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Case report

A Classic Mimicker of Systemic Vasculitis☆

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ABSTRACT

Background and objective: Embolic and constitutional manifestations of intracavitary cardiac tumors are included within the classic mimickers of systemic vasculitis, especially in those in which there are no cardiac manifestations. We present a case report of atrial myxoma in which the patient only presented systemic symptoms and in whom an initial diagnostic approach of systemic vasculitis was made. We also performed a literature search of the cases described.
Patient and method: A case report of atrial myxoma with atypical presentation manifested as a systemic disease with no concomitant cardiac symptoms is described. The case report is discussed and 11 cases of atrial myxoma pseudovasculitis described in the literature are reviewed, emphasizing their similarities and differences.
Discussion: Constitutional symptoms and cutaneous manifestations were the most common. Most of the cases showed partial response to glucococorticosteroid treatment, reinforcing the theory of the inflammatory role in its pathogenesis. Mean delayed time to diagnosis was 12.27 months.
Conclusion: Atrial myxoma is a systemic vasculitis mimicker, this being difficult to diagnose in the absence of cardiac manifestations. This delay in diagnosis entails serious complications.

Un simulador clásico de vasculitis sistémica

Palabras clave:
Mixoma
Tumor cardíaco
Vasculitis sistémica
Seudovasculitis
Simulador

RESUMEN

Fundamento y objetivo: Las manifestaciones embolígenas y constitucionales de los tumores cardíacos intracavitarios se engloban dentro de los mimetizadores clásicos de las vasculitis sistémicas, sobre todo en aquellas ocasiones donde no se presentan manifestaciones cardiológicas. Se describe un caso de mixoma auricular con clínica exclusivamente sistémica, cuya orientación diagnóstica inicial fue de vasculitis. Se revisan los casos descritos en la literatura.
Paciente y método: Se describe un caso de mixoma auricular con presentación en forma de manifestaciones sistémicas sin sintomatología cardiológica acompañante. Se expone el caso clínico y se compara con 11 casos de seudovasculitis por mixoma auricular descritos en la literatura, haciendo énfasis en las similitudes y divergencias.

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Discusión: Los síntomas constitucionales junto con las manifestaciones cutáneas fueron los más frecuentes. La mayoría de los casos presentaban respuesta parcial al tratamiento glucocorticoideo, reforzando la teoría del componente inflamatorio en su patogenia. La demora media en el diagnóstico fue de 12,27 meses.

Conclusión: El mixoma auricular es un simulador de vasculitis sistémica y es de difícil diagnóstico cuando no presenta manifestaciones cardíacas. La demora diagnóstica puede conllevar complicaciones graves.

Introduction

The systemic manifestations of intracavitary cardiac tumors—fever, weight loss, joint and muscle pain, and Raynaud's phenomenon—lead to this entity's being mistaken for immunological, neoplastic or infectious diseases, especially in those cases in which there are no cardiac manifestations.[1] There is a large group of classical mimickers of systemic vasculitis, including atrial myxoma, atherosclerosis, systemic AL amyloidosis, bacterial endocarditis, antiphospholipid syndrome and thromboangiitis obliterans.[2] This masking phenomenon is well known, although there have been few reports of cases in which the cause was atrial myxoma. We present a case of atrial myxoma with embolic and systemic manifestations, but no symptoms or signs of cardiac involvement, in which the initial findings pointed to a diagnosis of systemic vasculitis. We report the details of a search for the cases described in the literature, analyzing similarities and differences.

Clinical Observation

The patient was a 60-year-old man with a history of hypertension, which is being treated with an angiotensin receptor blocker, and dyslipidemia, which is being treated with a statin. He presented with low back pain of acute onset, as well as acrocyanosis of the right hand associated with ischemic pain in the fingers of that hand. Physical examination revealed acrocyanosis of the right hand; the peripheral pulses were present and symmetric. The heart and lung sounds were normal. He underwent chest radiography, electrocardiography and computed tomographic angiography of thorax and abdomen, all of which were normal. Laboratory tests, including complete blood count, kidney and liver function, coagulation tests, antiphospholipid antibodies, cryoglobulins and serological tests for hepatitis B and C viruses, parvovirus and antineutrophil cytoplasmic antibodies (ANCA) were normal. Antinuclear antibodies were detected at low titers (1/80) in a homogeneous pattern, with negative anti-double-stranded DNA antibodies and no complement consumption. Nailfold capillaroscopy revealed signs of angiogenesis with internal bleeding in the fingers of the right hand. Treatment consisted of oral antiplatelet therapy and intravenous prostaglandins, which achieved the complete resolution of the symptoms. Three months later, the patient developed bilateral Raynaud's phenomenon. One month after that, he presented with acute, stabbing, epigastric pain, as well as ischemic pain in the 4th toe on his left foot, hypesthesia in the 2nd–4th toes on the same foot and pain in the plantar fascia upon stepping with that foot. He reported proximal muscle weakness in lower extremities and undetermined weight loss over the preceding 2–3 months. Physical examination revealed that his left foot was colder than his right foot, a mild erythematous lesion in left plantar fascia, bilateral absence of posterior tibial and pedal pulses, and palpable popliteal pulses. The first heart sound had a greater intensity and there were no extra sounds.

The results of the laboratory tests included a hemoglobin level of 10.4 g/dL, hematocrit 34%, mean corpuscular volume 78 fL, creatine kinase 602 U/L, creatine kinase-mb 26 U/L, platelets 506,000/mm^3, γ-glutamyl transpeptidase 216 U/L, alkaline phosphatase 200 U/L, C-reactive protein 128.1 mg/L and erythrocyte sedimentation rate 88 mm/h. The remaining laboratory findings were normal. As we suspected medium to small vessel systemic vasculitis, the patient was admitted to the hospital to complete the study.

An electromyogram revealed mononeuritis of left posterior tibial nerve with no evidence of myopathy. To rule out arteriographic abnormalities, contrast-enhanced computed tomographic angiography of thorax and abdomen was scheduled. The patient received glucocorticoids (60 mg of methylprednisolone in 2 doses) to prevent a probable allergic reaction to the contrast material, which resulted in a notable improvement in his symptoms. The imaging study revealed an intracavitary filling defect at the level of left atrium. This finding led to the performance of transthoracic

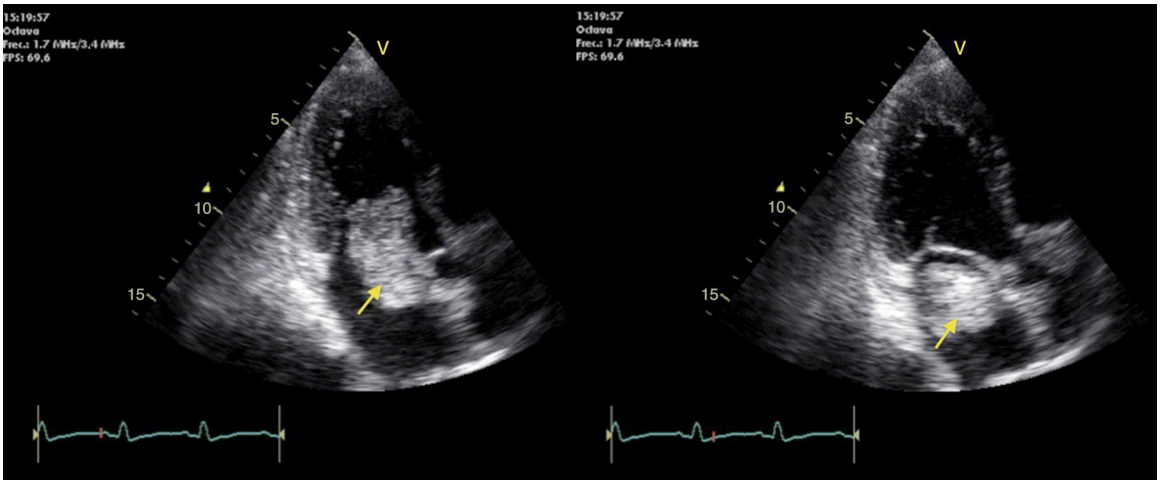

Fig. 1. Transthoracic echocardiogram with an apical view of 3 chambers showing a tumor in left atrium, in diastole (left) and in systole (right).