Fulminant Brain Necrosis From Atrial Myxoma Showers

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Background: Atrial myxoma is uncommon and may be associated with brief isolated ischemic events. We describe a patient with atrial myxoma and an incompletely evaluated ischemic stroke followed 1 year later with almost complete brain necrosis.

Summary of Report: A 63-year-old woman presented with a rapidly progressive illness resulting in coma within 24 hours from multiple myxomatous emboli. Cranial computed tomographic scan demonstrated multiple hypodensities in both cerebellar and cerebral hemispheres. Postmortem examination revealed virtually complete brain necrosis, multiple peripheral artery embolizations, embolization in the coronary arteries with acute myocardial infarction, and tumor emboli in the spleen and kidneys.

Conclusions: Although uncommon, atrial myxoma may present as a fatal multiple organ embolization that includes both anterior and posterior circulation territories of the brain. (Stroke 1993;24:1090-1092)

Key words • cerebral infarction • myxoma • tomography, x-ray computed

Atrial myxoma may represent a major source of neurological morbidity. Clinically apparent cerebral emboli have been reported in the range of 25%.1-3 In most patients with atrial myxoma, embolization of tumor fragments results clinically in isolated nonfatal ischemic strokes, but in others sudden plugging of the mitral valve opening may cause syncope or sudden death.4-5 Although atrial myxomas are rare, failure to recognize them may deprive patients of potential life-saving tumor resection. We describe a patient with a left atrial myxoma and an incompletely evaluated ischemic stroke that unfortunately led to fatal multiple organ emboli 1 year later.

Case Report

A 63-year-old woman was found unresponsive at home. She had been in good health until 1985, when she was briefly hospitalized for atypical chest pain. No serial electrocardiographic changes were found. Transthoracic echocardiography revealed a mild prolapse of the anterior leaflet. (No intracardiac masses were seen, even after review following this tragic event.)

One year before her present admission she had a small ischemic stroke in the inferior division of the left middle cerebral arteries. Physical examination showed a right hemiparesis and right facial droop with no speech difficulties. Cardiac auscultation was normal. Duplex scan revealed nonstenotic plaques in both carotid arteries. Echocardiography was not repeated. She was left with a mild residual right arm weakness.

On arrival to the emergency room, physical examination revealed shallow regular breathing with a respiratory rate of 26, blood pressure of 120/60 mm Hg, and temperature of 37.7°C. She was unresponsive to verbal stimuli and had no spontaneous motor movements. Eyes were deviated to the right. With painful stimuli she grimaced and withdrew her left arm and both legs. Pupils were 3 mm in size and minimally reactive to light. No papilledema was found. Corneal reflexes were absent. Muscle-stretch reflexes were increased in both the upper and lower extremities with bilateral extensor responses.

Laboratory examination revealed a respiratory, compensated metabolic acidosis with a lactate level of 6.8 mmol/L. Creatinine phosphokinase was 2300 U/L. Levels of urea nitrogen, creatinine, calcium, electrolytes, hematocrit, and white cell count were normal. The electrocardiogram showed a normal sinus rhythm with no acute ST segment changes. A radiograph of the chest showed scattered alveolar infiltrates in the left lower and right middle lobe of the lung consistent with aspiration pneumonitis.

Cranial computed tomographic scan revealed low attenuation areas within both cerebral hemispheres punctuated with multiple hemorrhages scattered throughout both cerebral hemispheres and effacement of cerebral sulci, consistent with massive cerebral infarction (Fig 1). The patient rapidly progressed to unresponsive coma. A cold, pulseless left arm and lower extremity preceded death.

Postmortem examination revealed a large left atrial myxoma with evidence of tumor emboli to multiple organs. The brain weighed 1420 g and showed generalized edema with bilateral uncal herniation and brain stem Duret's hemorrhages. Old and recent cerebral infarcts were found in the left superior and middle frontal gyrus extending to the precentral gyrus, the left inferior parietal lobule, left occipital pole, right superior inferior parietal

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lobule, right precentral gyrus, left dorsal cerebellum, bilateral basal ganglia, thalamus, lower midbrain, and pons. Multiple tumor emboli were found in the intracranial portion of the internal carotid (Fig 2) and vertebral arteries and in small leptomeningeal arteries. Additional tumor emboli were found in the coronary arteries, with multiple areas of recent and old myocardial infarction, and in the kidneys and spleen.

**Discussion**

Cardiac myxoma is an infrequent etiology of ischemic stroke. Studies suggest that atrial myxoma occurs in 0.5% of acute stroke patients, with women in the fifth decade at greatest risk. Transthoracic echocardiograms have been repeatedly negative in patients with atrial myxoma, and pathological proof of complete tumor embolization is on record. Transesophageal echocardiogram or, more likely, magnetic resonance imaging of the heart may increase the detection of atrial myxoma, but further study is warranted.

Atrial myxoma may present with many clinical syndromes. Patients are seen primarily by cardiologists with palpitations, exertional dyspnea, or syncope. In addition, the exceptional friable and spongy mass of atrial myxoma may easily dislodge fragments into the systemic circulation that may conceivably explain some of the nonspecific systemic complaints such as fever, fatigue, or weight loss. The central nervous system is one of the most susceptible areas of embolization, resulting in multiple ischemic strokes, and at times independent metastatic growth in the brain occurs.

Several unique clinical and pathological features prompted us to report this case. First, failure to recognize and treat atrial myxoma a year before this fatal illness resulted in many cerebral and myocardial infarcts that apparently were silent. Indeed, recent magnetic resonance imaging studies in patients with atrial myxoma have demonstrated the presence of multiple small cerebral infarcts that mimic lacunar disease. In our patient, repeat echocardiography at the time of her first stroke would have been appropriate rather than attributing the ischemic event to a previously demonstrated mitral valve prolapse.

Second, our patient arrived at the medical intensive care unit in a moribund condition and demonstrated an unrelenting process of embolization. An overwhelming shower of myxomatous material resulted in occlusion of both carotid arteries and branches of both anterior and posterior cerebral circulation. More frequently, patients with atrial myxoma are seen with small and single

**Fig 1.** Computed tomographic scan with a massive hypodensity in both cerebral hemispheres and multiple hypodensities in the cerebellar hemispheres. White-gray matter discrimination and cortical sulci have disappeared.
territory cerebral infarcts. Bilateral large-vessel occlusion has not yet been reported, to our knowledge.

Surgical resection is indicated when cardiac myxoma is detected. Cardiac surgery carries a comparatively low morbidity of approximately 5%, and later recurrences are virtually eliminated.10

References

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