Multiple Strokes Due to Atrial Myxoma With a Negative Echocardiogram

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We report a patient with three cerebrovascular events involving the posterior circulation, one of which occurred in conjunction with central retinal artery occlusion. The patient had constitutional findings but had a normal cardiac examination, negative echocardiogram, and negative blood cultures. Angiography on two separate occasions revealed left vertebral artery occlusion. Repeat echocardiography 10 months later, at the time of the third stroke, revealed a left atrial mass consistent with myxoma. In patients with repetitive unexplained embolic events in different vascular distributions and constitutional findings suggestive of atrial myxoma, repeat noninvasive studies are indicated even if the initial workup is negative. (Stroke 1988; 19:1570–1571)

Primary cardiac tumors are found in 0.05% of routine autopsies and are a rare cause of cerebral emboli. Approximately 50% of these tumors are myxomas, and about 75% occur in the left atrium. The diagnosis may be suggested by the typical triad of constitutional, embolic, or obstructive symptoms, but cardiac imaging studies are usually required since clinical evaluation rarely discloses specific findings.

We report the case of a 60-year-old woman with an atrial myxoma and recurrent cerebral embolic events. She had anemia, an increased erythrocyte sedimentation rate (ESR), and hypergammaglobulinemia but no cardiovascular symptoms or signs. Her initial echocardiogram was reported to be normal. Ten months later, after her third ischemic event, echocardiography demonstrated a left atrial mass; review of the initial echocardiogram failed to reveal any abnormalities that had been overlooked. This case suggests that atrial tumors cannot always be ruled out as a cause of embolic events even with an initial negative echocardiogram.

Case Report

While dancing, a 60-year-old right-handed woman had the acute onset of dizziness, nausea, and vomiting followed by transient paresthesias of her left face and hand. Initial examination demonstrated normal heart sounds and no cardiac murmurs. She had a left Horner's syndrome. An incomplete conjugate upgaze paresis was noted. Left beating nystagmus was seen in the primary position with bilateral gaze-evoked nystagmus. There was mild weakness of her left face, arm, and leg. Sensory testing revealed a decreased sensation to pinprick on the left side of her face. Finger-to-nose and heel-to-shin testing demonstrated dysmetria on the left. The patient was symmetrically hyperreflexic and had a left Babinski's response.

Laboratory studies included a white blood cell count of 14.0 x 10^3, a hemoglobin concentration of 10.0 g/dl, and a hematocrit of 31.1%. Electrocardiography demonstrated normal sinus rhythm with first-degree atrioventricular block. Two-dimensional echocardiography revealed normal aortic, mitral, and tricuspid valves, normal chamber size, and normal wall motion; no intracardiac masses were visualized. Increased signal intensity of the left cerebellar hemisphere and possibly the left posterolateral medulla was noted on T2-weighted magnetic resonance imaging (MRI). Angiography revealed complete occlusion of the distal left vertebral artery. Additional laboratory tests obtained following discharge included a hematocrit of 35%, an ESR of 54 mm/hr, and a polyclonal increase in γ-globulins.

Nine months later the patient developed the sudden onset of an inability to speak, drooping of the right side of her mouth, and leaning to her right. Examination revealed normal heart sounds without any murmurs. The patient was lethargic and disoriented with dysarthric speech. Sensation was decreased on the right side of her face and her left body. Right-sided weakness was present in her face and arm. Dysmetric movements were noted on the left. Reflexes were increased on the left, with bilat-
eral Babinski’s responses. Angiography was repeated and again demonstrated a left vertebral artery occlusion. MRI revealed the previous lesion in the left cerebellum and left medulla and new abnormalities involving the right cerebellum, right pons, left thalamus, and right occipital lobe.

Approximately 1 month after discharge, the patient had the acute onset of nausea and vomiting. She was found leaning forward and became unresponsive. On examination, there was evidence of a left central retinal artery occlusion. Her head and eyes were deviated to the right, and she had spontaneous movement on the left but only minimal withdrawal to noxious stimulation on the right. Repeat two-dimensional echocardiography revealed normal mitral, tricuspid, and aortic valves; cardiac chamber size and wall motion were normal. A mobile left atrial mass was seen attached near the fossa ovalis region of the atrial septum; in diastole the mass extended through the mitral valve.

On the day after admission the patient developed a temperature of 104°F. She was placed on antibiotics for an aspiration pneumonia and an E. coli urinary tract infection. On the fourth hospital day she suffered a cardiac arrest and died. No autopsy was performed.

Discussion

We report a patient with recurrent embolic events involving the posterior circulation, one in conjunction with central retinal artery occlusion. Ten months after the initial event, and after her third embolic episode, echocardiography revealed a left atrial mass suggestive of myxoma.

Experienced laboratories report a 100% sensitivity in diagnosing atrial myxomas studied by two-dimensional echocardiography.\footnote{1} False-negative results have been reported when only M-mode echocardiograms were performed\footnote{2} or if the tumor was sessile.\footnote{3} To our knowledge, no false-negative results have been reported with two-dimensional echocardiography. The failure of an initial two-dimensional echocardiogram to detect the atrial mass in our patient may have been related to its size. Ten months after the original event and after a significant increase in size, the mass was detected.

Most cardiac myxomas occur in the left atrium, with 75% of these attaching to the atrial septum near the fossa ovalis. Myxomas may be sessile or pedunculated.\footnote{4} The friable, pedunculated tumors are more likely to embolize because of their mobile state.\footnote{5} Emboli may consist of either myxomatous tissue or thrombus on the surface of the mass.\footnote{6} Embolic myxoma has been described as invading, displacing, and destroying normal elements of arterial walls.\footnote{7} This process was accompanied by connective tissue proliferation and a mild inflammatory reaction, potentially resulting in aneurysm or pseudoaneurysmal formation. Rupture of an aneurysm could thus be responsible for hemorrhagic events in the absence of emboli.\footnote{8} In our patient the vertebral artery occlusion may have been composed at least partly of myxomatous embolic material with superimposed thrombus formation leading to additional embolic events in the same vascular territory.

This case illustrates that in patients with recurrent embolic events and constitutional signs, with or without cardiac symptoms, cardiac myxoma must strongly be considered. Repeat echocardiography or further cardiac imaging studies (i.e., computed tomography, MRI, or radionuclide studies) should be performed if an initial echocardiogram is not revealing to rule out this potentially curable cardiac tumor when clinical suspicion is high.

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References


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