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CASE REPORT

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Carney syndrome and cardiac myxoma complicated with polycystic embolism

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Introduction

Described in 1985 [1], Carney syndrome is a genetic disorder characterized by the association of pigmentary skin abnormalities, myxomas, dysfunction of endocrine tumors and schwannomas. Skin abnormalities are the most characteristic manifestations and occur in the form of a black or brown skin lens that increases at puberty. Cardiac myxomas occur at any age and occur in any cardiac cavity. Complications include systemic and pulmonary embolism, heart failure, myocardialinfarction, and cardiac arrest. The myxomas also occur in the skin, breast, oropharynx, and female genitalia, and one-quarter of these patients have Cushing's syndrome. The diagnosis of this rare pathology is usually made around 20 years [2-4].

Clinical case

A 41-year-old male patient hospitalized for febrile conditions accompanied by a significant biological inflammatory syndrome, asthenia, weight loss, dyspnea and temporo-spatial disorientation, with a cardiovascular history: of the 4 vascular accidents Cerebral palsy, cerebral palsy, cerebral palsy, cerebral palsy, cerebral palsy, cerebral palsy, cerebral palsy, cerebral palsy, dyslipidema.

On clinical examination

Patient disoriented, febrile, dyspneic (stage II of NYHA), temperature at 39°C, blood pressure at A 160/90 mm hg, weight 100kilos, IMC 32, saturation 85%.

Heart

Regular heart sounds, mitral systolic murmur 2/6, accompanied by a diastolic roll, sharper when changing position, absence of gallop and pericardial friction. Abdomen supples to palpation, painless, no masses swinging or viscero-megalia, normal peristaltic noises. Extremities: no edema, absence of clinical signs of thrombo-phlebitis (no decrease in passive ballooning and cord endured), absence of signs of lymphangitis, contralateral left and right distal left pulse, left lower limb temperature In relation to the reduced duty.

Neural system

Presence of signs of motor and sensory deficit in the lower left limb; no laser, no leri, no sphincter deficit, discrete intermittent claudication of the lower limb.

ECG

Sinus rhythm, at 80 bpm, left axial deviation with left ventricular hypetrophy, absence of rhythm and Atrio-Ventricular conduction disorders, presence of antero-septal sequelae. Computed tomographic scan of the thoracic contour: control after radiographic examination of ambiguous images: no pleural effusion or pericardial effusion, small microcalodic images entirely calcific in the 2 hemi fields pulmonary evoking sequellar images, no mediastinal lesion or pleuro-parenchymatous with an evolutionary appearance Biology: VS>85, CR 90.

A cardiac ultrasound examination was requested for vegetation research to eliminate endocarditis following the onset of an inflammatory syndrome and an infectious clinical condition. Cardiac ultrasound pre op: Device: MK60 Kontron put into service on 2/07/10 Probe: 3.5 Mhz. Echocardiography (cardiac ultrasound) in 4-cavity section and transesophageal ultrasound reveals in the left atrium the presence of a voluminous pedicle-shaped mass, bilobed, mobile, of gelatinous form inserted on the septum atrial with a Collet measured at 18mm (overall size 71 × 27 mm) proliferating the heart cycle in the mitral orifice, without clear signs of obstruction of the valvular play or obstruction phenomena, absence of suspicious images similar on the rest of the walls And cardiac cavities, free pulmonary veins.Heart dilated cardiac cavities, overall hypokinesia predominantly septal. Mean systolic dysfunction of LV with FA calculated by the Simpson method between 30 and 50% pre and postoperatively, absence of associated congenital abnormalities, dry pericardium. At the continuous doppler absence of pathological gradient between the left ventricle and the left atrium, mitral spectral curve profile I with an E/A ratio of 0.90, minimal mitral insufficiency, absence of aliasing zone at distal end of mass. At the tissue doppler absence of abnormal fills, peak S wave of 0.15 and wave L 0.20 m/s.

MRI confirms this formation, in hypersignal, particularly well enhanced at the late time after injection of gadolinium, docked at the interatrial septum by a short pedicle (Figure 1) which often penetrates the mitral orifice during ventricular filling on cinematic sequences

Different arterial embolic complications were diagnosed by cerebral MRI, angioscanner of legs and coronary angiography. Cerebral MRI performed urgently during hospitalization to show recent ischemia in the territory of the left middle cerebral, associated with possible ischemia

Straight cerebellar moderate and two sequelae of stroke, postoperative postoperative MRI, is stable compared to previous examinations. Coronary angiography to show a left and right coronary artery without plates, nor suspect image endoluminal.

Aortic arteriography and lower limbs showed occlusion by a fragment of left femoral thrombus of 5 × 0.6 cm. The patient was operated rapidly because of the severity of his clinical condition

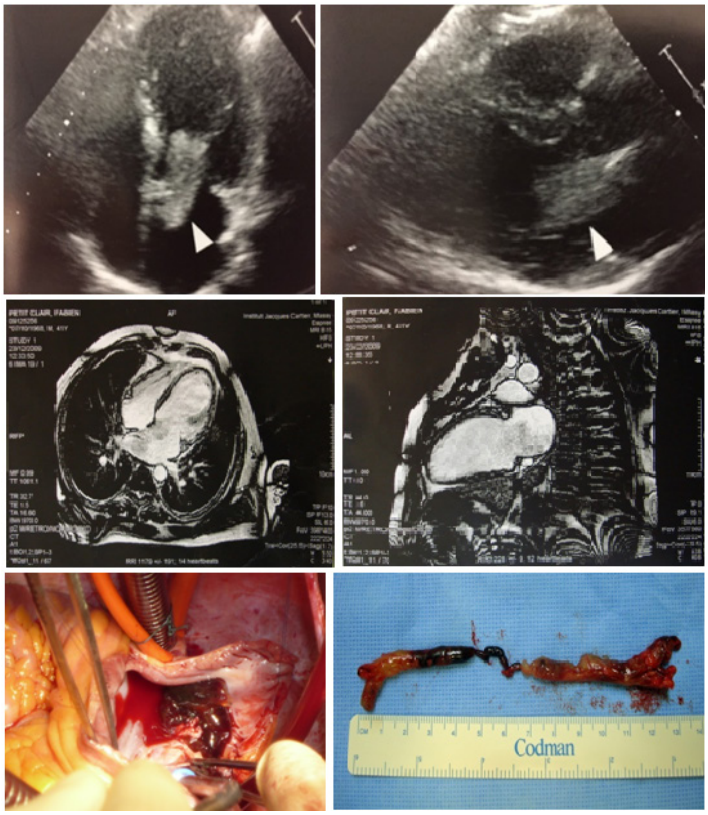

Figure 1. Imaging cardiacecho BD 4 cavities.

and the history of systemic embolism, unfortunately during the surgery, a new embolic complication occurred and this time in the left coronary, leading to an anterior Myocardial infarction and a worsening of Left ventricular systolic function. The histopathological examination of the cardiac tumor sample of 9 × 3.5 ×2.5 cm, to confirm the histological appearance of myxoma and the left femoral thrombus fragment of 5 × 0.6 cm histological appearance of recent fibrino-cruoric clot.

Discussion and conclusion

Myxoma is the most common primary cardiac tumor (40%). It is a benign tumor that is most common in Left Cardiac Headset (75%), more rarely in Right Cardiac Headset (20%) and exceptionally in a ventricle, its macroscopic appearance is variable; Irregular, sometimes lobulated or smooth and gelatinous, but it is usually connected to the endocardium by a rather characteristic pedicle, its discovery is usually between 30 and 60 years with a slight feminine predominance. It is most often isolated (95%), but it can be seen in carney syndrome or it is in multiple rule, atypical localization and sometimes associated with cutaneous and mammary myxomas, most often complicated cerebral arterial embolism. Lower limbs, splenic, renal and coronary arteries, which may go undetected in patients with carney syndrome with mental retardation.

Before the echocardiogram, which reveals it, signs very often present can evoke a myxome of the OG and are therefore to be known.An atypical cardiac murmur at the mitral orifice, often positional, linked to the frequent obstruction by the tumor of the diastolic orifice, sometimes giving a diastolic rolling. General signs such as moderate fever, arthralgia, asthenia, and embolism in 30% or dominant stroke (20%), but may also affect coronary arteries, spleen, kidney, and lower limbs, such as in our observation. The surgical procedure must be performed rapidly given the very emboligenic nature of the myxoma in these patients with carney syndrome. The surgical follow-up was simple and the patient evolved very favorably from the cardiovascular point of view. Patients with Carney's syndrome must systematically have a complete cardiovascular assessment, associated with a cardiac ultrasound to eliminate a common pathology pathology tumor in these patients.

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