Thrombosis of a Structurally Healthy Aorta with Systemic Embolic Event

The aorta is a very rare cause of systemic arterial embolism, especially if debris or aneurysms are ruled out. However, in some patients with systemic embolism it is possible to find a thrombus in an apparently normal aorta, whose natural course and best treatment are still not defined. We describe two cases of aortic thrombosis and embolism in different territories, reporting their treatment outcomes.

Case 1

We report the case of a 70-year-old female patient, with history of rheumatoid arthritis and schizophrenia, who was admitted in the emergency department due to abdominal cramp-like pain with episodes of hematemesis. The patient was afebrile and normotensive and the physical examination showed a soft abdomen, with diffuse pain on palpation and no signs of peritoneal irritation. An abdominal CT scan with oral and i.v. contrast evidenced lack of filling in the colic artery, a branch of the superior mesenteric artery; and an exploratory laparotomy ruled out suspected mesenteric ischemia. The patient presented a torpid course on subsequent days, with abdominal distension and digestive intolerance. Therefore, the abdominal CT scan was repeated, together with a chest CT scan, which revealed lack of filling in the descending thoracic aorta consistent with intra-aortic thrombus (Figure 1 a and b) and also lack of filling in the colic artery. The aortic MRI ruled out aortic wall abnormalities, and anticoagulant therapy with continuous heparin infusion was started. Lab tests were negative for thrombophilia. Due to poor clinical course of the abdomen, i.v. anticoagulation was temporarily discontinued. Another laparotomy revealed intestinal ischemia, and therefore an enterectomy and right hemicolectomy with discharge ileostomy was performed. A week after the abdominal surgery, and again under i.v. heparin therapy, the patient presented acute arterial ischemia of the left lower limb. The arteriography revealed an acute thrombotic occlusion in the distal portion of the common femoral artery, leading to thromboaspiration and balloon angioplasty. Administration of i.v. heparin was restarted. After 45 days of heparin treatment, a follow-up chest CT scan with contrast revealed that the intra-aortic thrombus was no longer present. The patient was discharged on aspirin 100 mg/day, SC enoxaparin 60 mg/12 hours, and cilostazol 100 mg/day. At 6-month follow-up, the patient has not presented further embolic episodes under oral anticoagulant treatment.

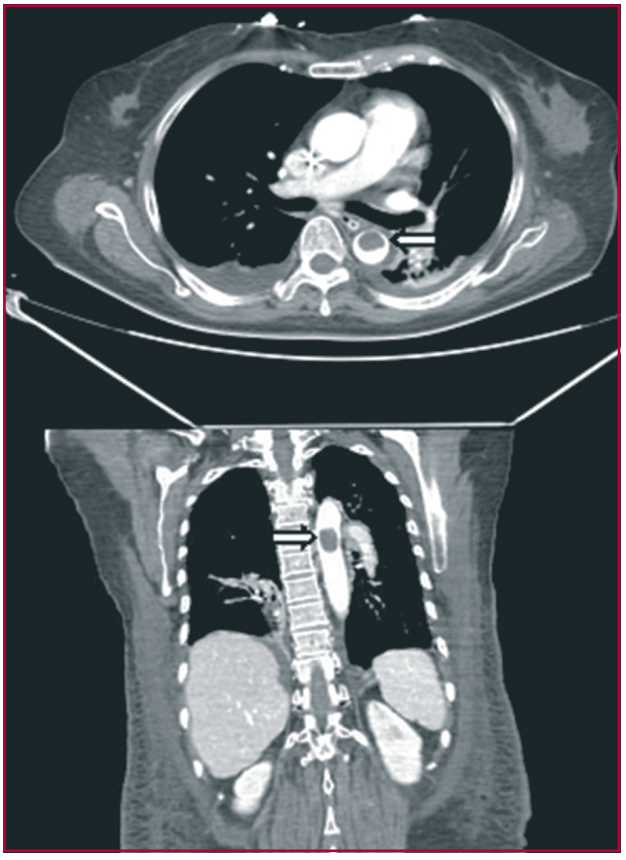

Fig. 1. Chest and abdominal CT scan. The arrows indicate thrombus

Case 2

We report the case of a 46-year-old patient, with no relevant medical history, who was admitted due to dizziness and vomiting. In view of suspected posterior fossa stroke, a MRI was performed, evidencing focal cerebellar ischemia. Physical examination showed cold left upper limb, and a Doppler ultrasound revealed acute humeral artery occlusion, requiring left upper limb bypass. Given the two ischemic foci and the suspected cardioembolism, a transesophageal electrocardiography was performed, showing preserved diameters and LV function, and moderate concentric LV hypertrophy. A highly mobile image consistent with thrombus, 1.8 cm long and 0.9 cm diameter, was detected in the descending aortic artery. An MRI was performed to assess aortic anatomy, showing an iso-hypointense lesion in T1 and T2 sequences, distal to the emergence of the left subclavian artery and in close contact with the external lateral wall, with a maximum diameter of 12 mm x 4 mm. The finding was interpreted as an intra-aortic thrombus (Figure 2). The patient was administered i.v. heparin since admission. The patient presented a com-

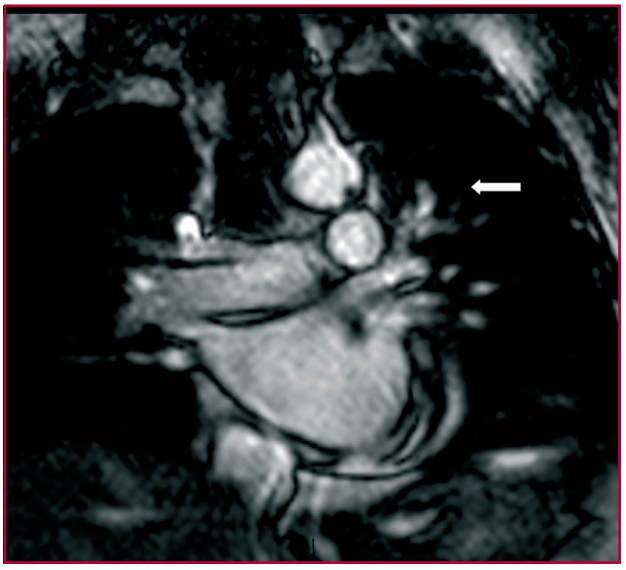

Fig. 2. Cardiac MRI. The arrow indicates thrombus

partment syndrome complication of the left upper limb that required temporary discontinuation of anticoagulant therapy and discharge incisions. The administration of i.v. heparin was later restarted and, after 4 days of clinical stability, it was replaced by enoxaparin for 14 days and then changed to acenocoumarol. Thrombophilia tests were negative. At 40-days of follow-up after discharge, the patient has not presented further embolic episodes under oral anticoagulant therapy.

Non-cardiac causes for aortic embolism include aortic thrombus associated with complicated atherosclerotic plaques, aortic aneurysms, dissection, traumatic lesions and hypercoagulability. (1) Aortic thrombosis occurring in an apparently normal aorta (non-atherosclerotic, non-aneurysmal) is a very rare cause of systemic embolism, with very few cases reported so far. (2) The etiology of thrombus formation in an apparently normal aorta is not well understood and has been associated with many disorders, such as cancer, cocaine use, hypercoagulable states, heparin-induced thrombocytopenia, aortic tumors, and even acute pancreatitis or blunt trauma. The diagnosis is best made -while evaluating cardioembolic sources- with transesophageal echography (3) in symptomatic patients, and then by MRI or CT angiography with contrast for better assessment of the aortic wall. Nowadays, CT scan seems to be the method of choice, since it also rules out the presence of thoracic or abdominal masses. (4) It is necessary to assess the existence of procoagulant states. (5) As for treatment, there is no guideline that establishes what approaches to follow, but in most cases, initial anticoagulation therapy is administered. Thrombolytics have been administered in one case reported (a treatment questioned by other authors for the potential risk of fragmentation and increased embolic risk) and, in several cases, surgical resection has been performed or –more recently– a stent graft has been used. Surgical embolectomy has been used in young patients with large mobile thrombus or multiple embolisms despite anticoagulation, and current experience with stenting is limited to very few cases reported. (6) The duration of anticoagulant therapy and the type and route of administration are not standardized.

The two patients described were initially treated with i.v. heparin and both required temporary discontinuation of anticoagulant therapy and surgery due to complications derived from embolism (mesenteric ischemia and ischemia of the left upper limb with compartment syndrome after the bypass). Once they were stable and prothrombotic factors were ruled out, subcutaneous enoxaparin followed by acenocumarol were used. In both cases, CT scan showed resolution of the thrombotic mass: in the first case, the CT scan was performed at 45 days, and in the second case, at 15 days after starting anticoagulation therapy. Both patients had positive outcomes at discharge.

Conflicts of interest

None declared.

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Yanina Castillo Costa, Flavio Delfino, Víctor Mauro, Agostina Barsotti, Francisco Saltos, Carlos Barrero
Clínica Santa Isabel - CABA
Directorio 2037 - 6.to piso
E-mail: yanu_c@hotmail.com

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Primary Undifferentiated Sarcomas of the Left Atrium and Right Ventricle

Primary malignant cardiac tumors have an incidence

of 0.0001% to 0.0003%, and most of them correspond to angiosarcomas, rhabdomyosarcomas, and undifferentiated sarcomas. (1) Diagnosis is usually late, clinical presentation is erratic, and prognosis is unfavorable. We describe two cases of primary cardiac sarcomas.

Case 1

A 58-year old hypertensive woman presented with symmetrical polyarthralgias, progressive weight loss, night sweats, and 3-month history of evening fever. Chest-abdomen-pelvis CT scan showed a 23 mm heterogeneous, hypodense image, with lobulated borders, in close contact with the roof of the left atrium. No axillary or mediastinal lymph nodes, and no pleural or pericardial effusion were observed. Transthoracic echocardiography found a 13 mm pedunculated mass in the atrial roof, at the junction with the lateral wall (Figure 1A). Cardiac magnetic resonance imaging (MRI), performed for better tissue characterization and mass localization, showed a 14.4 mm × 8.4 mm homogeneous, hypodense mass inside the left anterior appendage, a second 20 mm × 10 mm mass with irregular borders in the postero-superior region of the same atrium, and a third 22 mm × 14 mm postero-superior mass with regular borders (Figure 1B). Also, left-to-right blood flow through the atrial septum was detected, consistent with atrial septal defect (ASD). In view of suspected secondary origin, a PET-CT scan was performed. The clinical condition suddenly progressed to FC IV dyspnea, with poor respiratory function and peripheral perfusion. Transesophageal echocardiography showed an increase of one of the masses protruding through the mitral valve into the ventricle, leading to an emergency surgery. Following left atrial access, three masses were observed: one located in the left atrial appendage, another at the exit of the pulmonary veins, and the most voluminous one arising from the postero-inferior portion of the atrial septum protruding to the mitral valve. The three masses were resected, the atrium was reconstructed with a CorMatrix® patch, and the ASD was closed. The pathological diagnosis was high-grade undifferentiated sarcoma (grade 3 in the WHO classification), with morphological features suggesting adipose histogenesis. The patient remained under joint follow-up with the oncology service, who decided to start chemotherapy with doxorubicin despite her poor prognosis. Ten months after surgery the patient is still alive.

Case 2

A 65-year-old woman with history of dyslipidemia and smoking, presented with 1-week history of progressive FC III-IV dyspnea and chronic stable angina with moderate exertion. Physical examination, ECG, and lab tests provided no relevant data. Transthoracic echocardiography revealed normal left chambers, enlarged right chambers and apical thrombus in the right ventricle (RV), with estimated pressure of 100 mmHg in the pulmonary artery. Helical CT scan showed a hypodense image consistent with thrombus in the pulmonary artery extending to the RV, with >70% occlusion (Figure 3A). Thrombolytic therapy (tPA) was started, without improvement. Transesophageal echocardiography found a heterogeneous mass in the RV outflow tract with passage to the pulmonary artery. Due to presumptive diagnosis of RV tumor, right ventriculotomy and mass resection were performed, with good postoperative course (Figure 3B). Pathology revealed a vimentin (+), desmin, ac-

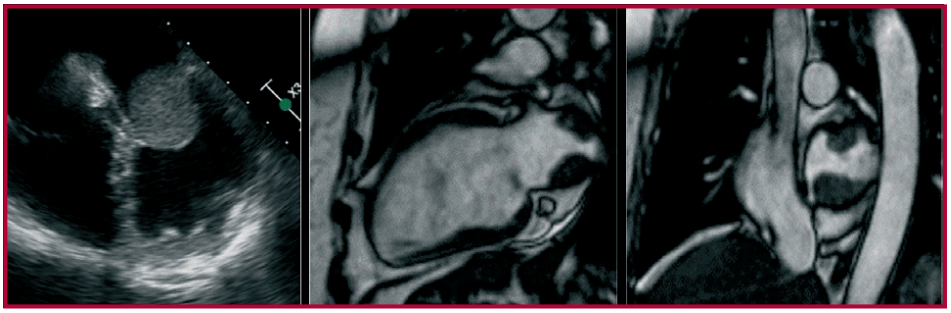

Fig. 1. A. Echocardiography showing a mass in the left atrium. **B** and **C.** Two magnetic resonance images showing two masses in the left atrium. (Case 1)

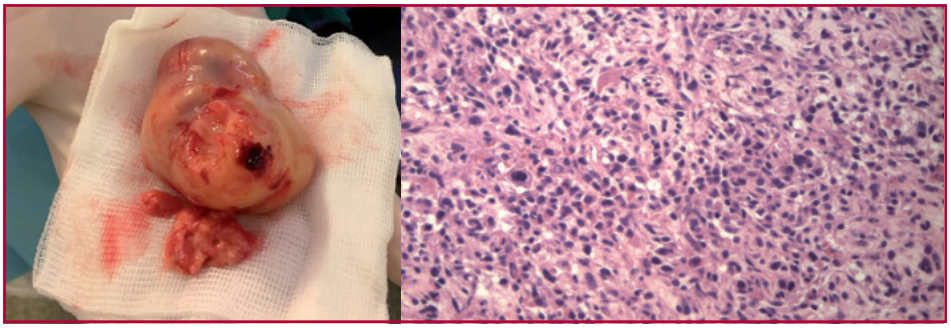

Fig. 2. Macroscopic image of the tumors and undifferentiated sarcoma histology. Morphological features can be observed suggestive of adipose histogenesis. (Case 1)

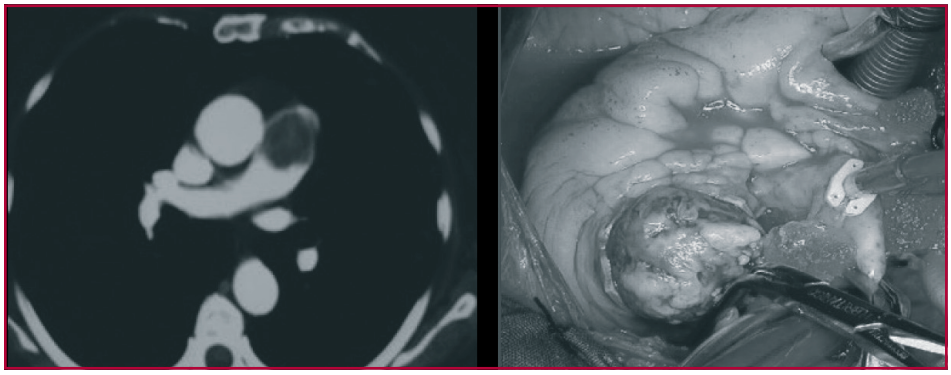

Fig. 3. A. Computed tomography showing right ventricular sarcoma invading the left pulmonary artery. **B.** Intraoperative image showing the sarcoma implanted in the right ventricle at the time of removal. (Case 2)

tin, factor VIII, CD31 and CD34 (-) undifferentiated sarcoma. The patient died eight months after surgery.

Primary malignant tumors of the heart occur between the fourth and fifth decades of life regardless of sex, and can be distributed in any heart chamber. Depending on the cell line, 37% of tumors can be angiosarcomas, 24% malignant fibrous histiocytomas, 9% leiomyosarcomas, 7% rhabdomyosarcomas, or 7% poorly differentiated sarcomas. (2) Some studies propose certain preference of some types of tumors for a specific location; thus, poorly differentiated sarcomas and leiomyosarcomas are more frequently found in the left chambers (66% and 50%, respectively), as opposed to angiosarcomas, which are typically located in the right atrium. (3) Their clinical presentation is erratic, and usually produce symptoms due to the effect of mass growth, causing obstruction to blood flow and drop in cardiac output. Patients may also present with dyspnea, chest pain, or symptoms resembling other cardiovascular conditions. Constitutional symptoms such as fever, weight loss, or night sweats are also common. (4)

Echocardiography is the most common study to detect these tumors and differential characteristics with benign tumors have been described, such as their origin outside the atrial septum, their extension to the pulmonary veins, the occurrence of multiple masses, their broad fixation to the atrial wall, and their semisolid consistency. As in our case, these data are suggestive of a diagnosis different from myxoma, and lead to further studies. Cardiac CT scan and MRI accurately show mass location, extension, and characteristics, making them very useful tools not only for disease characterization, but also for surgery planning. (4) Surgical treatment is mandatory and its purpose is the complete resection of the tumor, although the result is not curative given the high rate of local recurrence or distant metastases these tumors have.

In recent years, chemotherapy has been used as adjuvant therapy even in patients with local recurrence. (5-6) As its effectiveness is unknown, there is no accurate information on the optimal scheme. Doxorubicin is used in view of its benefit in soft tissue sarcomas in other locations; however, in some series it does not appear to prolong survival. (5) The histological lineage does not correlate with prognosis in the different series, but FC III-IV, high degree malignant histology, more than 50% tumor necrosis, presence of distant metastases at the time of diagnosis, and the amount of mitosis observed in more than 10 cells per 10 high-power fields are associated with poor survival. (2) Despite treatment, the prognosis for these tumors remains unfavorable.

Conflicts of interest

None declared.

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Osvaldo Tenorio Núñez[1, 2], Michel David[1, 2], José Gabriel Gargiulo[3], Esteban Ludueña Clos[3], Héctor Alejandro Machain[2]

[1] Department of Cardiovascular Surgery, Hospital Alemán.
[2] Hospital Británico.
[3] Department of Cardiology, Hospital Británico, Buenos Aires.
Av. Pueyrredón 1640 -C1118AAT-Buenos Aires;
email: omtenorio@hotmail.com

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