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CEREBRAL ISCHEMIA AS INITIAL NEUROLOGICAL MANIFESTATION OF ATRIAL MYXOMA

Case report

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ABSTRACT - Cerebral infarctions of cardiac etiology are observed in around 20% of patients with ischemic stroke. Cerebral ischemia is the first clinical manifestation in 1/3 of cases of atrial myxomas. Although almost half of patients with atrial myxoma show changes at neurological exam, non-hemorrhagic cerebral infarction is seen in computed tomography in practically all cases. We present the case of a 40 year-old woman whose first clinical manifestation of atrial myxoma was an ischemic stroke. We point out to the possibility of silent cerebral infarction in atrial myxoma patients.

KEY WORDS: brain embolism, atrial myxoma, ischemic stroke.

Isquemia cerebral como manifestação neurológica inicial de mixoma atrial: relato de caso

RESUMO - Infartos cerebrais de etiologia cardíaca são observados em cerca de 20% dos pacientes com acidente vascular cerebral isquêmico. Infarto cerebral ocorre como manifestação clínica inicial em um terço dos casos de mixoma atrial. Embora quase metade dos pacientes com mixoma atrial apresente alteração ao exame neurológico, infarto cerebral não hemorrágico é visto na tomografia computadorizada em praticamente todos os casos. Os autores apresentam o caso de uma paciente, cuja primeira manifestação clínica do mixoma atrial foi um acidente vascular cerebral isquêmico e chamam a atenção para a possibilidade de infarto cerebral silencioso em pacientes portadores de mixoma atrial.

PALAVRAS-CHAVE: embolia cerebral, mixoma atrial, infarto cerebral silencioso.

Cerebrovascular diseases (CVD) are the main cause of death and permanent handicap in Brazil[1,2]. Around 14% to 20% of ischemic CVDs are of cardioembolic etiology, among which the most important ones, considering emboligenic potential, are atrial fibrillation, acute myocardium infarction, cardiac valvular disease, infectious endocarditis, and atrial myxoma, which is responsible for 0.4% of cases[3,4].

Atrial myxoma usually manifests as a mitral valve obstruction. Neurological symptoms are not frequent as initial showing of this tumor[5,6].

We report the case of a patient with atrial myxoma whose first clinical presentation was due to cerebral ischemia.

CASE

40-year-old white woman showed light frontal cephalea and vomits, followed by right hemiparesis, mental confusion and disorientation for 8 hours, with a partial improvement in the subsequent hours. She had been a smoker for 15 years and had arterial hypertension for 4 years. Clinical exam showed an arterial tension of 110x80 mmHg, pulse frequency of 80 per minute, unaltered cardiac auscultation, preserved peripheral pulses, clean lungs, and normal abdomen. The patient was confused, apathetic, with right hemiparesis with brachial predomination (degree IV+). Fundoscopy was normal in both sides. The exams of the senses and the cranial nerves were normal. Meningeal signs were absent[8]. Laboratorial exams showed peripheral blood white cells (9.2 x10^3/mm^3), red cells (5.17x106/mm^3), hematocrit (46.2%), hemoglobin (15.5%), platelets (265000/mm^3), glycemia (151.0 mg/dL), electrolytes (calcium: 9.0 mg/dL; magnesium: 2.20 mg/dL; potassium: 3.70 mmol/dL; sodium: 140.0 mmol/dL), cardiac enzymes (CKMB: 1.00 U/L; CPK: 25.00 U/L, prothrombin activity (87.5%), INR (1.15) and lipid profile (total cholesterol: 168.00 mg/dL; LDL-cholesterol: 116.40

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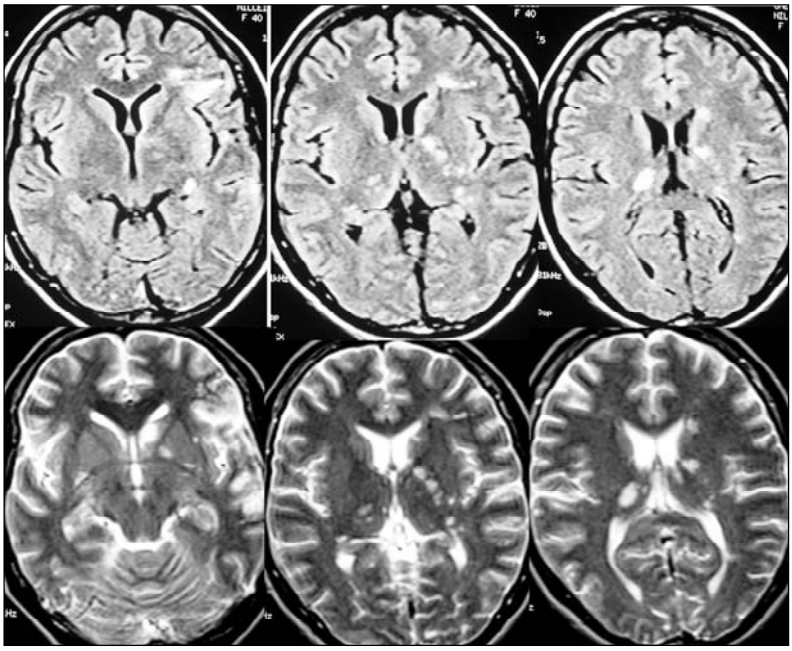

Fig 1. Magnetic resonance in FLAIR and T2 sequences, showing multiple areas of cerebral embolism.

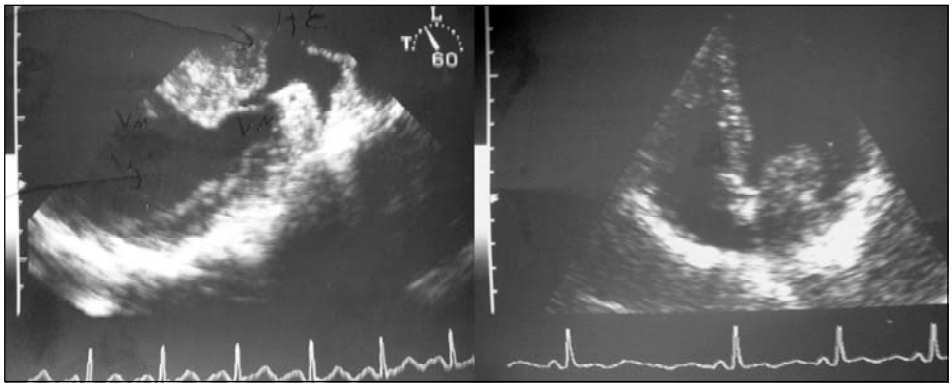

Fig 2. Transesophageal echocardiogram: left atrial myxoma measuring 38x26 mm, irregular surface, jutting out at entrance of the left ventricle during diastoles, with normal mitral valve flux; normal cavities, dimensions and dynamics, without spontaneous contrast in atrium; septum without solution of continuity; aorta with normal morphology.

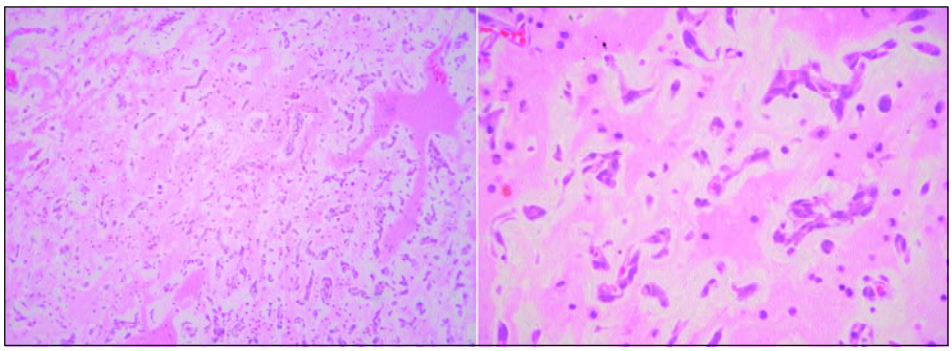

Fig 3. Microscopy (H-E): histological cuts reveal cellular proliferation with formations of small cords or isolated cells, showing large eosinophilic cytoplasm, with indistinct edges, round nuclei with open chromatin and inconspicuous nucleoli, arranged on a myxoid background. Hemosiderophages are found in the lesion. Pleomorphism and atypical mitoses are not viewed.

mg/dL; HDL-cholesterol: 32.00 mg/dL; triglycerides: 98.00 mg/dL). VDRL and Chagas disease (ELISA) were not reactive. The electrocardiogram (ECG) was normal. Magnetic resonance (Fig 1) showed multiple areas of cerebral infarction. Transesophageal echocardiogram (Fig 2) revealed the presence of a left expansive lesion, possibly indicating atrial myxoma.

The patient had surgery and the diagnosis of atrial myxoma was confirmed (Fig 3). In the postoperative, the patient did not show other neurological events and she was kept for observation in the ward.

DISCUSSION

Atrial myxoma accounts for approximately 50% of surgeries conducted for cardiac tumors[7] and preferably strikes women (3 women to 2 men) from the third to the sixth decade, with an average of 43 years of age[8]. This tumor is infrequent during childhood[9]. It usually comes up as symptomatic triad, constituted by: mitral valve obstruction symptoms (67%), manifesting itself as cardiac insufficiency and weakness; embolism symptoms (29%), especially for brain and peripheral vessels; and as systemic symptoms (34%), such as fever and weight loss.

Neurdological manifestations in patients with atrial myxoma are reported in 25% to 45% of cases[10,11] and may be secondary to cerebral infarction, cerebral hemorrhage and, more rarely, subarachnoid hemorrhage. Other neurological manifestations observed are syncopes (28%), psychiatric symptoms (23%), cephalea (15%) and epileptic fits (12%)[8,12,13]. Recurrent cerebral infarctions are common before the resection of this tumor and are caused by emboli through myxomatous material or through thrombi[6,14-16]. Fusiform or saccular aneurysms may be observed distally in intracranial arteries[6,17,18]. These aneurysms can be asymptomatic[19], or even not detected by angiography[16,17].

Neurological manifestation as initial presentation of atrial myxoma is found in 36% of cases, although 45% of cases have abnormalities on neurological exams and practically all patients present non-hemorrhagic cerebral infarction at computed tomography[6].

Cardiac auscultation is normal in 36% of patients and ECG can show only unspecified alterations[6]. There is a correlation between tumor size and the alterations at cardiac auscultation and at ECG, which occur predominantly in larger tumors[5]. The diameters of these tumors range from 1 to 15 cm and weigh from 15 to 180 grams (average: 37 g) and have a friable surface or villosities in 35% of cases[5]. Our patient had normal cardiac auscultation and ECG, and the diagnosis was done after transesophageal echocardiogram. Magnetic resonance techniques were not necessary, but may be used if necessary.

The use of recombinant tissular plasminogen activator (rt PA) in the acute phase may be an option for patients with atrial myxoma because there are evidences of thrombi adhered to the tumor. However, its use should be preferably done intra-arterially owing to risk through the presence of asymptomatic aneurysms, which could be detected through angiography before procedure[16,20,21]. Our patient came to the hospital with 8 hours of evolution and with a deficit in partial regression, which excluded reperfusion. Although there is uncertainty as to the use of anticoagulants in these patients, we opted for intravenous heparin to prevent new cerebral emboli.

The prevalence of silent infarctions in the general population is estimated at 11% to 18%[22]. In the patients with atrial myxoma, there are already evidences on neuroimaging of multiple asymptomatic infarctions at diagnosis[6], showing that probably in these cases, as it is for our patient, there may be a higher risk of future cognitive disturbance owing to the multiple cerebral ischemias, especially if the tumor is not operated on or if the diagnosis is done late. Therefore, surgery has to be performed as soon as possible, even on asymptomatic patients, as secondary prevention of cerebral infarction[6,23]. Surgical excision is generally curative and may be done in 69% of cases[8]. Neurological events after surgery are rare[6]. Our patient underwent surgery and remained hemodynamically stable and was released from the hospital without complications or new manifestations of cerebral ischemia.

We conclude that ischemic cerebral vascular accident may indicate the presence of an atrial myxoma, and that silent infarction patients should undergo investigation of this cardiac pathology, since its diagnosis is important to establish a quick surgical conduct, in order to avoid the occurrence of new cerebral events.

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