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Cerebrovascular Accident Caused by Embolic Atrial Myxoma

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Background

Atrial myxomas, the most common type of cardiac tumors, can cause life-threatening complications. As most cardiac myxomas are surgically curable, early diagnosis is crucial. Cardiac tumors can present with cardiac and embolic manifestations, and should be considered in the differential diagnosis of patients presenting with such symptoms. In this case report, we describe a young, healthy patient who presented with stroke symptoms secondary to embolic atrial myxoma.

Case Presentation

A 45-year-old male with no past medical history presented with an acute onset of dysarthria, described as garbled speech. He also experienced weakness and heaviness of his right hand, which impaired his ability to brush his teeth and write. He denied left-handed or lower extremity weakness, head trauma, facial droop, dysphagia, migraines, diplopia/visual changes, confusion, or memory loss. He denied tobacco, alcohol, or drug use. Review of systems was otherwise negative, although the patient reported that whenever he participated in strenuous physical activity over the past few years, he would become nauseated.

Investigations

The patient initially presented to an outside hospital after his symptoms persisted for over 24 hours. Computed tomography (CT) of the head was negative for intracranial bleed and he was discharged. While his dysarthria improved, he continued to have weakness and went to another hospital for evaluation. Brain magnetic resonance imaging (MRI) at the second hospital showed two acute left basal ganglia infarcts. As part of the stroke workup, he had an echocardiogram, which revealed a 5 cm left atrial myxoma. He was transferred to Thomas Jefferson University Hospital (TJUH) for further management.

On presentation to the TJUH Cardiac Critical Care Unit (CCU), the patient's vital signs were as follows: temperature 98.2°F, blood pressure 142/80 mm Hg, pulse 100 beats per minute, respiratory rate 19 breaths per minute. Cardiac examination was notable for a soft diastolic rumble heard best in the left upper parasternal area. Neurological examination revealed minor dysarthria. The remainder of the physical examination was normal. Complete blood count and basic metabolic panel were within normal limits, and electrocardiogram showed sinus rhythm. C-reactive protein (CRP) was elevated to 7.9 mg/dL. Chest radiograph was unremarkable. CT head showed left basal ganglia and corona radiata hypodensities consistent with recent infarction. CT thorax revealed a hypoattenuating 3.7 x 2.4 cm lesion in the left atrium. Transesophageal echocardiogram (TEE) confirmed a large (5 x 3.2 cm), irregular, gelatinous mass, with frond-like, mobile edges attached to the fossa ovalis, and broad 1 cm stalk. This mass prolapsed into the mitral valve orifice during diastole, likely representing a left atrial myxoma.

Treatment

Four days after presentation to TJUH, the patient underwent robotic-assisted excision of the myxoma. There were no intraoperative complications, and post-procedure TEE revealed resolution of the left atrial myxoma without mitral valvular insufficiency. The patient required pressors and inotropic support for 2 days post-operatively for persistent hypotension. He was discharged home in stable condition on post-operative day #3.

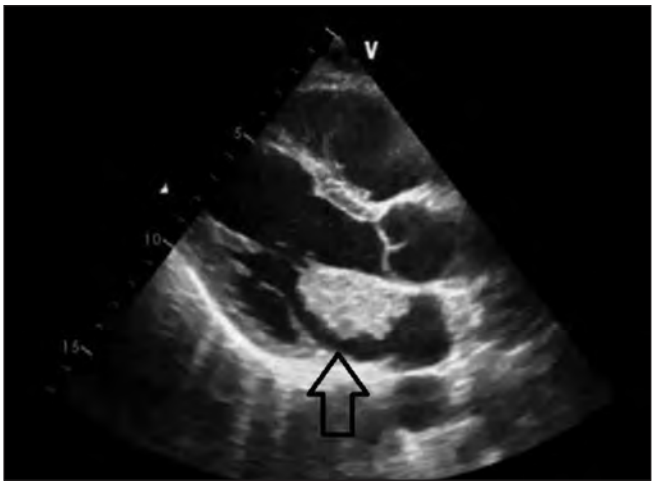

Figure 1. Three-chamber parasternal long axis view on echocardiogram with arrow indicating left atrial myxoma

Discussion

Myxomas, the most common type of benign primary cardiac tumor, account for approximately one-third of benign tumors of the heart.[1-3] They occur across all age groups, more commonly in women and during the third through sixth decades.[1,4-7] 90% of cases are sporadic, while the remainder are transmitted in an autosomal dominant fashion. This inheritance typically occurs as part of the Carney syndrome complex that includes myxomas, lentigines/pigmented nevi, and endocrine overactivity.[1,8-12] The NAME (nevi, atrial myxoma, myxoid neurofibroma, and ephelides) and LAMB (lentigines, atrial myxoma, and

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blue nevi) syndromes likely represent subsets of the Carney complex.[1,13] While the genetic origin of the Carney complex is unclear, patients frequently have inactivating mutations of the PRKAR1A tumor-suppressor gene.[1,13]

Most myxomas arise in the left atrium from the interatrial septum near the fossa ovalis.[4,14] They are gelatinous structures comprised of myxoma cells in a glycosaminoglycan-rich stroma, often pedunculated on a fibrovascular stalk.[1,14,15] They arise from multipotent mesenchymal cells that persist from embryonic cardiac septation and have been found to produce vascular endothelial growth factor (VEGF), which is implicated in the early stages of tumor growth.[13-16]

Depending on location, myxomas can cause the signs and symptoms of corresponding valvular disease; atrial myxomas can mimic atrioventricular (AV) valve stenosis due to valve obstruction, while ventricular myxomas can cause outflow obstruction similar to subaortic or subpulmonic stenosis.[1,3] Diastolic murmurs result from obstruction of the filling ventricle, while systolic murmurs arise when valve closure or outflow is interrupted.[3] The characteristic "diastolic tumor plop," a low-pitched sound during early to mid-diastole, is thought to represent the physical impact of the tumor hitting the AV valve or ventricular wall.[1] Repetitive trauma to the valve may also cause damage and regurgitation due to a "wrecking ball" phenomenon.[17] Congestive heart failure (CHF) may result from obstruction of cardiac flow and/or impaired contractility from direct invasion of the myocardium (myocardial damage may also cause arrhythmias, heart block, pericardial effusion or tamponade).[2,18,19] Symptoms may provide a clue to the tumor's location: left-sided tumors may present with dyspnea on exertion, orthopnea or flash pulmonary edema; right-sided tumors may present with syncope, sudden death or right heart failure.[2, 18-21] Of note, these symptoms can be sudden and/or positional due to the effect of gravity on the tumor's position.[1,2,18,19] Fragments of myxomas may embolize and cause signs of peripheral or pulmonary emboli, as well as fever, weight loss, malaise, arthralgias, rash, digital clubbing, and Raynaud's phenomenon.[1,2,22-28] Laboratory abnormalities associated with myxomas include anemia, leukocytosis, thrombocytopenia/cytosis, elevated erythrocyte sedimentation rate (ESR) and CRP.[1,2,22] Rarely, myxomas may become infected; the incidence of cerebral and systemic emboli from infected myxomas is greater than that from non-infected myxomas.[29] Management of these patients is often complicated by bacteremia, septic shock, disseminated intravascular coagulation (DIC), multi-organ failure, and cerebral infarction.[30]

Echocardiography is useful in diagnosing myxomas; tumor size, shape, and attachment can be assessed, all of which assist in surgical planning for excision.[1,3,18,31] TEE offers superior imaging compared to transthoracic echocardiography (TTE) owing to the proximity of the esophagus to the heart and use of high frequency transducers.[31] Cardiac CT and MRI offer noninvasive imaging options that can also help evaluate the characteristics of myxomas.[1,2] In the past, cardiac catheterization and angiography were routinely performed prior to excision; their role has diminished in favor of noninvasive imaging due to the additional risk of tumor embolization during catheterization.[1,32]

Definitive treatment for a myxoma is surgical excision; medical therapy is only indicated for management of concomitant CHF and/or arrhythmias.[1,33,34] Septal defects and/or valvular damage that occur during surgery may require repair.[33,34] A potential complication of excision is fragmentation and embolization of part of the tumor.[33,34] While thrombolytic therapy is reasonable for stroke secondary to blood clot emboli from cardiac myxomas, it has little effect on emboli composed of actual tumor.[35] Overall post-surgical prognosis is excellent, with a 3% rate of operative mortality.[1,19,33,34] Recurrence is rare in sporadic cases (1-2%) and usually due to inadequate resection; familial myxomas recur in 12-22% of cases and are likely due to multifocal lesions.[1,2,19,27] Follow-up includes routine semi-annual echocardiography. Screening of first-degree relatives is appropriate due to the possibility of familial inheritance.[1]

Key Points

Although an uncommon cause of stroke, cardiac myxoma should be considered in the differential diagnosis. Definitive treatment is surgical resection, which offers excellent cure rates with rare recurrence and low mortality. Medical management is supportive. Once a cardiac myxoma has been identified, arrangements for surgical resection should be made immediately.

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