interpreted as rhabdomyoma. The tumor was composed of large cells with abundant clear cytoplasm and scattered spider cells (Fig 6). Scattered foci revealed striking nuclear enlargement, hyperchromasia, and multinucleation. Mitotic activity was not identified. Typical cytoplasmic glycogen was demonstrated on the periodic acid-Schiff stain (Fig 7). The muscle characteristics of the cells were confirmed by strong immunohistochemical reactions to myoglobin (Fig 8), desmin, and muscle-specific actin. The material was sent to the Armed Forces Institute of Pathology for consultation. A diagnosis of rhabdomyoma was also favored by the Armed Forces Institute of Pathology. Three weeks after surgery, an echocardiogram revealed normal left ventricular function, a moderate-sized pericardial effusion, and no evidence of recurrent disease (Fig 9). The patient is currently recovering well from his surgery and remains in sinus rhythm with no evidence of ventricular arrhythmia since the time of surgery.

**DISCUSSION**

Rhabdomyoma is a benign striated muscle tumor that is usually classified as cardiac or extracardiac based on its location and unique histology. The adult extracardiac form of rhabdomyoma is extremely uncommon, with < 100 cases reported in clinical literature. These lesions mostly occur in the head and neck. Cardiac rhabdomyoma is the most common cardiac tumor seen during infancy and childhood, with approximately 75% occurring prior to the age of 1 year. There is a well-described association with tuberous sclerosis, a familial multisystemic syndrome characterized by mental retardation, epilepsy, adenoma sebaceum, and hamartomas in multiple organs. About 40% of patients with pathologically confirmed cardiac rhabdomyomas have tuberous sclerosis; conversely, up to 50% of patients with tuberous sclerosis have evidence of cardiac rhabdomyomas on echocardiography. The lesions of rhabdomyoma are often multiple and can originate from the ventricular septum or the free wall of either ventricle. The atria are involved in up to 30% of cases. Lesions are yellow-gray in appearance and although not encapsulated, are well circumscribed from surrounding tissue. The classic microscopic finding is the “spider cell,” a large cell with a central cytoplasmic mass surrounded by fibrillar strands that give the appearance of a spider hanging in a net.

Adult cases of cardiac rhabdomyoma are extremely rare, with reported cases mostly associated with clinical syndromes, including tuberous sclerosis and Behcet’s disease. Though the majority of cardiac rhabdomyomas are multiple, our patient had evidence of only a single lesion by extensive diagnostic evaluation and on frozen section. The pathology seen in our patient is also unique. While the majority of the specimen has the classic histology of rhabdomyoma, more cellular regions with much smaller

![Figure 1](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/21953/) Rhythm strip obtained while the patient was in the emergency department demonstrating multiple premature ventricular contractions and nonsustained ventricular tachycardia.

![Figure 2](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/21953/) Apical four-chamber echocardiographic view focusing on the ventricles. A right ventricular mass is noted to be attached to the interventricular septum. LA = left atrium; LV = left ventricle; MV = mitral valve; RV = right ventricle; RV mass = right ventricular mass.

![Figure 3](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/21953/) CT scan of the chest demonstrating right ventricular mass attached to the interventricular septum. AO = aorta; IVS = interventricular septum; see Figure 2 legend for other abbreviations.
cells are also seen. These cells have abundant eosinophilic cytoplasm more characteristic of the adult (extracardiac) rhabdomyoma. A case of cardiac rhabdomyoma with pathology resembling the extracardiac variant has been described.\(^6\) Our case, however, contains densely packed regions and multinucleated cells not previously seen in the extracardiac variants. While a benign course is suggested, the absence of previous experience with such tumors makes time the ultimate determining factor of a benign tumor biology.

Less invasive medical imaging techniques, particularly high-resolution CT, transthoracic and transesophageal echocardiography, and MRI have mostly replaced cineangiography for the imaging of cardiac masses. Cardiac catheterization still remains a useful clinical tool to evaluate coronary artery patency in patients who require this prior to cardiac surgery.

Tumors are highly vascular structures, often outstripping their capillary blood supply through rapid growth. Despite this feature, the appearance of an arterial confluence, or tumor blush, on angiography is unusual. The reported tumors with a blush on angiography include renal cell carcinoma, hepatic carcinoid, hemangioma, meningioma, insulinoma, and paraganglioma.\(^7\)–\(^12\) Reports of cardiac tumors with a blush on coronary angiography have previously been limited to hemangioma, myxoma, and pheochromocytoma.\(^13\)–\(^15\) We believe this is the first report of a cardiac rhabdomyoma with tumor blush on coronary angiography.

The widespread use of imaging techniques such as echocardiography has permitted the early detection of cardiac rhabdomyomas, particularly in patients with tuberous sclerosis. This has facilitated a better understanding of

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**Figure 4.** Right anterior oblique, caudal view from left coronary angiogram demonstrating a right ventricular tumor blush emanating from the septal perforator vessels of the left anterior descending coronary artery.

**Figure 5.** Gross pathologic specimen of resected right ventricular mass with interatrial septal attachment.

**Figure 6.** Light microscopy of spider cells demonstrating abundant fibrillar cytoplasm with extensions to the cell membrane (hematoxylin-eosin, original \(\times\) 680).

**Figure 7.** The abundant intracytoplasmic glycogen content of the tumor cells (periodic acid-Schiff, original \(\times\) 680).
the natural history of these lesions. It is now known that these lesions can spontaneously regress and even resolve completely without the need for intervention.\(^{16}\) Although these tumors are benign and the majority of patients are asymptomatic, cardiac rhabdomyoma can present with a variety of symptoms including congestive heart failure, tachyarrhythmias, and sudden death. Congestive heart failure can result from flow obstruction by larger masses, mechanical interference with valve function, and arrhythmias.

The pediatric literature reports several types of arrhythmias with rhabdomyoma including supraventricular tachycardia, Wolff-Parkinson-White syndrome, atrial flutter and fibrillation, variable atrioventricular block, ectopic atrial tachycardia, premature extrasystoles, and ventricular tachycardia.\(^{17}\) The mechanisms for the majority of these rhythm disturbances still remain to be determined. Tachyarrhythmias may result from valvular interference of the tumor mass or reentry about its border with the myocardium. Accessory bundles resulting in ventricular preexcitation have been seen in certain patients, and one group of investigators has used pace mapping to localize the origin of ventricular tachycardia.\(^{18}\) There is growing evidence that resecting the tumor, as in our case, can result in a sustained resolution of symptoms and likely a cure.\(^{19,20}\) Thus arrhythmias refractory to medical management, in addition to the obstructive effects of the tumor mass remain strong indications for resective cardiac surgery.

In conclusion, cardiac rhabdomyomas are extremely rare occurrences in the adult population, mostly seen in association with complex clinical syndromes. Clinical symptoms can include arrhythmias and congestive heart failure resulting from vascular obstruction. Lesions can spontaneously regress, but surgery is often necessary. Echocardiography, cardiac catheterization, CT scanning, and MRI can all be helpful in detection and anatomic definition.

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### Figures

**Figure 8.** The presence of intracellular myofilaments consistent with myocyte origin (myoglobin stain, original × 400).

**Figure 9.** Apical four-chamber view of an echocardiogram performed 3 weeks postoperatively. The pericardial patch is well seen. Pericardial effusion is not well visualized. RA = right atrium. See Figure 2 legend for other abbreviations.
Pheochromocytoma Crisis, Cardiomyopathy, and Hemodynamic Collapse*

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Pheochromocytoma is a notorious clinical entity. Although suspicion is aroused by severe hypertension in young patients, this sign is often absent. We present a case in which early absence of hypertension and nonspecific signs and symptoms led to failure of prompt diagnosis. The delay proved fatal when the patient developed fulminating pheochromocytoma crisis. This case illustrates a variety of clinical features seen from the vantage of the evolution of the disease as it went unrecognized. The patient’s course underscores the importance of familiarity with the gamut of manifestations for timely diagnosis, and the priority of the latter given the looming risk of overwhelming complications.

Key words: cardiomyopathy; hypertension; marijuana; myocarditis; noncardiogenic pulmonary edema; pheochromocytoma; pheochromocytoma crisis

Pheochromocytoma is a tumor of special renown in clinical medicine. Its hallmark—severe, labile BP elevation—leads to its frequent consideration as a potential etiology in young hypertensive patients, despite the low prevalence of the condition. Yet, pheochromocytoma commonly does not behave in the classic manner, and the nonspecific nature of its manifestations may render prompt recognition elusive. Detailed knowledge of the protean presentations of pheochromocytoma provides the best chance at early detection. Failure may lead to a catastrophic outcome.

CASE PRESENTATION

A 28-year-old man presented to our emergency department with headache, epigastric discomfort, nausea, and vomiting. One year earlier, he had visited another emergency department, complaining of nausea, vomiting, and headache. He was noted to have mild glucose intolerance and was discharged with “gastritis.” Four months later, he came to our emergency department with complaints of a right temporal headache. A diagnosis of sinusitis was made prior to discharge. Two months thereafter, the patient returned after awakening with “heart pounding” and vomiting. He received volume repletion for postural hypotension and was released with “gastritis.” Ten days before admission, recurrent symptoms led to an upper GI series. Detection of mild reflux esophagitis prompted omeprazole treatment.

On the night of admission, the patient noted a throbbing right occipital headache. After playing basketball, he developed bilious vomiting and presented to our emergency department. He complained of lightheadedness, dull abdominal pain, and precordial discomfort without radiating pain or pleurisy. Initial examination showed labile hypertension and epigastric tenderness, but the patient soon became pale, diaphoretic, and dyspneic. Following 100 mL of hemoptysis, he required intubation for hypoxia. Chest radiography revealed diffuse bilateral alveolar infiltrates.

The patient denied smoking or drinking, but admitted to marijuana use. Examination (ICU) showed a sedated, profusely diaphoretic man. BP ranged from 85/55 to 290/170 mm Hg, pulse 140 to 180 beats/min, and temperature 39.1°C. He required 100% fraction of inspired oxygen and positive end-expiratory pressure of 15 mm Hg. A pulmonary artery catheter yielded the following: right atrial pressure, 9 mm Hg; pulmonary artery pressure, 30/22 mm Hg; mean pulmonary capillary wedge pressure, 22 mm Hg; and cardiac index, 2.1 L/min/m². Labs had diffuse cracks bilaterally. There was a soft systolic murmur at the left sternal border. Abdomen was soft; no discrete masses were appreciated. Laboratory values included the following: bicarbonate, 22 mg/dL; anion gap, 18 mg/dL; urea nitrogen, 15 mg/dL; creatinine, 2.7 mg/dL; glucose, 270 mg/dL; creatine kinase, 355 mg/dL; MB fraction, 14.7 mg/dL; and leukocyte count, 14,800 cells/mL. Room-air arterial blood gas measurements (preintubation) revealed a pH of 7.4; Pco₂, 32 mm Hg; Po₂, 61 mm Hg; and bicarbonate, 21 mg/dL. Toxicology screens were positive for tetrahydrocannabinol. ECG showed sinus tachycardia. Echocardiography revealed normal left and right ventricular size and wall thickness, but severe biventricular dysfunction (left ventricular ejection fraction, 20%).

The patient continued to have marked swings in BP and pulmonary capillary wedge pressure requiring titration of sodium nitroprusside. Abdominal ultrasonography (Fig 1) and CT (Fig 2) showed a hemorrhagic 12-cm left suprarenal mass. The patient received normal saline solution, 4 L IV, followed by phenoxybenzamine, 30 mg nasogastrically, and phentolamine boluses of 0.5 mg to 1.0 mg IV, totaling 7 mg. On his second hospital day, however, the patient suffered a terminal bradyasystolic arrest. Spot urine analysis subsequently showed the following: epinephrine, 4.32 mg/L (normal range, 0 to 25 µg/d); norepinephrine, 8.76 µg/L (normal range, 0 to 100 µg/d).

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