An Unusual Cause of Stroke in a Young Adult

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Cerebral vascular accidents of uncertain origin in any patient pose a burden on the family physician to uncover the underlying cause by means of a thorough and concise workup. Even though cardiac tumors are rare, missing the diagnosis may lead to devastating results. The case reported here is of a 40-year-old man with a left atrial myxoma as the cause of his embolic cerebral infarct.

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Cerebral vascular accidents are uncommon in young or middle-aged adults, and when they occur, the more common risk factors for stroke include hypertension, diabetes mellitus, hyperlipidemia, acquired immunodeficiency syndroms (AIDS), heavy alcohol consumption, and a family history of stroke.[1] Less commonly known risk factors and causes of stroke in the young or middle-aged adult include inactivity (3.1 strokes per 1000 men per year compared with 0.5 strokes per 1000 men per year for men actively engaged in vigorous exercise)[2]; cigarette smoking in men (relative risks of nonfatal and fatal stroke were 2.52 and 1.24, respectively, for men smoking 20 or more cigarettes per day)[3]; cocaine use[4]; atrial fibrillation with other clinical risk factors (ie, recent congestive heart failure, history of hypertension, or previous atrial thromboembolism)[5]; atrial myxomas[6]; and being black (a substantially higher risk of intracerebral and subarachnoid hemorrhage than in whites).[7]

Case Report

A 40-year-old white man developed a sudden change in mental status; he was unable to communicate or follow commands, and was in a state of confusion. Paramedics were summoned to his home and found the patient to have no evidence of trauma, but he had global aphasia with right-sided weakness of the upper extremity more than the lower extremity; his blood pressure was 210/140 mm Hg. The patient had a past medical history for hypertension and hyperlipidemia and was regularly taking antihypertensive medication, ie, lisinopril 20 mg daily. He had no history of diabetes mellitus, heart palpitations, alcohol or drug use, prior thromboembolic episodes, cigarette smoking, or family history of stroke. He was treated for hypertension en route to the emergency department.

In the emergency department, the patient was awake but disoriented. He was aphasic, unable to follow simple commands, and made inappropriate responses to questions posed by emergency department personnel. The patient's blood pressure was 120/56 mm Hg, his pulse 95 beats per minute, oral temperature 96.6°F (35.9°C), and respirations 16 per minute. The right side of the patient's face drooped and his tympanic membranes were clear, with no hemotympanum. There was no thyromegaly. No carotid bruits were auscultated. An examination of the heart and lungs revealed no abnormalities. The abdomen was soft and nontender, and there was no hepatosplenomegaly.

A neurologic examination revealed that the pupils were sluggish but reactive to light. Extraocular muscles were intact; accommodation could not be tested. Fundi showed no papilledema, and visual fields were hard to assess. Conjugate gaze was with left preference. There was minimal flattening of the right nasal labial fold. Other cranial nerves could not be assessed. Deep tendon reflexes of the brachial radialis, triceps, and patella were 3+ on the right side and 2+ on the left. The Achilles tendon reflex was 2+ bilaterally with a questionable Babinski sign on the right side. Decreased motor function and muscle tone were greater on the right upper extremity than on the right lower extremity, with strong sensory pain withdrawal on the left side compared with the right.

Laboratory evaluations revealed the following

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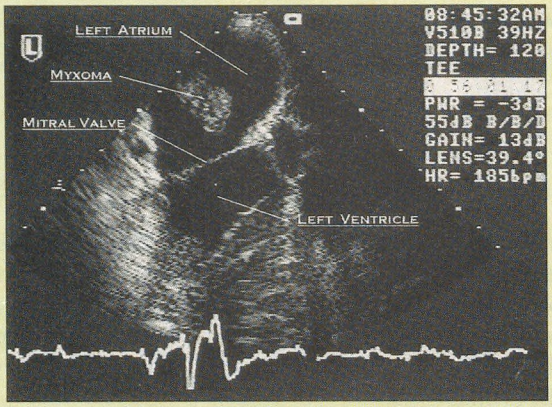
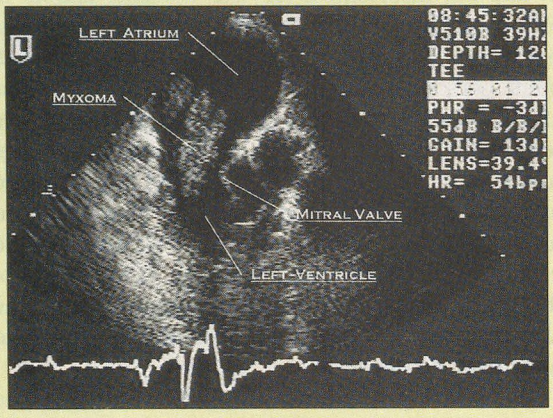
FIGURE 1

Transesophageal echocardiogram shows (A) tumor sitting in left atrium with mitral valve closed, (B) prolapsing into left ventricle, and (C) retracting back to left atrium.



A



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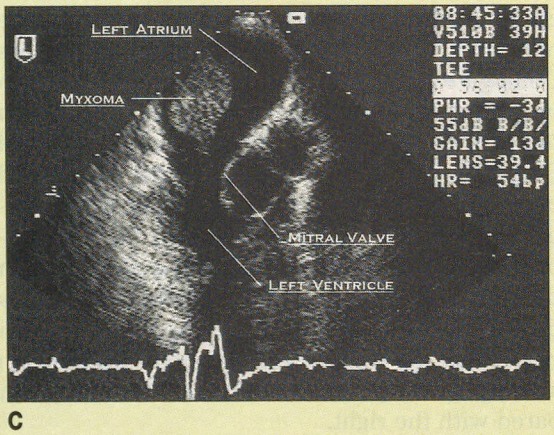


C

values: white blood cell count (WBC) 8.6 x 10^3/mm^3, hemoglobin 14.4 g/dL, hematocrit 42.0%, platelet count 220 x 10^3/mm^3, neutrophils 84%, lymphocytes 8%, monocytes 7%, and eosinophils 1%. Sodium level was 135 mEq/L, potassium 3.7 mEq/L, chloride 100 mEq/L, CO_2 23 mEq/L, BUN 15 mg/dL, creatinine 1.0 mg/dL, glucose 120 mg/dL, and sedimentation rate 6 mm/h. Total cholesterol was 207 mg/dL and triglycerides 168 mg/dL. The drug and alcohol screening tests were negative. An ANA test was within normal limits. Arterial blood gases on room air showed a pH of 7.4, P_{CO_2} at 38 mm Hg, P_{O_2} at 96.8 mm Hg, and bicarbonate 25 mEq/L.

A lumbar puncture was within normal limits. A computed tomographic (CT) scan of the brain showed no evidence of hemorrhage. Doppler ultrasongraphy of the carotid artery was negative. Transthoracic echocardiography showed no mass, thrombi, or valvular abnormality. An electrocardiogram (ECG) showed normal sinus rhythm with no ectopy or ST changes. A magnetic resonance imaging scan (MRI) of the brain 2 days later showed basal ganglia and temporal and parietal ischemic changes on the left side compatible with a left middle cerebral artery infarct.

Although the standard workup of the patient revealed no abnormalities, including a transthoracic echocardiogram that failed to show any cardiac thrombus or tumor, other conceivable causes had to be investigated before the patient's condition was diagnosed as an idiopathic stroke and he was sent for rehabilitation.[6] In this case, the patient had a transesophageal echocardiogram that revealed a 3.0-cm left atrial myxoma, the cause for his embolic stroke. This was done as a result of wanting to leave "no stone unturned." If the patient had been sent off to rehabilitation without this diagnosis, stroke recurrence would be extremely high with devastating results in the young adult, such as permanent brain necrosis from atrial myxoma showers.[6] The patient was transferred to a tertiary care medical center, where open heart surgery and removal of the benign atrial myxoma was completed without incident.

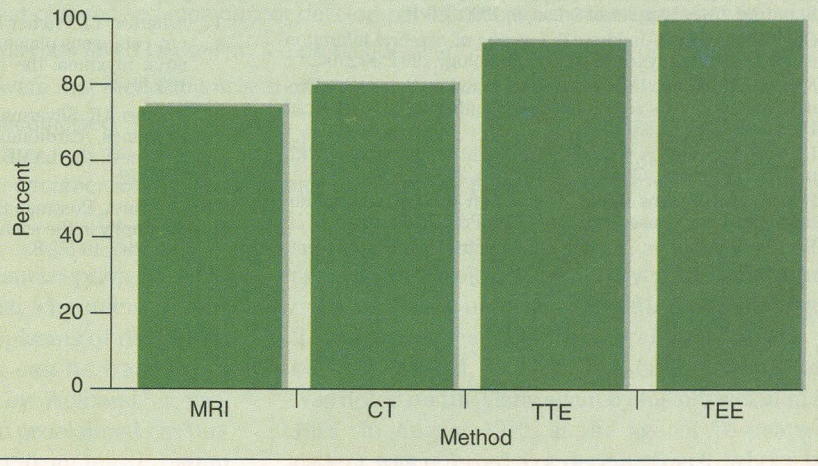
At present, the patient is not taking any anticoagulation medication, he is in speech and occupational rehabilitation, and doing quite well. He is currently taking lisinopril 10 mg daily, digoxin 0.25 mg daily, metoprolol 50 mg 1/2 twice daily, and hydrochlorothiazide 25 mg/triamterene 37.5 mg daily.

Atrial Myxoma

All primary tumors of the heart are rare (0.001% to 0.28% of autopsy cases).[8] Atrial myxomas are the most common, with 80% of them found in the left atrium. Other primary cardiac tumors include rhabdomyomas, fibrous histiocytomas, hemangiomas, and different types of sarcomas. The more common metastatic tumors to the heart include bronchogenic carcinoma, breast carcinoma, malignant melanoma, lymphomas, renal cell carcinoma, and Kaposi's sarcoma.[8]

FIGURE 2

Rate of detection of atrial myxomas, by type of radiology. MRI denotes magnetic resonance imaging; CT, computed tomography; TTE, transthoracic echocardiology; and TEE, transesophageal echocardiology.



Atrial myxomas are usually pathologically benign with the potential to embolize systemically, and are diagnosed more frequently in middle-aged women than men. Diagnosis is usually accomplished with the aid of echocardiography, MRI,[8,10] or cutaneous biopsy of emboli material.[11] Cardiac myxomas have surgical cures, low recurrence, and good long-term survival.[12] Patients may present with fever, weight loss, and malaise, with an increased erythrocyte sedimentation rate and leukocytosis, or with cardiac obstruction of blood flow or signs of peripheral, cerebral, or pulmonary embolization (depending on the tumor location). With cardiac obstruction, the patient may present with right-sided heart failure (right-sided tumors) or transient pulmonary edema exacerbated in the upright position (gravity pulling the tumor into the mitral valve) with left atrial tumors (Figure 1).

On physical examination, a diastolic "tumor plop" sound or a mitral stenosis-like murmur may be auscultated. Numerous cutaneous manifestations may also be present. For example: livedoid macules,[11] an erythematous papular eruption of the extremity associated with claudication,[13] nonblanching lesions of the fingertips,[14] necrosis to the toes,[15] Raynaud's phenomenon,[16] and erythema and petechiae of the hands and feet.[13] Also, complex syndromes where myxomas and cutaneous lesions are included are: NAME syndrome (nevus, atrial myxoma, and neurofibroma ephelides[17]), and LAMB syndrome (lentigines, atrial myxomas, and blue nevi[18]). Tachyarrhythmias with or without ST changes on ECG may also be subtle clues.

Atrial myxomas are difficult to diagnose given their rarity, and between 5% amd 9% are missed on transthoracic echocardiography.[8,19] However, transesophageal echocardiography (TEE) has a 100% diagnosis rate of atrial myxomas,[8] followed by 78.4% by CT and 70% by MRI[8] (Figure 2). There must be a high index of suspicion on the part of the physician and a persistent pursuit to uncover the cause of a stroke in a concise and well-thought-out workup.

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