Embolism to the central nervous system from cardiac myxoma

Report of two cases

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Two cases of left atrial myxoma are reviewed, both presenting as embolic phenomena. Neither patient gave a history compatible with pre-existent cardiac dysfunction. Sudden collapse and subsequent right hemiplegia resulted in one patient when an embolus lodged in the left middle cerebral artery. The second patient presented with headache and transient visual obscuration in the left eye. She showed evidence of embolism to the central retinal artery, and particulate matter could be seen within the retinal arterioles. Attention is drawn to the fact that echocardiography now constitutes a simple, noninvasive, and highly reliable method of making this diagnosis. The propensity for embolic tumor fragments to grow and invade cerebral arterial walls is discussed along with its possible neurosurgical significance.

Key Words • cerebral embolus • cardiac myxoma • echocardiography • pseudoaneurysm

Since 1962, seven cases of cardiac myxoma have been diagnosed antemortem and surgically excised with pathologically confirmed diagnoses at our institution. Two of the seven experienced central nervous system dysfunction as the initial manifestation of their disease. One patient sustained catastrophic neurological damage when an embolus occluded his left middle cerebral artery. The second patient demonstrated embolism to the central retinal artery, with transient monocular visual obscuration. A third patient, not reported here, suffered a right middle cerebral embolus intraoperatively during cardiac surgery.

Primary cardiac tumors are rare, being found in only 0.05% of routine autopsies. Metastatic neoplasms to the heart are more common, occurring in 0.6% of autopsied material. Myxoma is the most common primary tumor of the heart. About 75% of these occur in the left atrium, usually near the fossa ovale. Approximately 20% originate in the right atrium, while the remaining few arise from the ventricle, a valve leaflet, or from the wall of the pulmonary artery. Cardiac myxomas have occurred in the newborn as well as the elderly, but the greatest age incidence is between 30 and 60 years. Silverman, et al., in a review of the literature through 1962, presented a total of 33 cases of cardiac myxoma demonstrating embolization. Of that number, 30 tumors arose in the left atrium. Fifteen of the 33 cases (45.5%), at
some point in their clinical course, were associated with embolism to the brain. Only two cases (6%) reviewed were found to have had embolism to the eye. For unknown reasons, women are affected with cardiac myxoma three times as frequently as men. Sudden death can occur in as many as one-third of the cases of atrial myxoma.

Case Reports

Case 1

On June 9, 1975, this 17-year-old boy suddenly collapsed. His eyes would open, but he responded to his father only by moving his head. There was no history of seizure. He was seen by his physician and referred to our hospital. His past history was unremarkable.

Examination. On admission, his blood pressure was 160/118 and heart rate was 68. Respirations were not labored. He was moderately obtunded. When aroused he would attempt to follow simple commands. Neurological examination revealed a profound right hemiparesis and right central seventh nerve palsy. He had global aphasia. Pupils were equal and reactive with no gaze disturbances. Plantar responses were normal, and no sensory deficit was discernable. His neck was supple. General physical examination was negative. There was no cardiac murmur. An echoencephalogram was midline. Admission hemogram revealed 10,500 white blood cells, and a hemoglobin of 15 gm%. Electrolytes, blood urea nitrogen, and blood glucose were normal. Special hemorrhagic studies were normal. Erythrocyte sedimentation rate was moderately elevated at 38 mm/hour (normal = less than 15) and C-reactive protein measured 3+. Rheumatoid factor and antinuclear studies were negative. Antistreptolysin O titer was normal, as was an electrocardiogram. Serum protein electrophoresis showed mild elevation in alpha 1 globulin at 0.36 gm/100 ml (normal levels = less than 0.28 gm/100 ml) and a borderline elevated alpha 2 globulin. Gamma fraction was normal. Blood cultures revealed no growth. Chest and skull films on admission were normal. Emergency left carotid arteriogram demonstrated abrupt occlusion of the left middle cerebral artery at its trifurcation, with minimal collateral flow.

Operations. The patient underwent craniotomy, and a superficial temporal artery to middle cerebral artery bypass was performed within 8 hours of admission. Cortical softening in the left hemisphere, proven by biopsy, was found. His subsequent evaluation was directed at discovering a source for the embolus. Phonocardiography and carotid pulse tracings were unremarkable. Echocardiography suggested an echo mass in the left atrium compatible with cardiac myxoma. Angiocardiography verified the presence of a left atrial tumor mass which approached and withdrew from the mitral valve with each heartbeat.

On the 16th hospital day, he underwent median sternotomy with successful excision of a large pedunculated left atrial myxoma arising from the atrial septum.

Postoperative Course. His postoperative course was uncomplicated. During the ensuing days, there was partial resolution of motor deficit, so that he could walk without assistance. Dysphasic speech persisted. He was discharged on the 32nd hospital day to the care of his parents and a speech therapist.

Case 2

A 56-year-old woman with a long history of glaucoma presented to her ophthalmologist on December 13, 1968. She complained of three episodes of severe bifrontal headache, worse on the left, one bout of associated diziness, and dimming of vision in her left eye. She gave no history suggestive of previous cardiac disease. Examination of the left fundus revealed several retinal arterioles occluded by what appeared to be embolic particulate matter. She was referred to our hospital for neurosurgical evaluation.

Examination. Complete neurological examination was normal. There were no cervical bruits. Auscultation of the heart revealed a soft systolic murmur, heard best at the apex.

Admission laboratory data demonstrated mild elevation of liver enzymes, an elevated serum lipid level, and an abnormal glucose tolerance curve with a fasting blood sugar of 122 mg%. Complete blood count was normal. Admission chest film showed moderate cardiomegaly with congestion of the central pulmonary vasculature. Calcification was noted in the region of the left atrium. Electrocardiogram was abnormal with frequent atrial premature contractions and nonspecific ST-T abnormalities. Phonocardiography...
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demonstrated a mitral regurgitation murmur and a loud third heart sound. Vectocardiogram was within normal limits. Right retrograde brachial and left direct carotid arteriograms showed no evidence of cervical or intracranial vascular disease.

Cardiac cinefluoroscopy suggested a left atrial enlargement. A rounded, partially calcified mass was also visualized residing within the left atrium which, on occasion, was seen to prolapse into the left ventricle. A diagnosis of cardiac myxoma was made.

Operation. On December 23, 1968, at surgery, a pinkish-tan pedunculated tumor was removed from the left atrium. Histological study documented a cardiac myxoma, partially calcified, with some organizing thrombosis. Convalescence was uncomplicated, and the patient was discharged on January 9, 1969.

Discussion

Several authors have summarized the clinical features of left atrial tumors. Unfortunately, there is no single syndrome characteristic of this disorder. A patient may present with sudden intractable cardiac decompensation associated with a murmur of mitral stenosis. The denial of previous rheumatic heart disease may be helpful in diagnosis. Absence of calcification in the mitral valve and a normal-sized left atrium on chest film favors myxoma over mitral valve disease. Bed rest and digitalis generally will not help such a patient. The cardiac murmur may change in character with alterations in the patient's position. Prolapse of a pedunculated tumor with obstruction of a valvular orifice can produce sudden alterations in consciousness associated with a marked reduction in cardiac output. Yufe, et al., describe such a case which was misdiagnosed as a seizure. It should be emphasized that neither of our patients presented with primary cardiac symptoms.

Constitutional effects of myxomas have been previously described and are common. Such signs include elevated sedimentation rate and positive C-reactive protein as our first case exemplifies. Fever, anemia, and abnormal serum proteins have been noted. Such a clinical presentation may mimic a collagen vascular disease.

Peripheral embolism is a frequent manifestation of atrial myxoma, occurring in about 45% of cases. The brain is affected in about 50%, but multiple organ involvement can be seen. Maroon and Campbell have emphasized that a surgically excised embolus from any location should always be examined histologically, since myxomas have been diagnosed from tumor emboli removed from a variety of sites. Stoane, et al., have summarized the cerebral angiographic appearance of myxomatous emboli and alluded to the presence of two basic patterns: 1) filling defects of varying size with or without interruption of flow, and 2) local changes in the arterial walls and perivascular tissue ranging from slight dilatation and irregularity to pseudoaneurysm formation.

In a clinicopathological study, Price, et al., found that at many sites of myxomatous embolism to the brain, there was actually local invasion of the arterial wall by the myxoma that led to aneurysmal dilatation. They believed that the massive cerebral hemorrhage causing the death of their patient probably resulted from the rupture of such an aneurysm. How frequently this occurs in patients with myxomatous cerebral emboli is unknown. It suggests the need for long-term neurological follow-up study in patients with surgically "cured" cardiac myxomas. Such a study might lead to a decision for embolectomy using microscopic technique if a tumor fragment is located in an accessible site and diagnosed early enough.

Echocardiography is an accurate noninvasive procedure now considered to be a highly reliable screening test in the diagnosis of cardiac myxoma.

Summary

Two cases with central nervous system embolization from cardiac myxoma have been reviewed. The need for a high index of suspicion is stressed when any patient presents with a cerebral embolus of which the origin cannot be readily discerned. Echocardiography, an atraumatic and highly reliable screening test, should be employed as soon as possible.

References


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