Medical management of a suspected atrial myxoma in a nonagenarian

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Abstract

Cardiac myxomas are the most frequently encountered tumors of the heart. However, they are unusual to be newly diagnosed in the geriatric population. Myxomas are among the great mimickers, with a myriad of clinical presentations related to heart failure, embolic events, and constitutional symptoms. We describe a rare case of a giant atrial myxoma in a nonagenarian presenting with heart failure, which was medically managed.

Keywords

Atrial myxoma, nonagenarian, heart failure, polypharmacy

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Introduction

Myxomas are the most frequently encountered intra-cardiac tumors, representing 50% of primary cardiac tumors. They are generally located in the left atrium (85%) and predominantly originate from the interatrial septum, near to the *fossa ovalis*.^{1,2}

They have a female predilection, usually manifesting in middle-age to late adulthood. Some common differential diagnoses include pedunculated thrombus, metastatic sarcoma, and melanoma.³ Despite myxomas being relatively rare in the elderly population, the incidence has steadily increased due to longer life expectancy coupled with more frequent imaging studies being performed.⁴

Patients can be relatively asymptomatic or symptomatic based on tumor location and burden. The classical triad of obstructive physiology, embolization, and constitutional signs account for a myriad of clinical presentations, including heart failure, arrhythmias, and sudden cardiac death.⁵ Geriatric patients often present with non-specific symptomatology that can often go unnoticed, thus, delaying accurate diagnosis.

We describe a rare case of a giant atrial myxoma in a nonagenarian presenting with heart failure, which was medically managed.

Case report

A 94-year-old South Asian female with a medical history of atrial fibrillation with a prior cerebrovascular event (stroke) approximately 2 months prior, chronic kidney disease stage

3b and moderate dementia initially presented (March 2019) with severe dyspnea and worsening pedal edema during the preceding week. Her vital signs indicated systolic blood pressures of 160 s mmHg, heart rate of 143 beats per minute, respiratory rate of 24 breaths per minute with an oxygen saturation of 92% on room air. Her physical examination revealed atrial fibrillation, an elevated jugular venous pressure of 12 cm H₂O, an S₃ gallop with occasional bibasilar crackles and moderate pitting edema. A 12-lead electrocardiogram confirmed atrial fibrillation with a rapid ventricular response and non-specific ST-T changes. Portable chest radiography revealed florid pulmonary edema with diffuse Kerley B lines and prominent hilar congestion. Pertinent diagnostic laboratory investigations included a d-dimer 323 ng/dL (normal $\leq 500 \text{ ng/mL}$), pro-brain natriuretic peptide 3468 pg/mL (normal $\leq 300 \text{ pg/mL}$), cardiac biomarkers, CK-MB 7 U/L (normal \leq 20 U/L), troponin I 0.03 ng/

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Figure 1. Two-dimensional transthoracic (TTE) series: (a) 2D-TTE parasternal long-axis view displaying the atrial myxoma during diastole (enveloped by the red circumferential border), (b) 2D-TTE parasternal long-axis magnified/zoomed-in view illustrating the extent of tumor burden and the myxoma's attachment to the left atrial-free wall, and (c) 2D-TTE parasternal long-axis view illustrating the tumor dimensions of $59 \text{ mm} \times 43 \text{ mm}$ in the top left corner.

mL (normal ≤ 0.15 ng/dL). Routine investigations revealed a hemoglobin of 9.2 g/dL (normal 12–15 g/dL) with a normal white cell count and serum creatinine of 1.6 mg/dL (normal 0.6–1.2 mg/dL). She was initiated on renally dosed enoxaparin, low-dose beta-blockade, and diuresis with a furosemide bolus, and infusion in the emergency department and subsequently admitted for further hospitalization.

Bedside transthoracic echocardiography demonstrated a large $59 \text{ mm} \times 43 \text{ mm}$ mass arising from the left atrial free wall (see Figure 1(a)–(c)). There was preserved left ventricular systolic function (ejection fraction of 65%) without regional wall motion abnormalities. In addition, there was moderate left atrial enlargement (38 mL/m^2) with severe diastolic dysfunction and restrictive physiology with an estimated left atrial pressure of 22 mmHg and moderate pulmonary hypertension of 52 mmHg. There was no overt prolapse of the myxoma through the mitral valve orifice during the time of the study.

Due to the patient's advanced age, high clinical frailty scale, and moderate dementia, a non-invasive, conservative medical strategy was adopted after achieving a consensus among family members. Diagnostic coronary angiography, advanced imaging with computed tomography and magnetic resonance modalities, and open-heart surgical excision were deferred. She was instituted on daily low-dose apixaban, bisoprolol, rosuvastatin, amlodipine with hydralazine and nitrates, and furosemide to attenuate polypharmacy while maintaining euvolemia.6-10 Sacubitril/valsartan and spironolactone were avoided due to her chronic kidney disease and concern of precipitating electrolyte derangements such as hyperkalemia. Ivabradine was not utilized as the patient had chronic atrial fibrillation. The patient's ensuing 4-day hospitalization course was uneventful, and her blood pressures and heart rate gradually normalized. She was safely discharged on an individualized medical regimen with routine follow-up appointments and full-time home-health nursing (see Figure 2). The patient was asymptomatic and hemodynamically stable at her



Figure 2. Censored photograph of the patient a few days after hospital discharge. The white arrow indicates that her pedal edema has since mostly resolved.

3-month appointment (June 2019); however, unfortunately, expired during sleep (October 2019) before her 6-month appointment (after her index hospitalization), and the family deferred a post-mortem.

Discussion

This case is unique, as to the best of the authors' knowledge; it is the eldest patient described to have a suspected atrial myxoma. Unfortunately, it could not be verified by histopathology or imaging modalities such as computed tomography or cardiac magnetic resonance imaging (cMRI) due to the conservative approach. The tentative diagnosis was entirely reliant on transthoracic echocardiography, which, albeit possesses a 95% sensitivity.^{2,11} This is considered the first-line diagnostic test in clinching a prompt and accurate diagnosis, whereas its more invasive counterpart, transesophageal echocardiography, provides 100% sensitivity. Cardiac MRI is rapidly gaining traction as a novel imaging modality in the assessment of cardiac myxomas; however, was not performed in this case due to patient's claustrophobia. It provides superior tissue characterization by differentiating myxomas from thrombus, and benign from malignant lesions, without exposure to ionizing radiation.^{12,13}

The patient's overall presentation was consistent with that of heart failure with preserved ejection fraction (HFpEF) exacerbation. The etiology was considered multifactorial and likely contributed by a pseudo-stenotic effect of the myxoma, hypertensive urgency, and atrial fibrillation with a rapid ventricular response, which may or may not have been related to the myxoma. Dyspnea and palpitations were the most common symptoms on index presentation, being 70% and 35%, respectively.¹⁴ Our patient had atrial fibrillation, which was coincident in almost 30% of patients.^{5,15} The emergence of heart failure is contingent on tumor location and burden in creating a pseudo-stenotic effect due to malapposition of mitral valve leaflets and obstructive physiology within the left ventricular outflow tract.¹⁶ Also, it has been postulated that a paraneoplastic cytokine milieu may induce myocardial inflammation and resultant ischemia.17,18 Coronary embolization of tumor fragments may also be implicated in the pathogenesis of incident heart failure; however, angiography was deferred in preference to a non-invasive approach. Our patient did also report a plethora of non-specific constitutional symptoms, which can often mimic rheumatologic disease and be overlooked, especially in the geriatric population, obfuscating the eventual diagnosis.17,19,20

Due to the advent of novel imaging technologies and, generally, longer life expectancy, the incidence of myxomas in the geriatric population has increased.²¹ Atrial myxomas presenting in the seventh decade is rare, with only a paucity of published case reports. At the turn of the last century, Bire et al.²² discovered only 19 confirmed cases of septuagenarians and elder with myxomas. An updated review reported just more than 60 individuals with myxoma, with their ages ranging from 68 to 88, and the eldest patient undergoing successful resection being a Japanese man, aged 90.^{4,23} In an older series of 100 individuals with atrial myxoma, almost one-fifth were aged 70 and older.²⁴ With respect to geriatric patients, a recent study evaluated elective surgery in elderly

patients with myxomas.¹⁵ During the time frame, 1985–2012, 17 consecutive patients (mean age ~69 \pm 4 years) were evaluated. Overall, 13 patients survived resection with a relatively high mortality rate of nearly 25%. The patient's calculated EuroSCORE II was approximately 8%.²⁵ In light of this information, and the fact that our patient was almost 20 years older than the mean study population, a non-invasive management plan was adopted.

The patient also had a medical history of the prior cerebrovascular event 2 months earlier, which could have possibly resulted from systemic embolization (10%–45%) from the atrial myxoma or a cardioembolic attributed to atrial fibrillation, which in turn could be sequelae of the myxoma itself. As there was no mitral valve disease, the patient was initiated on low-dose daily apixaban for direct oral anticoagulation, given her chronic kidney disease 3b and HAS-BLED score of 4, clinically translating to a near 9% risk of a major bleeding event.^{26,27} As the patient possessed several features for excessive polypharmacy such as frailty, multimorbidity, and dementia, meticulous care was taken in individualizing evidence-based therapies with minimal drug–drug interactions.^{28,29}

Conclusion

We describe a rare case of a giant atrial myxoma in a nonagenarian, which was medically managed. Myxomas are among the great mimickers, with a myriad of clinical presentations and should be considered as a differential diagnosis for heart failure in a geriatric population.

Key clinical message

The clinician should be aware of atrial myxoma as an etiology for heart failure in geriatric patients.

Authors' contributorship

J.K., R.S., V.S., S.P., S.M., and N.A.S. all contributed equally in writing the manuscript. All authors read and approved the final manuscript.

Compliance with ethics guidelines and standards

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

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The patient's power of attorney (the patient has moderate to severe dementia) has provided written, informed consent to have the details of her case published. Institutional approval was not required for publication.

Data sharing statement

All available data can be obtained by contacting the corresponding author.

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Ethics approval

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