Case Report

Left atrial myxoma swinging like a ball valve thrombus: "The wrecking ball effect"

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ABSTRACT

Left atrial myxoma is the most common benign primary tumor of the heart, more common in females, and often presents with non-specific symptoms with incidental detection in echocardiography. The bimodal mode of presentation of the left atrial myxoma is as follows: one group of patient's presents with symptoms mimicking infective endocarditis and the other group of patient's presents with symptoms of typical mitral stenosis. Large left atrial myxoma acting as a ball valve thrombus causing complete obstruction of the mitral valve and causing pulmonary venous hypertension with acute pulmonary edema has not been described in the literature so far. We report a rare case of large left atrial myxoma swinging like a ball valve thrombus across the mitral valve in a 76-year-old female presenting with acute pulmonary edema and desaturation. The patient underwent emergent surgical excision of the left atrial myxoma which relieved the pulmonary edema and desaturation as well. Left atrial myxoma being completely occlusive led to passive pulmonary venous hypertension and acute pulmonary edema: a rare phenomenon to be observed in a clinical scenario.

Key words: Atrial, Ball, Myxoma, Thrombus

Atrial myxomas are the most common primary tumors of the heart accounting for 40–50% of all cardiac tumors. Early diagnosis remains a challenge in atrial myxomas because of associated non-specific symptoms. Approximately, 90% of atrial myxomas are sporadic in origin, solitary, and pedunculated; 75–85% of atrial myxomas occur in the left atrial cavity and approximately 25% of those occur in the right atrium (RA). Approximately, 10% of atrial myxomas are familial and transmitted in an autosomal dominant fashion [1]. Familial atrial myxomas are frequently multiple and are more frequently located in the ventricle (13% vs. 2% in sporadic cases). The peculiarity of presentation of atrial myxoma is that they often present with symptoms with a change in the body position.

General symptoms of atrial myxomas mimic that of infective endocarditis; they present with fever, malaise, cachexia, chest pain, or tightness [2]. Approximately, 20% of atrial myxomas can be completely asymptomatic. Symptoms of the left atrial myxoma often mimic mitral stenosis in the form of cough due to pulmonary venous stasis, palpitation due to associated atrial fibrillation, exertional shortness of breath (75%), paroxysmal nocturnal dyspnea, pulmonary edema, hemoptysis, and dizziness or syncope (20%) secondary to cerebral embolization.

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Few patients of the left atrial myxoma present interestingly with platypnea, that is, patient feels difficulty in breathing in an upright position with relief in the supine position. Pedunculated left atrial myxoma swings like a "ball attached to a thread" across the mitral valve known as the "wrecking ball effect" which was typically observed in our case and the left atrial myxoma being completely occlusive and acting like a ball valve thrombus causing pulmonary venous hypertension and flash pulmonary edema amounting to urgent surgical removal is a rare phenomenon to be described in the literature so far.

CASE REPORT

A 76-year-old elderly female presented to the emergency department with complaints of acute onset shortness of breath and orthopnea with systemic desaturation (SpO_2 74%) for the last 1 h.

On clinical examination, she had a pulse rate of 118 beats/min, regular in nature, and blood pressure of 100/60 mm Hg in the right arm supine position. She had coarse leathery crepitations all over both lung fields (>50% of bilateral lung fields) suggestive of acute pulmonary edema and had early diastolic murmur (tumor plop) over the precordium which was changing in intensity with changing patient's position. Tumor plop in atrial myxoma is due to halt in excursion of tumor mass during early diastole and

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impingement of the tumor mass with endocardium during early diastole.

Serum chemistry in our case revealed increased ESR of 80 mm/h, anemia of chronic diseases with hemoglobin of 8.4 gm/dl, leukocytosis (total leukocyte count 12000/dl), and increased C-reactive protein of 8 ng/ml and normal renal profile. Increased interleukin (IL)-6 levels in atrial myxoma are an indirect clue to the histological variety of cardiac tumors as atrial myxomas produce more IL6 which is responsible for producing Raynaud's phenomenon in 50% of cases.

Chest X-ray revealed acute pulmonary edema with classical "Bats Wing" appearance of the pulmonary hilum. Electrocardiogram (ECG) revealed left atrial enlargement and echocardiography revealed a pedunculated left atrial myxoma variegated in the appearance of size 62 mm×24 mm attached to the mid interatrial septum with a narrow stalk and swinging like a ball valve thrombus across the mitral valve being completely occlusive (Fig. 1) causing acute pulmonary venous stasis, pulmonary edema, and desaturation.

The patient was managed with non-invasive ventilation and intravenous diuretic which improved oxygen saturation to 92% and the patient was immediately taken for emergency cardiac surgery for surgical resection of the tumor mass to relieve the desaturation. The atrial myxoma was excised with the standard *operative* technique for dealing with a *cardiac myxoma* with a median sternotomy approach which allows the removal of the mass under cardiopulmonary bypass (CPB) under general anesthesia. On arrival into the operating room, the patient was placed in a 15° left tilted supine position, a large-bore peripheral IV line was inserted, and the standard monitoring for cardiac surgery of our institution was applied (5 leads ECG, femoral arterial catheter, and state entropy).

General anesthesia of the patient was induced after 3 min of preoxygenation and ventilation by means of a Sellick maneuver, using target-controlled intravenous anesthesia with a combination of Remifentanil (Minto model) and Propofol (Schneider model) to ensure a level between 40 and 60 state entropy. After intubation and mechanical ventilation set up, an ultrasound-guided right internal jugular catheter and a bladder catheter were placed. The mean arterial blood pressure target before CPB was set at 70 mmHg. Anticoagulation therapy by heparin (400 U/kg) was administered before aorta cannulation.

Surgery was initiated by a median sternotomy, and CBP was established using ascending aorta-RA bicaval cannulation.

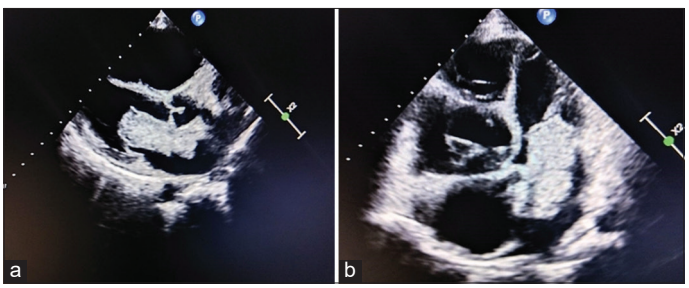

Figure 1: (a) Left atrium myxoma like a ball valve thrombus across the mitral valve; (b) variegated appearance of completely occlusive left atrium myxoma

Perfusion strategy consisted in pulsatile flow, ranging from 2.9 to 3.1 L/min/m^2 (theoretic cardiac index = 2.8 L/min/m^2), and high perfusion pressure (66–81 mmHg) under normothermic conditions (36–37°). A gradual transition from corporeal to fully extracorporeal circulation was realized. The right atrium was opened along the atrioventricular groove. The interatrial septum was incised at the lower border of the fossa ovalis. A hanging stitch was placed and the interatrial septum was excised keeping 0.5–1 cm from the stalk of the tumor. The tumor was removed easily without any adhesion with the residual atrial wall or mitral valve. The atrial septal defect was closed directly using continuous polypropylene 4/0 running sutures. The right atrial wall was closed. All wounds were closed and one chest drain was placed in the right pleural cavity. Total operative time was 4 h; and total bypass time was 150 min, respectively. Intraoperative transesophageal echocardiography (TEE) showed good results; there was no residual atrial defect and a resultant good competent mitral valve.

The patient was extubated the next morning 12 h after the operation; then she had a normal recovery with no significant complications. The chest drain was removed on the 2nd post-operative day and the chest film was normal. The patient was discharged after 7 days in a stable hemodynamic condition with advice to follow-up after 1 month.

DISCUSSION

Myxomas have been described across all ages from 30 to 83 years; the mean age of presentation of atrial myxomas is 53 years in sporadic cases and 25 years in familial cases. About 75% of sporadic atrial myxomas occur in females [2]. Familial atrial myxomas have no female sex predilection. Our case was unique in presentation as the patient was of 76 years with index presentation of flash pulmonary edema with desaturation.

Echocardiography revealed the mass to be polypoid in nature as myxomas can be polypoid, round, or oval. They are gelatinous, white, yellowish, or brown in color with a smooth or lobulated surface. Although myxomas most commonly originate from fossa ovalis, they can originate also from the posterior atrial wall, anterior atrial wall, or atrial appendage. Myxomas become more symptomatic when they are more than 5 cm in diameter. The friable nature of the myxomas accounts for frequent tumor embolization which occurs in 30–40% of cases. Ha and associates reported a more frequent occurrence of systemic embolism in polypoid tumors as compared to round ones (58% vs. 0%) [3].

Myxomas are vascular tumors and may also be neovascularized by a branch of the coronary artery [4]. Hemorrhage in atrial myxoma has also been reported secondary to profound neovascularization [5]. Myxomas express an increased amount of vascular endothelial growth factor and IL-6 [6-8]. Carney syndrome [9,10] accounts for 7% of all atrial myxomas due to abnormality of the short arm of chromosome 2 (Carney) where multiple atrial myxomas may be found in both atria and ventricles.

Atrial myxomas acting as a ball valve thrombus may lead to sudden cardiac death, occlusive atrial myxoma, and acute cerebral embolism accounting for sudden cardiac death in myxomas

in 1.5% of cases. The right-sided atrial myxomas present with typical signs of right heart failure with raised jugular venous pressure, ascites, pedal edema, and increased fatiguability. The patients with familial myxoma may have a variety of features known as syndrome myxoma or Carney syndrome [11] which constitutes the following: Cutaneous myxomas, lentigines (i.e., flat brown discoloration of the skin), endocrine hyperactivity, that is, cushing syndrome and multiple cerebral fusiform aneurysms. Other described syndromes associated with atrial myxomas include the following: NAME syndrome with features of nevi, atrial myxoma, myxoid neurofibroma, and ephelides (i.e., freckles [tanned macules found on the skin]) and LAMB syndrome with features of lentigines, atrial myxoma, and blue nevi. The risk of recurrence is higher in familial myxoma syndrome [12].

Differential diagnosis of the left atrial myxoma includes mitral stenosis, tricuspid stenosis, pulmonary embolism, and idiopathic pulmonary hypertension. Myxoma also damages the valve leaflets and annuli causing mitral regurgitation if it is left-sided and tricuspid regurgitation if it is right-sided which also stand as a differential diagnosis in rare cases.

Echocardiography remains as the gold standard to diagnose an atrial myxoma. An atrial myxoma must be differentiated from a left atrial thrombus; the thrombus is usually situated in the posterior portion of the atrium and has a layered appearance. The presence of a stalk and mobility also favors atrial myxoma. TEE has better specificity and 100% sensitivity compared to trans-thoracic echocardiography in diagnosing atrial myxoma. TEE is usually advisable for myxoma syndrome, as there may be multiple less-common sites with minuscule myxomas which can be better picked up in TEE. The point of attachment of atrial myxoma is best visualized by cardiac magnetic resonance imaging. Fluorodeoxyglucose-positron emission tomography scanning is not typically indicated in the evaluation for myxoma which reveals hypermetabolic hypodense areas [13].

Cardiac catheterization is usually performed to exclude coexistent coronary artery disease in patients older than 40 years and in our case, elderly female had normal coronaries. If petechiae are present, a skin biopsy may reveal the presence of spindle-shaped, myxomatous, endothelial-like cells with round or oval nuclei, and prominent nucleoli. Histologic studies of atrial myxomas are characterized by the presence of lipidic cells embedded in a vascular myxoid stroma [14]. Lipidic cells characteristically polygonal to stellate in shape with scant eosinophilic cytoplasm. We treated the patient with emergency surgical resection due to the complete occlusion of atrial myxoma. Conventional treatment of atrial myxoma is surgical removal by median sternotomy and the parietal pericardium was used to close the surgical defect.

Minithoracotomy with robotically-assisted surgery results in a shorter length of hospital stay and is considered a safe and feasible method for atrial myxoma excision [15]. A case report of total endoscopic robotic resection of a left atrial myxoma in an elderly patient with persistent left superior vena cava also demonstrated a good outcome [16]. Gur and Aykac recommend resection of the myxoma with the surrounding healthy margin of minimum of 5 mm to prevent recurrence, followed by annual echocardiography to assess for possible relapse [17]. Excision of atrial myxoma is associated with early post-operative mortality of 2.2%. Post-operative atrial fibrillation is seen in 23–33% of patients [18,19]. Independent predictors of outcomes included advanced age, left atrial diameter, and mitral valve surgery. Because of the risk of tumor fragmentation and embolization, vigorous palpation or manipulation of the myxoma should be performed only after cardioplegia.

The recurrence rate in sporadic cases is 1–5% while the recurrence in familial cases is 20–25% and primary myxoma occurring at atypical cases is more likely associated with recurrence [20]. Younger age at diagnosis, smaller tumor dimension, and the localization of the tumor to the ventricles were predictors of recurrence [21]. Recurrence is usually attributed to incomplete excision of the tumor, growth from a second focus, or intracardiac implantation from the primary tumor. Damaged valves may require annuloplasty or prosthetic replacement.

CONCLUSION

We represent a rare case of large atrial myxoma swinging like a ball valve thrombus across the mitral valve presenting as flash pulmonary edema which was successful treated with emergency surgical excision. Interesting presentation of a large left atrial myxoma acting as a ball valve thrombus – can be rather regraded and coined as a “Ball Valve Tumor.” Diversity is the rule in the presentation of nature for ages.

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