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Vascular Diseases Quiz – Case 36

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A 53-year-old female presented to the Emergency Department with a pale and cold right lower extremity. The symptoms appeared one hour ago and the patient transferred immediately to the nearest hospital. Triage referred her directly to the Vascular Department Unit after a short clinical exam, as well as a fall blood and biochemistry check.

The patient presented in distress, slightly tachycardic, but hemodynamically stable. Prior to this incident, the patient was well and fit. Past medical history included hypothyroidism, osteoporosis and total hysterectomy performed 15 years ago for symptomatic uterine fibroids.

On physical examination, the right extremity was cold and pale, without palpable pulse at both the popliteal and the pedal arteries.

Emergency vascular ultrasound and angiography were performed and the patient underwent a surgical embolectomy. A small amount of occlusive material was recovered and was sent for histopathologic analysis.

What's the diagnosis?

Comment

A wide variety of cardiac pathology potentially leads to acute limb ischemia (ALI), with atrial fibrillation being the most common cause. Other causes include myocardial infarction, left ventricular aneurysms, non-bacterial thrombotic endocarditis and infective endocarditis, aortic mural thrombi and atheroembolism.

In the present case, the patient underwent rigorous investigation for potential embolic sources during the days hospitalized. Computed tomography (CT) scan of abdomen revealed infarction of the spleen and the right kidney. CT of the chest and brain did not show any evidence of embolisation.

Cardiac ultrasound scan revealed a mass in the left atrium, with normal cardiac mobility. Findings were consistent with a cardiac tumor, probably a myxoma. The patient was referred to a cardiac surgeon for further evaluation and possible tumor resection by open-heart surgery.

Cardiac myxoma is rare in the general population with an incidence of approximately 0.0017%, but it is the most frequent benign tumor of the heart, representing 50–83% of all primary cardiac tumors. Histologically, these are real tumors, derived from multipotent mesenchymal cells of the subendocardium. Most of sporadic myxomas are solitary, affect females with a prevalence of 3:1, and patients usually are in their fifth decade of life.[4–6] On the contrary, familiar myxomas mainly affect young males. In 75–85% of cases, myxomas are located in the left atrium. Despite its benign nature, myxoma complications may lead to catastrophic results, making this entity an important diagnostic and therapeutic challenge. Patients typically present with one or more of the classic triad of signs such as cardiac obstruction (67%), systemic symptoms (34%) and embolic signs (29%).[7] Cardiac obstruction may cause dyspnea, cough, hemoptysis, pulmonary edema, congestive heart failure, syncope or cardiac arrest, while constitutional symptoms such as fever, weight loss, and immunologically induced myalgia, arthralgia, muscle weakness and Raynaud syndrome are the most common systematic symptoms. Emboli usually migrate to the central nervous system, but the whole arterial bed, from coronary to lower extremities arteries, may be affected. Embolisation of multiple organs, as in this case, is less frequent and it is associated with a tumor surface that is either irregular or covered in friable villi.

As physical examination may be misleading, diagnosis is often reached with cardiac ultrasound that constitutes the diagnostic method of choice. Surgical resection is usually curative and aims to relieve symptoms and avoid complications.

In the absence of a profound diagnosis in a patient with ALI, thorough medical history and physical examination should not be neglected, as atypical symptoms and signs may imply other sites of embolization as well. Once ascertained, multiple emboli should raise suspicion for a cardiac cause and physicians should urgently proceed to prompt evaluation. Given the rarity but also the significant mortality of cardiac tumors, even in the absence of arrhythmias, valvular or coronary heart disease, in patients without previously known significant risk factors, evaluation with a heart ultrasound is warranted and should never be omitted.

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