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Right Atrioventricular Myxoma Presenting with Recurrent Syncopal Attacks

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Conflict of interest: None declared

Patient: Female, 38-year-old
Final Diagnosis: Myxoma
Symptoms: Dyspena • syncope
Medication: —
Clinical Procedure: Surgery
Specialty: Cardiac Surgery • Cardiology

Objective: Rare disease

Background: Myxomas are rare benign tumors of the heart. These tumors are commonly located in the left atrium, but rarely can affect the right side of the heart. Although it is a relatively rare tumor, it is the most common primary cardiac tumor, accounting for 75-80% of them. Secondary or metastatic cardiac tumors are much more common than primary tumors, accounting for more than 95% of cardiac tumors.

Case Report: A 38-year-old woman presented with shortness of breath and syncope. Upon investigation, she was found to have a right atrioventricular myxoma. It was associated with tricuspid regurgitation, right-sided heart failure, and pulmonary hypertension.

The syncopal attacks and shortness of breath resolved completely after tumor resection. Tricuspid regurgitation (grade 1) and mild pulmonary hypertension (right ventricular systolic pressure 35 mmHg) remained as sequelae of delayed presentation. These may be due to recurrent embolization of tumor fragments to segments of the pulmonary artery.

Conclusions: Cardiac myxomas should be considered in patients presenting with unexplained shortness of breath and syncope. Early diagnosis and tumor resection may prevent serious complications such as pulmonary hypertension and right-sided heart failure.

Keywords: Hypertension, Pulmonary • Jordan • Myxoma • Syncope

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Background

Myxomas are rare benign tumors of the heart, accounting for less than 0.2% of all tumors found in humans [1]. Although it is a relatively rare tumor, it is the most common primary cardiac tumor, accounting for 75-80% of them [2].

The most common location of myxomas is the left atrium (80%), followed by the right atrium (15-20%), right ventricle (2-4%), and left ventricle (2.5%) [3-5]. In the left atrium, myxomas originate from the interatrial septum near the fossa ovalis and are attached to the interatrial septum by a thin stalk [1].

Secondary or metastatic cardiac tumors are much more common than primary tumors, accounting for more than 95% of cardiac tumors [4]. Histopathologically, these tumors originate from multipotent mesenchymal cells of subendocardial connective tissue consisting of fibroblast cells in a background of gelatinous mucoid tissue.

Depending on their location, myxomas can mimic different diseases (ie, mitral stenosis, infective endocarditis, vasculitis, and pulmonary embolism) [6-9]. Here, we report a case of right atrioventricular myxoma presenting late that led to right ventricular (RV) outflow obstruction, pulmonary hypertension, and early manifestations of right-sided heart failure.

Case Report

A 38-year-old female patient without a significant past medical history presented to the emergency department of our hospital in September 2019 with a history of exertional shortness of breath and regular palpitations since 1 month before admission. She had 2 attacks of syncope during this period. She also reported easy fatigability and 8-kg weight loss within the last 2 months.

On physical examination, at the time of admission, she looked healthy without shortness of breath or cyanosis. She was afebrile and normotensive. Her heart rate was 112 beats/min (bpm) and regular in rhythm. O_2 saturation at room air was 93%. Heart examination revealed a pansystolic murmur at the lower left sternal edge that increased with inspiration concordant with tricuspid regurgitation (TR).

Her chest was clear to auscultation. Abdominal examination revealed a firm and tender palpable liver edge 2 to 3 cm below the right costal margin. There was no raised jugular venous pressure or lower limb edema.

Electrocardiography (ECG) showed sinus tachycardia, right bundle branch block, and frequent ventricular ectopic rhythm.

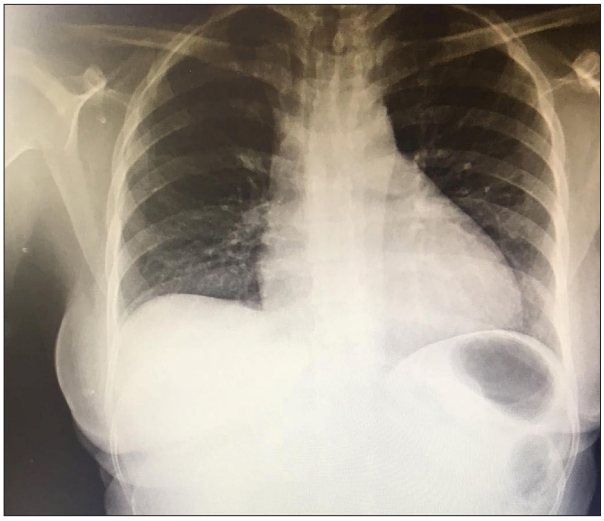

Figure 1. Chest X-ray at the time of admission showing borderline cardiomegaly with clear lung fields.

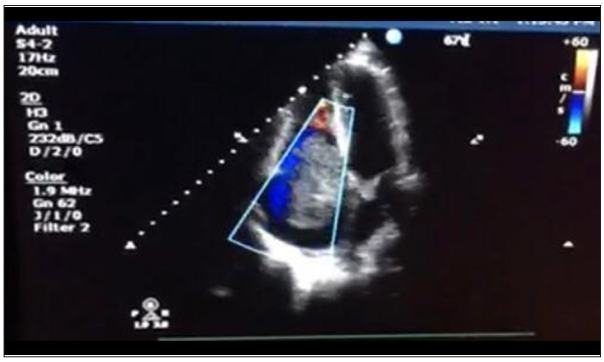

Figure 2. Apical 4-chamber view showing right atrial myxoma prolapsing into the right ventricle.

Holter ECG monitoring (24-h) showed sinus tachycardia reaching 136-140 bpm and frequent ventricular premature complexes. Chest X-ray showed borderline cardiomegaly with clear lung fields as shown in **Figure 1**.

Transthoracic and transesophageal echocardiography (TTE and TEE respectively) revealed a large mass measuring 5.6×3.8 cm in the right atrium originating below the fossa ovalis, affecting the tricuspid valve, leading to severe TR and extending to the RV wall causing RV outflow obstruction and pulmonary hypertension.

The right side of the heart was dilated; RV dimension was 3.2 cm (N=1.2-2.3 cm), with TR grade 2-3 and RV systolic pressure (RVSP) of 50-55 mmHg (n=8-20 mmHg) as shown in **Figure 2**.

Laboratory workup showed the following values: hemoglobin (Hb) 11.3 g/dL (n=12-14 g/dL); erythrocyte sedimentation rate (ESR) 53 mm/h (n=13-27 mm/h); C-reactive protein 145 mg/L (N=0-5); international normalized ratio 1.6 (n=0.9-1.2);

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alanine and aspartate transaminases 61 and 94 IU/L, respectively (n=35-38 IU/L), serum albumin 26 g/L (n=35-52 g/L), and brain-type natriuretic peptide 822 pg/mL (n<100 pg/mL).

Liver ultrasound revealed a homogenously enlarged liver (18 cm) with no focal defects.

Thyroid function tests, fasting blood sugar, blood urea nitrogen, urinalysis, and serum lipids were within normal limits. Hepatitis markers hepatitis B surface antigen and hepatitis C virus antibodies were nonreactive. Also, human immunodeficiency virus antibodies were negative.

An initial diagnosis of cardiac myxoma was made and surgical resection of the mass was planned as soon as possible.

The patient underwent cardiac surgery after 4 days of admission. Intraoperatively, 400 mL of pericardial effusion was found and drained. The right atrium and right ventricle were enlarged. A 12×8-cm huge dark-red to gray color, vascularized, gelatinous mass filling both the right atrium and the right ventricle was found. It was attached to the interatrial septum below the fossa ovalis by a thin stalk. It was extending down through the tricuspid valve with part of the mass adherent to the interventricular septum. We delivered and freed the mass, identified and resected the stalk, and then cauterized the base of the stalk.

The patient came out of cardiopulmonary bypass without the need for direct-current shock. She was intubated for 12 h postoperatively and kept in the intensive care unit for 24 h, where she was given 1 unit of packed red blood cells and 5 units of fresh frozen plasma.

The mass was sent for histopathologic examination and showed macroscopically a gray-colored mass measuring 9×4.5×3 cm. Gross sectioning showed myxoid areas with other areas of hemorrhage and necrosis. Microscopic examination showed stellate myxoma cells with abundant eosinophilic cytoplasm in the background of abundant mucopolysaccharide (myxoid) ground and areas of hemorrhage and necrosis and no evidence of malignancy. The intrapericardial drain was removed on the third postoperative day and the patient was discharged without any complications.

She was seen in the clinic at 1-month, 2-month, 5-month, and 1-year intervals postoperatively. She was doing well without any complaint. Her liver transaminases returned to normal levels. Repeated TTE 5 months and nearly 1 year postoperatively was free of any cardiac masses. It showed grade 1 TR; the right atrium and right ventricle were still dilated, with RVSP of 35 mmHg, as shown in **Figure 3**.

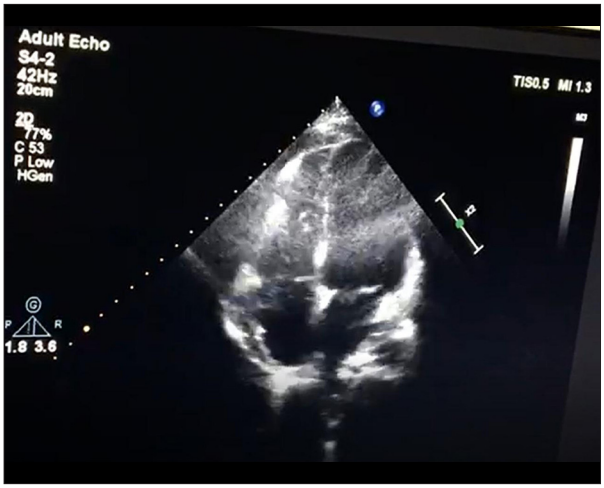

Figure 3. Apical 4-chamber view with color flow mapping, postoperatively, showing grade 1 tricuspid regurgitation.

Discussion

Myxomas are rare benign tumors of the heart, accounting for 5% of all tumors affecting the heart. The rest are metastatic tumors [1,4]. The most common location of myxomas is the left atrium (80%) [4], but rarely they can be found in the left ventricle, right atrium, or right ventricle, in that order of frequency [4,5].

The clinical features of myxomas are determined by their location, size, mobility, and fragility. Myxomas of the left side present with obstructive symptoms (orthopnea and paroxysmal nocturnal dyspnea and even pulmonary edema) mimicking mitral valve stenosis or congestive heart failure. When systemic embolization occurs, patients may present with acute stroke, brain abscess, or peripheral emboli. Recurrent pulmonary embolization may lead to pulmonary hypertension.

Patients with tumors on the right side of the heart may present with shortness of breath and easy fatigability, as in our case, due to pulmonary hypertension and right-sided heart failure. Embolization of right atrial myxomas may lead to pulmonary embolism [6,7]. Constitutional symptoms may include fever, loss of weight, anemia, and high ESR, mimicking infective bacterial endocarditis or systemic vasculitis [8,9]. These symptoms occur as a result of production and release of cytokines, interleukin-6, and interleukin-8 [10]. In our case, the presence of high-grade TR and pulmonary hypertension, along with anemia and weight loss, indicates that the tumor was there for a long time.

Initial investigations (ie, ECG and chest X-ray) are usually not helpful in establishing the diagnosis. The wide availability of echocardiography nowadays made the diagnosis ready and

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easy. Echocardiography (TTE and TEE) is also helpful for preoperative evaluation of the tumor regarding the size, location, and associated cardiac complications (pulmonary hypertension, TR, and right-sided heart failure, as in our case). TEE has superior diagnostic ability for cardiac tumors to TTE because of the proximity of the esophagus to the heart, the lack of intervening lung and bone, and the ability to use a high-frequency imaging transducer that affords superior special resolution.

When the diagnosis is made, the definite line of treatment is surgical resection of the tumor as early as possible to prevent serious complications such as pulmonary or systemic embolization or even sudden death. Outcomes of early surgery are largely favorable, with a mortality rate less than 5% [11-13].

Postoperative recurrence is rare and occurs in about 2-5% of cases reported in familial forms, or in cases of multicentric tumors due to incomplete resection of the tumor stalk [13]. Recurrence may happen 5 to 6 years after tumor resection [14]. The constitutional signs and symptoms of cardiac myxomas improve after resection of the tumor, as in this case [14].

Conclusions

Cardiac myxomas should be considered in patients presenting with unexplained shortness of breath and syncope. Early diagnosis and tumor resection may prevent serious complications such as pulmonary hypertension and right-sided heart failure.

Conflict of Interest

None.

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