Systemic manifestations of a cardiac tumour

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Abstract

The authors present a case report of cardiac myxoma, manifested by two cerebrovascular accidents and an event emulating a vasculitis process.

Considerations are made on clinical features, differential diagnosis, treatment and prognosis.

Key words: myxoma, cerebrovascular diseases, embolic effect.

Introduction

Primary cardiac tumours are rare with an incidence in autopsies series of 0.0017 to 0.05%, according to Pernot and 0.35, according to McAllister.[1] From these tumours, 75% are benign and 25% malignant. Regarding the benign tumours, 50% are myxoma and 20% rhabdomyoma.

Myxoma was recognised for the first time by Columbus in 1559 (in the necropsy exam), and only in 1952 Goldberg et al. made the first pre-surgical diagnosis, through angiocardiography.[2]

In a bibliographic review of the international literature (articles published from 1971-1994), myxoma incidence changes from one series to the other, with a higher frequency being seen from the 3rd to the 6th decade of life, being very rare in children.[3,4,5,6,7,8,9]

Regarding its frequency according to gender, a predominance of 3:1 female compared to male is described.[3]

At national level and aware that the cases published do not indicate the frequency of myxoma cases diagnosed, we made a bibliographic research of the Portuguese literature (from 1989-1994) having only recorded 4 cases.[21,22,23,24]

Most authors consider myxoma as a true benign neoplasm with malignant potential, while others claim that this is the result of thrombus organisation, formed in the endocardial surface and suffered myxomatosous degenerescence.[4]

Cardiac myxoma can be occasional (90%), of the complex type or syndrome (7-10%).[10]

Clinically, they can emulate a variety of other cardiac diseases (cardiac failure; pericarditis; endocarditis; myocardiopathy; pulmonary hypertension) and non-cardiac (autoimmune diseases/vasculitides, infections; neoplasms; cerebrovascular accidents; haematology diseases, for instances, anemia, thrombocytopenia, polycythemia) being necessary a high degree of suspicion to achieve a diagnosis.[2,11,12,13]

Diagnosis should be made at the earliest possible stage, as it is a clinically entity potentially treatable with a reduction of the morbidity and mortality rates.

Case report

M. J. C. S., a 42 years-old patient, female, Caucasian, health worker, divorced, born and residing in Lisbon.

Admitted through the Emergency Service on the 14th December 1994, due to a condition of space and time disorientation, mental confusion and right hemiparesis.

Apparently in good health condition until 2 years ago, when she started complaining of persistent cervical-dorsal pain and the painful intensity was accentuated with postural movements, causing occasionally complaints of headaches and paresthesia in the upper limbs. Simultaneously, petechial lesions and subungueal ecchymoses appeared.

She had an appointment with a Rheumatology consultant being subject to several tests (in the sense of identifying any change of coagulation, autoimmune diseases, Lyme's disease) that, according to infor-

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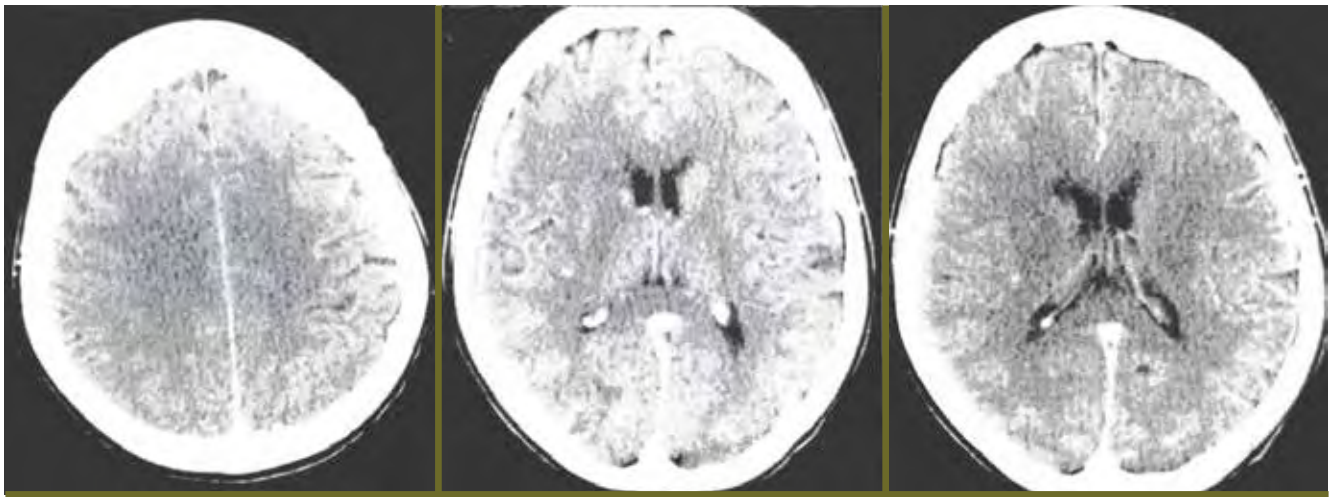


CE-CT Scan (18/09/94). Small hypodense images, right temporal and parietal and a heterogeneous image, right frontal polar (ischemic? neoformative?)

FIG. 1, 2, 3

mation collected, would all be negative. However, regarding the clinical condition, the hypothesis of being a vasculitis was kept, therefore corticosteroid therapy was started, and a reversion of the lesions and petechia was verified with an improvement of painful complaints.

In March 1994, cervical pain restarted with increasing intensity associated to paresthesia and decrease of muscular strength in the upper limbs. Radiography and CT scan of the cervical spine were carried out, being diagnosed a disk hernia between C6-C7 reason why she underwent surgical intervention. During the post-surgical period small hemorrhages in the hands subungueal bed reappear (keeping clotting tests within normal ranges) which revert with an increase on the corticosteroids dosage.

At that time, a depressive condition sets in with periods of mood swings. In an attempt to clarify such condition, she is seen by a Neurologist and subject to several diagnosis additional tests, including lumbar puncture with cytochemistry, bacterial and immunology tests and serologic research of several viruses (tests we did not have access to) which did not reveal changes (according to information supplied by relatives and confirmed by the Neurology colleague). After therapy with amitriptyline and anxiolytics an improvement on symptoms was seen.

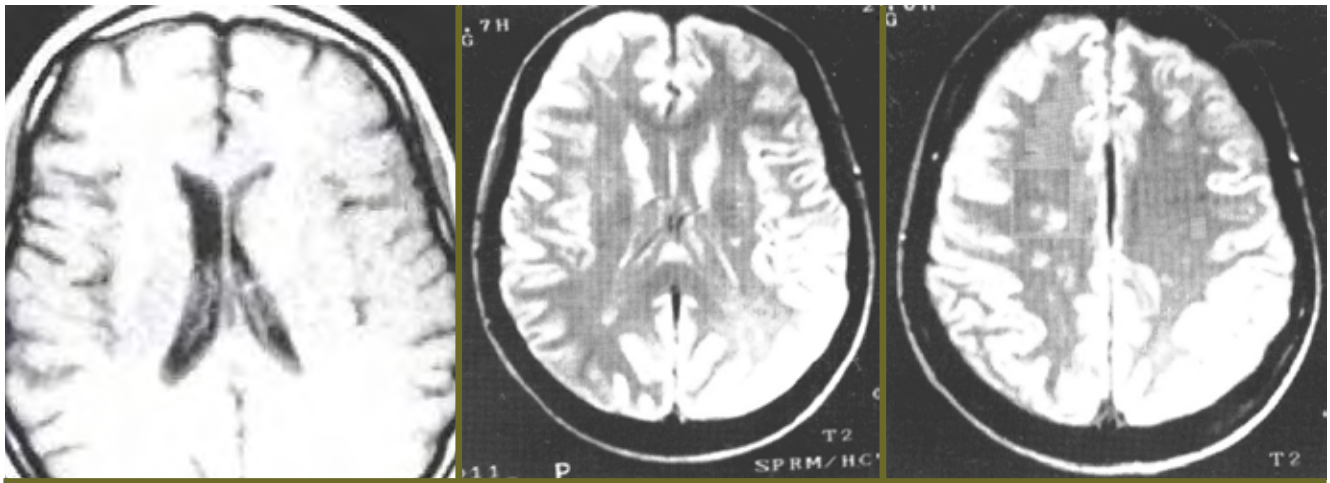
Suddenly, in September 1994, the following condition appears with left hemiparesis, left facial paresis, of central type and sphincter incontinence. She is admitted to hospital undergoing CE-CT scan (*Fig. 1,2,3*) and CE-NMR (*Fig. 4, 5, 6*) showing :"small ischemic lesions, vasculitis type, on the right cerebral hemisphere". After recovery therapy, she makes a progressive and total recovery, keeping a wide base of gait.

On the 13th Dec 1994, during an episode of emotional stress and high anxiety, emerges a condition of mental confusion, space and time disorientation, incoherent speech and right hemiparesis, of brachial predominance emerges, reason why she is admitted to hospital.

There is no previous history or following febrile episodes, dizziness, vertigo, hearing and visual changes, cardiac and respiratory complaints, as well as gastrointestinal or genital-urinary.

Personal background: at 16 years of age, she underwent appendectomy; in 1982 a fracture of the right femur (road accident); in 1989, a diagnosis of uterine myoma, she denies smoking, alcohol or illegal drugs intake, as well as promiscuous sexual behaviour, she denies hypertension, diabetes, diseases of cardiac and respiratory type, menarche at 12 years of age. I G I P – eutocia labor, male newborn alive.

Family background: irrelevant; objective exam: (positive elements); normal general condition. Looking the age she had. Skin and mucosa coloured and hydrated. Without petechial lesions or ecchymosis. No edema. Eupnea. Axillary temperature – 36.5°C.



CE-NMR (26/09/94). Suggestive aspects of ischemic lesions, vasculitis type.

FIG. 4, 5, 6

Blood pressure 150/80 mmHg. Pulse (radial, right) - 70 bpm, regular, rhythmic, wide. Carotid pulse without changes. Without visible venous jugular ingurgitation. Non palpable thyroid. The thorax showed no changes, namely at cardiac auscultation (carried out several times, in different positions) as well as the pulmonary. There were no palpable organomegalies in the abdomen. Neurologic test: patient awake, apathy, with monochordic voice and monosyllabic speech. Disoriented to time and space relating to herself. Marked naming deficit, kept understanding. Without meningeal signs. Funduscopy – pupillary pallor on the left. Right hemiparesis of brachial predominance. Without sensitivity changes. Decreased osteotendinous reflexes on the right. Cutaneous plantar reflexes while bending to the left and outlines extension on the right.

Additional tests carried out before admission: CE-CT scan (18/09/94): "Small hypodense images, on the temporal-parietal right and a heterogeneous image on the polar front right (Ischemic? Neoformation?). CE-NMR (26/09/94): "Aspects suggesting ischemic lesions, vasculitis type". Carotid Echo Doppler and transcranial Doppler (30/09/94): "Pattern of anatomic and hemodynamic normality of the supra-aortic trunks".

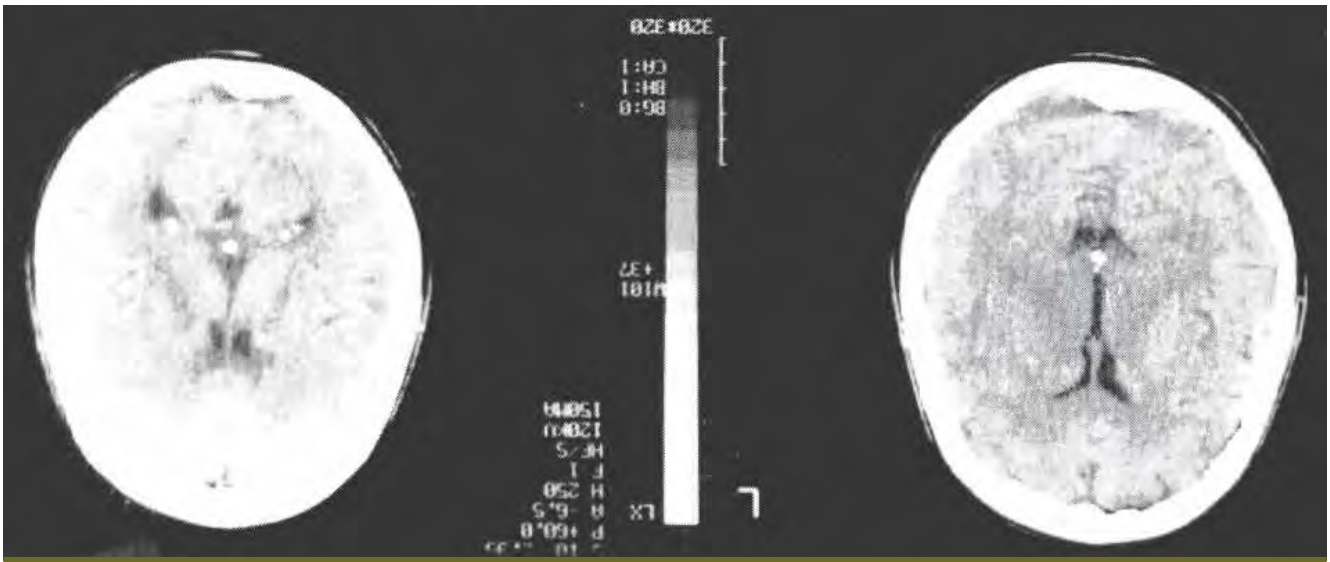
Additional tests carried out during hospitalization:

Hemogram: normal with ESR of 20mm in the 1st hour. Kidney and liver functions: normal. Total protein 7.7 g/dL with normal globulin. Mantoux 10 U negative. Negative serology (A, B and C Hepatitis; syphilis; HIV1 and HIV2; toxoplasmosis; CMV; Epstein-Barr; H. simplex; borrelia).

Autoimmune disease markers (C3; C4; CH100; C50; CRP; RA Test; Waller/Rose; ANA; anti-DNA; anti-RNA; anti-RNP; anti-Ssa; anti-SSm; anti-SSb; anti-cardiolipin antibodies) within normal ranges.

CSF: normal cytobiochemistry test; negative tests for bacteriology, direct and cultural, mycology, parasitology and KB (direct and cultural). Negative antibodies research for anti-borrelia, CMV, EBV, HSV, toxoplasmosis. Protein immune electrophoresis study without changes. Total study of coagulation with chromatographic profile of serum and urinary amino acids: within normal ranges. ECG without changes. Thoracic teleradiogram without changes. CE-CT scan (13/12/94). "Some scattered lacunar lesions, left temporal and right frontal. Without evidence of acute vascular lesion." (*Fig.* 7). CE-NMR (19/12/94): "Compatible aspects with vasculitis processes, reaching areas of vascular distribution of small vessels (perforating: perforating arteries/penetrating arteries function), predominantly on the left" (*Fig. 8,9*). Digital angiography of the supra-aortic vessels without changes.

EEG: "Diffuse slowing of the electrogenesis base, with focal marking on the temporal area of the left cerebral hemisphere. Scarce paroxysmal activity with the same location". Echocardiogram (05/01/95): "bulky intra-atrial mass on the left, with a partial



CE-CT scan (13/12/94). Some scattered lacunar lesions, left temporal and right frontal. Without evidence of acute vascular lesion

FIG. 7

prolapse to the left ventricle, in systole." (*Fig. 10*). Transeseophageal echocardiogram: "it is confirmed the existence of mass on the Left Atrium (23 x34), mobile, fringed, inserted in the IA septum, at the level of the foramen ovale, with a prolapse into the left ventricle".

Clinically, from the 2nd day of admission onwards, there is a reversion of the hemiparesis. She keeps periods of mental confusion and disorientation to space and time.

On the 6th January 1995, she is referred to the Cardiothoracic Surgical Service of Hospital Sta Marta, undergoing a surgical resection of the tumor.

The histological test has confirmed to be a myxoma (polypoid formation with around 1 cm diameter, of gel like soft consistency).

The echocardiogram carried out afterwards has revealed: "Valves without changes. Left ventricle of normal dimensions, with good overall systolic function. Normal left atrium and right cavities. No intracardiac images and no pericardial effusion.

Comments

Myxomas grow from the endocardium surface of any of the 4 cardiac chambers or septa and tend to invade them:[11] left atrium (75%); right atrium (18%); similar to each ventricle (7%). 85% cases, are originated in the inter-atrial septum (more common in the *fossa ovalis*).

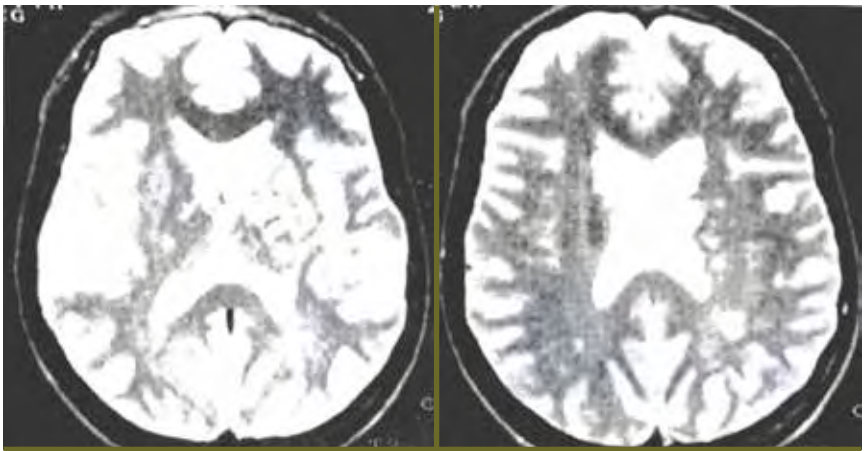
Macroscopically they are friable, 4/8 cm in diameter, gelatinous consistency, polypoids/viliforms (80%), globular (20%); often calcified. Poorly vascularized, it can have areas of necrosis or hemorrhage.[2]

Microscopically, they are made up by polyedric cells with a small and round core, scattered in a myxomatous eosinophilic stroma. In the stroma there is a predominance of mucopolysaccharides, lymphocytes, hystiocytes, plasmocytes and mastocytes.[2]

Before a background of 2 cerebral vascular accidents, located in different territories, in a female patient, young, without arrhythmias, with a coagulation study and normal immunologic markers and without changes in the study of supra-aortic vessels, the hypothesis of a possible cardiac etiology, in spite of the objective exam, the ECG and the thoracic teleradiogram did not reveal changes.

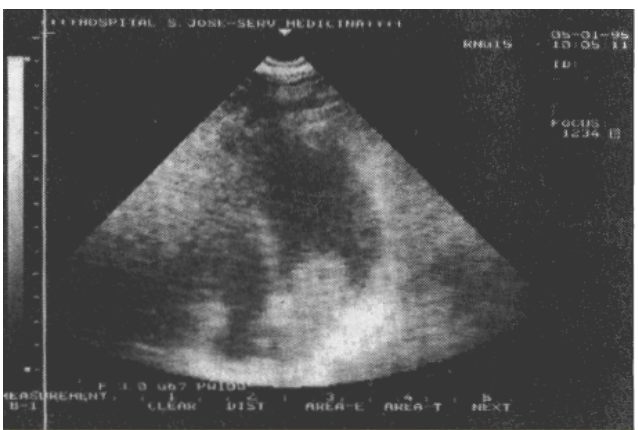
Carrying out the echocardiogram has enabled the diagnosis while making evident a bulky mass in the left atrium, confirmed through transesophageal echocardiogram.

Such tests are diagnosis additional tests crucial to such pathology. They enable to clarify the occasional existence of intracavitary lesions, endo- or myocardial, to identify the structure of the myxoma (necrosis foci, cysts); to detect bilateral myxomas; to enable the intra-operative monitoring of removing the tumor.[3,11,13,14,15]



CE-NMR (19/12/94). Compatible aspects with vasculitis process, reaching territories of vascular distribution of small vessels predominantly on the left side

FIG. 8, 9



Echocardiograma(5/01/95). Left intra-atrial bulky mass, partially prolapsing into the left ventricle during systole.

FIG. 10

After the surgical resection, the tumor histologic exam showed to be a left atrium myxoma.

Myxomas, due to their characteristics (number, location, dimensions, structure and invasive capacity) can show themselves with a variety of symptoms: obstructive, general or embolic. [2,11,12,13,16,17]

The obstructive symptoms result of the mass, mobility, invasion of valvular orifices and/or cardiac chambers which leads to a clinical condition suggesting heart failure (right or left), pulmonary hypertension, pulmonary infarction, arrhythmia and upper vena cava obstruction.

In the case presented, the patient denied, persistently, any complaint of the cardiorespiratory forum; the objective exam did not show signs of peripheral venous congestion nor changes in the cardiac auscultation. The same way, the ECG and the thoracic teleradiogram did not reveal any changes.

This fact is interesting, as there was a bulky myxoma, pediculated, protruding into the left ventricle and would lead to signs and symptoms of left heart failure, which was not verified.

Regarding the complaints of cervical and dorsal pain, paresthesia, petechial lesions and subungueal ecchymoses, which were construed initially as a possible vasculitis dysfunction leading to the start of steroid therapy, there were no more than some general and systemic symptoms and embolic which can emerge associated to the myxoma.

Embolic processes may be systemic or pulmonary. Systemic occur in 30-70% of patients with myxoma on the left atrium due to the obstruction of the arteries (brain, kidneys, lower limbs, aortic bifurcation) by tumor fragments or the thrombus adhering to the myxoma.[3,11,17]

In the literature it is described an incidence of embolism in the central nervous system in 10-45% of patients with myxoma, being affected more commonly the left hemisphere and the territory of the medial cerebral artery.[6]

In such situations, the neurologic symptoms represent often, the only manifestation of the existence of the myxoma and are different according to the location and the lesions extent. In fact, it can be stated the myxoma can be the treatable cause of the cerebral vascular accident.

From a neuroradiology point of view (CE-CT scan, CE-NMR and angiography) ischemic lesions may emerge, whether or not lacunar, hemorrhagic lesions, local changes in the arteries walls which can go from dilation and irregularity to the formation of aneurysms.[18]

Therefore, in the case presented, the first embolic process manifested itself with a left hemiparesis condition and homolateral facial paresis related with right

temporal-parietal facial paresis and the second with a condition of mental confusion, disorientation to space and time and right hemiparesis, due to scattered lacunar lesions. Angiography did not show any changes.

The myxoma can represent the biggest source of neurologic morbidity. Its resection reduces morbidity to 5% but can have neurologic complications a few years after intervention.

A myxoma reoccurrence of 5-7% of cases (total resection? Multiple development foci? Malignant transformation? Tumor metastization?) therefore a regular follow up is mandatory.[19]

This pathology diagnosis can be lengthy (cases evolving for 5-20 years before diagnosis and even cases where diagnosis was only achieved during necropsy are described). As a matter of fact, most small myxomas are asymptomatic and on the other hand, when acquiring dimensions that lead to symptomatology, can be misconstrued, if such nosologic entity is not present.

Prognosis will depend on the length of time the disease evolves, its consequences, the myxoma characteristics (dimension, location, size of the pediculum, possibility of causing obstruction or being a source of embolization by the tumor material).

In cases where only the medical therapy is carried out, the prognosis is reserved, and syncope or sudden death can occur; but when the radical tumor resection is performed with bypass a survival rate of 71% is described.[20] ■

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