Case Report

Gerstmann's Syndrome: Can Cardiac Myxoma Be the Cause?

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Key words:
Atrial myxoma, Gerstmann's syndrome, stroke.

Cardiac myxomas are primary cardiac tumours. Clinical presentations vary. Central nervous embolism has been a constant association. We describe a case of a 40-year-old female who presented with neurological signs and symptoms of Gerstmann's syndrome secondary to a left atrial myxoma.

Manuscript received:
October 15, 2007;
Accepted:
November 14, 2007.

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Cardiac myxomas are benign tumours of the heart.[1] Most myxomas (85%) are located in the left atrium; the remaining 15% are in the right atrium, in the ventricles or, rarely, arise from the valves.[2] Myxomas may produce symptoms related to obstructive valvular dysfunction, to embolism, and constitutional symptoms. The clinical manifestations of cerebral embolism vary, with infarction and ischaemia being the most frequent. We present the case of a female with neurological manifestations of Gerstmann's syndrome, probably secondary to left atrial myxoma, and review the pertinent literature.

Case presentation

A 40-year-old Caucasian female presented to the neurologist of our hospital complaining of headache, dizziness and a reported loss of consciousness. The preliminary neurological examination revealed a confusion of right and left, inability to identify fingers, dysgraphia, occasional errors in speech and dyscalculia when asked to perform simple mathematic procedures. Reflexes and cranial nerve examination were normal. Sensory, motor and cerebral examinations were normal. Physical examination revealed normal vital signs. There was no history of medication or drug abuse, nor of neurological disorders or coagulopathy. The results of blood cell count and biochemistry profile were within normal limits. The only parameter above limits was erythrocyte sedimentation (86 mm/h). Computed tomography (CT) of the brain at her presentation was interpreted as normal (Figure 1). Cardiac auscultation revealed a diastolic murmur. The electrocardiogram was normal, but transthoracic echocardiography indicated the presence of a left atrial mass measuring 3 x 3 cm, attached to the atrial septum, with a shaggy surface (Figure 2). Transoesophageal echocardiography confirmed the diagnosis of atrial myxoma. A new CT of the brain one week after her admission indicated a lesion on the left parietal lobe (Figure 3). Based on the physical examination and the diagnostic procedures, the diagnosis of Gerstmann's syndrome, probably secondary to left atrial myxoma, was established. The patient underwent excision of the atrial mass by a biatrial approach under total cardiopulmonary bypass, and closure of the atrial septal defect with a pericardium patch. The postoperative course was uneventful. The patient joined a follow-up program. One year af-

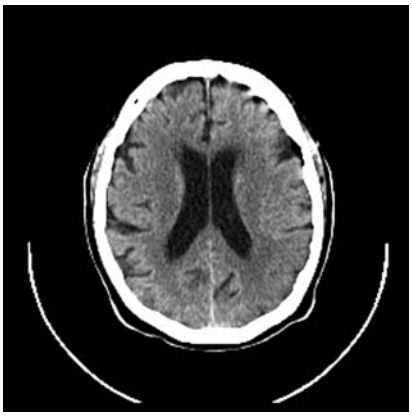

Figure 1. Computed tomography of the brain at the time of admission, which was interpreted as normal.

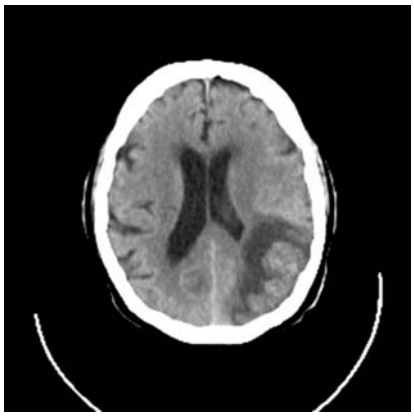

Figure 3. Computed tomography of the brain one week after the patient's admission, indicating a lesion on the left parietal lobe

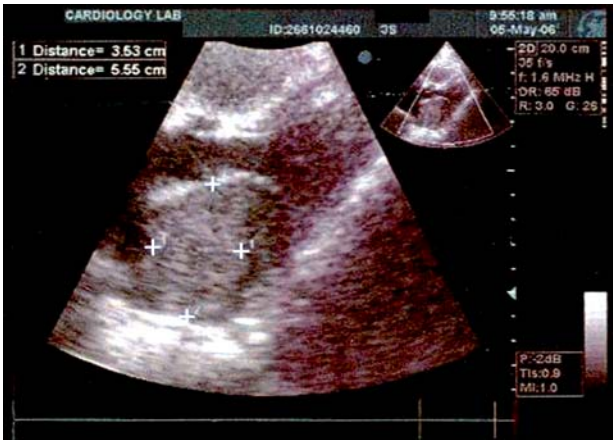

Figure 2. Echocardiography demonstrating the existence of an atrial myxoma.

ter the initial presentation and following speech therapy the patient is doing well, with the symptoms being diminished and no sign of recurrence of myxoma.

Discussion

Cardiac myxomas are primary cardiac tumours and make up approximately 50% of them in patients of all ages. They are intracavitary tumours occurring within any cardiac chamber, but mostly they have a predilection for the left atrium.[1,2]

Classic manifestations with which they present include constitutional, obstructive and embolic symptoms. Constitutional symptoms are the least common manifestation and include fever, myalgia, arthralgia, and weight loss. They can also produce symptoms of haemodynamic derangement from obstruction of the flow within the cardiac chambers, obstruction of the pulmonary or systemic venous drainage, or deformation of a cardiac valve. Systemic emboli occur in 30-45% of patients with left atrial myxoma. Embolisation to the spleen, adrenals, intestine, kidneys, abdominal aorta, coronary arteries, and mesentery and limb arteries have been reported in 10-45% of cases. The cerebral arteries, including the retinal arteries, are infected in most patients. Neurological manifestations of atrial myxomas are frequent and have been reported in 25-45% of cases. Unusual manifestations of atrial myxoma include the development of multi-infarct dementia due to recurring embolism, spinal artery embolism, spreading of tumour cells to the superior sagittal sinus and, rarely, leakage from peripheral myxomatous aneurysms.[3-5]

Gerstmann's syndrome is a neurological disorder characterised by four primary symptoms: a writing disability (agraphia or dysgraphia), a lack of understanding of the rules for calculation (acalculia or dyscalculia), an inability to distinguish right from left, and an inability to identify fingers (finger agnosia). In adults the syndrome may occur after stroke or in association with damage to the parietal lobe. This disorder is often associated with brain lesions in the dominant (usually left) side of the angular and supramarginal gyri near the temporal and parietal lobe junction. In addition to the above symptoms, many adults experience aphasia, difficulty in expressing themselves during speaking, in understanding speech, or in reading and writing. Development of Gerstmann's syndrome due to left atrial myxoma is extremely rare and to our knowledge has been reported only once in the medical literature.[5] In the case presented here, our patient was admitted with headache, dizziness, a reported lost of conscious, confusion of right and left, inability to identify fingers, dysgraphia, occasional errors in speech and dyscalculia when asked to perform simple mathematic procedures. The above symptoms indicated the presence of Gertsmann's syndrome. The auscultation finding of cardiac murmur was the

cornerstone in identifying the presence of a left atrial myxoma, which was the cause of the neurological disorders by embolisation.

The diagnostic procedures for identifying Gerstmann's syndrome are a CT scan and magnetic resonance imaging of the brain. In our case the initial CT scan was interpreted as normal and did not confirm the diagnosis with the radiological manifestations. This finding has been reported before.[6]

The treatment for Gerstmann's syndrome is symptomatic and supportive. In adults the symptoms diminish over time. Speech therapies and occupational therapies may help diminish dysgraphia and apraxia.

Transthoracic echocardiography is the most frequently employed non-invasive diagnostic procedure for detecting a cardiac mass. The test is accurate and highly sensitive. Better results have been reported from the use of transoesophageal echocardiography. Some recent papers have reported the usefulness of coronary angiography in further defining the diagnosis of myxoma by demonstrating signs of neovascularisation, and the usefulness of cardiac magnetic resonance imaging for demonstrating specific findings in favor of myxoma.[7]

Cerebral embolism and neurological signs and symptoms remain a frequent manifestation of left atrial myxoma. Although the existence of atrial myxoma is a rare entity, it must be considered in the differential diagnosis of the causes of stroke and cerebral ischaemia, especially in young patients. Cardiac auscultation and transthoracic echocardiography must be considered in all patients with no apparent causes for cerebral ischaemia.

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