



Case Report: The Myxomatous Mystery

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Introduction

Primary cardiac tumors are rare with an incidence rate of 1.38 new cases per 100,000 individuals per year. Of these tumors, 75% are benign in origin and the most common type is a myxoma with a frequency of over 50%. Myxomas most commonly arise from the left atrium followed by the right atrium. Clinical presentation of a myxoma varies depending on the location, size, and shape of the tumor. For example, a patient diagnosed with a myxoma may have constitutional symptoms, cardiovascular symptoms, pulmonary symptoms, etc., or may be entirely asymptomatic. Since there are no specific symptoms presented by patients with a myxoma, physicians often face challenges in diagnosing.

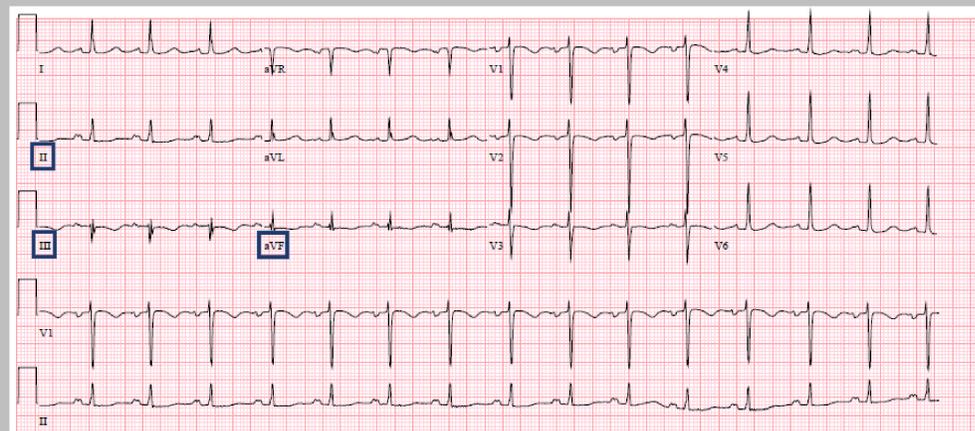
Case Report

A 72-year-old female with a past medical history of hypertension presented to the Emergency Department (ED) with progressive dyspnea on exertion of 1-week duration. She reported chest tightness, orthopnea, and paroxysmal nocturnal dyspnea. She denied experiencing similar symptoms in the past. Vital signs were unremarkable except for her blood pressure which was 178/103 mm Hg. Pertinent physical exam findings included jugular venous distension, a S3 heart sound, and bibasilar rales. Laboratory tests were ordered and all were unremarkable, including her troponin I and BNP levels. ECG was significant for T wave inversion in the inferior leads. Chest x-ray and CT were ordered and showed diffuse ground glass opacities throughout both lungs, bilateral pleural effusions, and mild cardiomegaly, as well as dilation of the main pulmonary artery. Lasix was given as patient appeared to be volume overloaded, which resulted in brisk diuresis and relief of symptoms. At this point, an acute exacerbation of left-sided heart failure was suspected and a transthoracic echocardiogram (TTE) was ordered. TTE revealed a large ovoid mobile echodense mass. The mass was 5.1 cm x 2.9 cm and attached to the lower half of the interatrial septum on the left atrium and moved in and out of the left ventricular inflow. This finding was most consistent with a left atrial myxoma and the patient was then transferred to a medical center capable of resecting the myxoma. The patient tolerated the myxoma excision well and was discharged to cardiac rehabilitation. The surgical pathology report confirmed cardiac myxoma with central hemorrhage.

Transthoracic ECHO



Electrocardiogram



Discussion

In the setting of predisposed hypertension, we suspect that the large size and mobile nature of the left atrial myxoma caused functional partial obstruction of the mitral valve, leading to secondary valvular heart disease. Our patient had normal BNP levels on presentation, and on TTE, had normal left atrial and ventricular sizes, a normal left ventricular ejection fraction of 60-65%, and only mild mitral regurgitation. However, the obstruction of the mitral valve was significant enough to raise pulmonary artery pressure to 38 mm Hg and cause progressive left-sided heart failure symptoms; the patient was presenting with both dyspnea on exertion and bilateral pleural effusions. The presence of mildly reduced right ventricular systolic function and mild tricuspid regurgitation suggests that right ventricular remodeling would have been inevitable in our patient if the myxoma remained untreated.

In a systematic review of 112 cases of left atrial myxomas, symptoms of mitral valve obstruction, the first arm of the classic triad of myxoma presentation, were present in 75 patients (67%), with mostly cardiac failure or malaise. Symptoms of embolism, the second most frequent presentation in the classic triad, were observed in 33 cases (29%). The third arm of the classic triad consisted of constitutional symptoms (34%) with fever, weight loss, or symptoms resembling connective tissue disease, due to cytokine secretion. The most frequent electrocardiographic sign discovered was left atrial hypertrophy (35%), whereas arrhythmias were uncommon. A review of laboratory data also suggested anemia and an elevated ESR, CRP, or globulin level could be present. Our patient displayed symptoms of mitral valve obstruction and mild cardiomegaly with unremarkable bloodwork, prompting the team to explore a diagnosis of acute heart failure originally. The long-term prognosis of left atrial myxomas is excellent and the recurrence rate is low. Long-term follow-up and serial echocardiography are advisable especially for young patients, and this was the recommendation for our patient as well.

Conclusion

Despite the rarity of the disease, myxomas, as well as other cardiac tumors, should always be included in the differential diagnosis when a patient presents with symptoms suspected of an acute exacerbation of heart failure. Echocardiogram is the diagnostic modality of choice to utilize when a myxoma is suspected. Once the diagnosis is confirmed, myxomas must be surgically resected in a timely manner in order to prevent complications of systemic embolization and other cardiovascular diseases, which may lead to sudden death.

References

1. Cresti A, Chiavarelli M, Glauber M, Tanganelli P, Scalese M, Cesareo F, Guerrini F, Capati E, Focardi M, Severi S. Incidence rate of primary cardiac tumors: a 14-year population study. *J Cardiovasc Med (Hagerstown)*. 2016 Jan;17(1):37-43. doi: 10.2459/JCM.0000000000000059. PubMed PMID: 25022931.
2. Gaasch WH, Vander Salm, TJ. Cardiac tumors. Colucci, WS, Vora, SR, Yeon, SB (Ed.), 9 Aug 2017. <https://www.uptodate.com/contents/cardiac-tumors>.
3. Paraskevaidis IA, Michalakeas CA, Papadopoulos CH, Anastasiou-Nana M. Cardiac tumors. *ISRN Oncol*. 2011;2011:208929. doi: 10.5402/2011/208929. Epub 2011 May 26. PubMed PMID: 22091416; PubMed Central PMCID: PMC3195386.
4. Pinede L, Duhaut P, Loire R. Clinical presentation of left atrial cardiac myxoma. A series of 112 consecutive cases. *Medicine (Baltimore)*. 2001 May;80(3):159-72. Review. PubMed PMID: 11388092.
5. Vasan RS, Wilson PW. Epidemiology and causes of heart failure. Colucci, WS, Yeon SB (Ed.), 20 Mar 2018. <https://www.uptodate.com/contents/epidemiology-and-causes-of-heart-failure>.