Atrial Myxoma Presenting as Acute Stroke: A Case Report and Review of Literature

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Research Article

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ABSTRACT

Cardiac myxoma is a rare cardiac tumor, but is notorious for emboli to the general circulation. Acute ischemic strokes are one of the most common complications of the emboli arising from the cardiac myxoma. Here, we describe an interesting case of acute stroke, which was later found to have myxoma.

Keywords: Atrial Myxoma, Emboli, Acute stroke.

INTRODUCTION

Cardiac myxoma is a source of emboli to the central nervous system and elsewhere in the vascular tree. However, nonspecific systemic symptoms and minor embolic phenomena may be over-looked in the absence of any history of cardiac problems. In this situation, cardiac investigations may not be performed and diagnosis of this rare condition may be delayed until the onset of more significant embolic disease, such as stroke with functional impairment, as in the case reported here. The clinical presentation of cardiac myxoma is discussed, along with appropriate investigations and treatment, which may prevent such sequelae.

Case Report

61 y/o Hispanic female with past medical history significant for Migraines headaches was brought by the emergency medical services (EMS) for altered mental status (AMS) and uncontrolled vomiting. The history was obtained by her husband. The patient was in her usual state of health and was driving with her husband when she suddenly started rubbing her face with her left hand while driving with the other hand and developed aphasia. She was making incomprehensible sounds but was able to follow her husband and pulled over the car on his command. The husband called EMS which arrived in 15 minutes. Prior to the episode she did not complain of any headache, blurry vision, palpitations, chest pain or seizure like activity. Her family history was significant only for Epilepsy.

In the ER patient was found obtunded with GCS (Glasgow coma scale) of 9 (E2V1M6). She was afebrile and hemodynamically stable with BP 131/73, HR of 65, RR 26 and saturating at 96%.

Examination of CNS revealed bilaterally equal (3mm) pupils reactive to light. Tonic flexion of Right elbow, moving all extremities to painful stimuli, brisk reflexes and Babinski reflexes bilaterally. Examination of other systems was normal. CBC and Basic metabolic profile were sent and a 12 lead EKG was performed which were normal. CT Scan of head was also done which showed no acute changes. Patient was admitted to medical intensive care unit (MICU) with presumptive diagnoses of Acute Brain Stem Stroke, Seizures or Migraine.

Frequent neurological evaluations were performed. Over next few hours, Patient’s GCS became seven and she got intubated for air way protection, her Right pupil was dilated, sluggishly reactive to light and she was not moving her Right extremities to pain. An MRI performed later revealed large left sided MCA infarct with midline shift but no hemorrhage. Patient was started on osmolar therapy and Neuro Surgery Consult was called for emergent hemi craniectomy and evaluation of hematoma.

Other workup was done to evaluate the causes of stroke which revealed normal MRA of head and Neck, no significant stenosis on carotid Dopplers. ESR and CRP remained negative. Echocardiogram was done to rule out cause of cardio-embolic stroke which showed a large well circumscribed mass on the intra-atrial septum consistent with a left atrial myxoma. Following the diagnosis of myxoma, patient was started on anticoagulation. Over the next
couple of days, the osmolar therapy was tapered down with slight improvement in neurological status and patient was weaned off the ventilator.

DISCUSSION

Atrial myxoma, the most common benign cardiac tumor, is found more commonly in young adults with stroke or transient ischemic attack (1 in 250) than in older patients with these problems (1 in 750) Hart et al, (1998). The annual incidence is 0.5 per million population [Pinede et al, (2001)], with 75% of cases occurring in the left atrium. There is a 2:1 female preponderance, MacGowan et al, (1993) and the age at onset is usually between 30 and 60 years.

The presentation of atrial myxoma often comprises a diagnostic triad. Active illness is often accompanied by elevation of ESR and C-reactive protein, hyperglobulinemia and anemia. Constitutional symptoms may be mediated by interleukin-6, produced by the myxoma itself [McCarthy et al, (1986)].

Strokes are often recurrent Kirschner et al, (2000) and may be embolic or hemorrhagic, the presentation ranging from progressive multi-infarct dementia Markel et al, (1987) to massive embolic stroke causing death Mercier et al, (1980). Because tumor fragments or adherent thrombus may embolize, anticoagulation may not be protective [Blondeau P. (1990)]. The multiple, bilateral fusiform aneurysms commonly found on peripheral arterial branches predispose the patient to cerebral hemorrhage Mendoza et al, (2001).

Transesophageal echocardiography, which has been reported as having 100% sensitivity for cardiac myxoma [Kessab et al, (1999)] is preferred over transthoracic echocardiography. Transesophageal echocardiography may also improve the detection of other major cardioembolic sources (e.g., intracardiac thrombus, vegetations or aortic arch plaque), as well as less common potential sources (e.g., patent foramen ovale, atrial septal aneurysm or left ventricular aneurysm) Hutton JT. (1981).

Cardiac MRI can assist in delineating tumor size, attachment and mobility [Bienfait et al, (2001)]. This information may be helpful in surgical resection, which, because of the risk of further embolization, should not be deferred even in asymptomatic cases discovered incidentally. Resection may lead to normalization of serum interleukin-6 levels and resolution of constitutional symptoms, and the intracranial aneurysms may regress and resolve [Knepper et al, (1988)].

Neurological sequelae after resection are rare but may occur without recurrence of the cardiac tumor. Instead of regressing, aneurysms may enlarge or appear for the first time [Price et al, (1970)]. Tumor fragments that have metastasized to the vessel walls may enlarge, causing vessel occlusion and delayed infarction, or they may penetrate through the vessel wall, forming intra-axial metastases. Adams et al, (2000).
Primary tumors recur in only 1% to 3% of sporadic cases, often because of inadequate resection. For patients with sporadic myxoma, annual review with echocardiography is suggested for a period of 3 to 4 years, when the risk of recurrence is greatest [Engberding et al, (1993)]. The recurrence rate is 1~3% after surgery. O'Rourke et al, (2003) For Carney complex, which has a recurrence rate of up to 25%, lifetime annual review with familial screening is recommended. Kapral et al, (1999)

CONCLUSIONS

The diagnosis of atrial myxoma can be elusive, especially when the symptoms suggest a systemic illness. In the case reported here, the significance of these symptoms became apparent when the patient presented acutely with a motor deficit as a result of cerebrovascular embolism. The presence of bihemispheric infarction focused subsequent investigations on the possibility of a proximal source of embolization, which resulted in identification of the causative atrial myxoma.

REFERENCE