Research Article

« Myxoma Of The Left Atrium Responsible For Fatal Cerebrovascular Accident »

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Received: 22 June, 2023
Accepted: 10 February, 2024
Published: 21 February 2024

Abstract:
Left atrial myxoma is a rare tumor. The polymorphism of its clinical presentation is sometimes confusing for the clinician. Although its histologically benign nature is recognized by the majority of authors, its intracardiac location gives it a significant lethal evolutionary potential in the absence of surgical treatment. We report here the case of a 38-year-old patient who presented with a massive ischemic stroke.

Key words: stroke, myxoma, tumor.

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Left atrial myxoma is a rare tumor. The polymorphism of its clinical presentation is sometimes confusing for the clinician. Although its histologically benign nature is recognized by the majority of authors, its intracardiac location gives it a significant lethal evolutionary potential in the absence of surgical treatment. We report here the case of a 38-year-old patient who presented with a massive ischemic stroke.

A 38-year-old truck driver was referred to the emergency department with an acute onset of right hemi-body deficit while climbing into his truck. The family history revealed no previous history. On clinical examination, the patient was drowsy, with no fever. Glasgow score was 8. Right hemiplegia with aphasia was noted. Osteotendinous reflexes were absent on the right. There was no meningeal syndrome or Babinski sign. Cardiac auscultation showed no murmurs or rubs. Blood pressure was 143/50 mmHg, pulse regular at 80 beats per minute. The electrocardiogram showed sinus rhythm at 80 beats per minute, minute with no repolarization or conduction abnormalities. The rest of the clinical examination was unremarkable. Biologically, the blood ionogram was normal, as was the complete blood count (hemoglobin: 12 g/dl, platelets: 150,000/mm). Fibrinogen was 5.7 g/L, C-reactive protein 10, sedimentation rate 8 mm at hour 1 and 15 mm at hour 2. Tests for deficiency of certain coagulation factors (antithrombin III, proteins C and S) were negative. The patient immediately underwent an injection-free cerebral CT scan, which revealed very discrete cortico-subcortical dedifferentiation in the left sylvian region (Fig. 1), and at the same time, a carotid echo-doppler, which showed, in the left internal carotid artery, a hyper-resistive spectrum of the bulbar region with absence of post-bulbar flow, reflecting thrombosis of the suprabulbar carotid artery. At the 24th hour of hospitalization, he underwent transthoracic cardiac echocardiography, which revealed a tumour.

A transesophageal cardiac ultrasound was then performed. This revealed a tumour in the left atrium (dimension 5 x 3 cm), the appearance of which was highly suggestive of a myxoma (figure 2).

At the same time, the neurological condition worsened, with the onset of respiratory pauses and neurovegetative disorders, necessitating tracheal intubation and mechanical ventilation. A new CT scan was then performed.

Two major ischemic lesions were visualized: one in the left sylvian territory and the other in the left posterior artery territory, associated with diffuse bi-hemispheric edema (figure 3). At the level of the foramen magnum, there was involvement of the cerebellar tonsils. Finally, the patient died at the 72nd hour in a clinical picture of cerebral involvement.

Discussion:
Cardiac tumors are relatively rare and generally benign: 70% of them are myxomas, whose frequency in the general population is around 0.035% in post-mortem series [1]. Myxomas are mainly located in the atria (15% in the right atrium, 85% in the left) [2]. It is a solitary, pedunculated tumor implanted on the inter-atrial septum. The friability of the myxoma explains the risk of migration of tumor fragments.

Functional signs reflect three different pathophysiological mechanisms: obstructive signs, of which exertional dyspnea is one of the most frequent, occurring in two-thirds of cases [3]. Lipothymia and syncope, and even sudden death, have been reported in cases of tumor entrapment in the lung.
General signs are relatively frequent (50-85% of cases) [5] and do not necessarily point to cardiac pathology, leading to diagnostic delays of several months or even years.

Weight loss and fever are the most frequent symptoms [3]. Our patient had cases of facial paralysis associated with aphasia also correspond to embolism in the left sylvian territory, and should not be distinguished from right hemiplegia with aphasia. Iterative embolisms occur in a third of cases. In most cases, they are complicated by cerebral softening. More rarely, fusiform aneurysms may occur as a result of infiltration of the arterial wall by myxomatous tissue, which destroys the internal elastic boundary and the media [1]. Secondary metastasis is exceptional [7].

Cerebral complications of cardiac myxoma have a mortality rate of around 20% in Bulkley's study of a series of 25 myxomas [8]. Although most myxomas are sporadic cases, around 7% of them run in families (with autosomal dominant inheritance), hence the value of systematically performing an ultrasound scan on all family members to look for them. This search proved unsuccessful in our patient's family.

No signs that might have attracted attention; functional signs related to an embolic phenomenon: arterial embolisms occur in 25 to 40% of cases, depending on the author [3]. In half the cases, these are cerebral embolisms, most often in the left sylvian territory. Neurological manifestations dominated by right hemiplegia or aphasia, as in our observation, often inaugurate the clinical picture. Dementia has also been described to set in, rapid [6], episodes disorientation.

Biologically, there is no definitive evidence for the diagnosis of atrial myxoma, only a cluster of arguments. An inflammatory syndrome is encountered in over 70% of cases (elevated sedimentation rate and C-reactive protein).

Anemia is often marked, of inflammatory or hemolytic origin, as a result of mechanical destruction of red blood cells by the blood stream.

Transesophageal cardiac ultrasound appears to be the gold standard for diagnosis. It provides better resolution and accuracy in the diagnosis of cardiac myxomas, but above all in the surgical approach, since it enables tumor implantation pedicles to be located more accurately.

Treatment is surgical as soon as the diagnosis is made, and as quickly as possible, with low operative and perioperative mortality for trained teams (2%) [9].

Other primary tumors are mainly ventricular myxomas, responsible for systemic embolisms, often cerebral, occurring in two-thirds of cases. In contrast, general signs are remarkably absent.

Rhabdomyomas and fibromas occur mainly in newborns and children. Most primary malignant tumors of the heart are sarcomas, angiosarcomas and rhabdomyosarcomas. The prognosis is generally poor, with very short survival. Secondary heart tumors are 20 to 40 times more frequent than primary tumors. They are by no means specific, and give rise to cardiac
symptoms. They most often reach the heart via the hematogenous or lymphogenous route, sometimes by direct invasion.

The originality of the case presented here lies in the absence of functional and biological signs prior to the acute episode, and in the fact that the disease is fatal within a few hours, whereas it is generally fatal several months after diagnosis. Although this is a rare event (less than 0.5%) [10] in the etiological assessment of a stroke, an intracardiac cause, such as myxoma, should be systematically sought, particularly in young subjects with no cardiovascular risk factors.

References: