

Vision loss, multiple cerebral infarction, ischemia of extremities: Systemic vasculitis or cardiac myxoma?

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Received June 12, 2020 accepted August 5, 2020

Introduction

Systemic vasculitis may involve multiple organs or systems including eye, ear, nose, kidney, lung and nervous system. Due to lack of specific biomarkers, the diagnosis of primary systemic vasculitis generally should exclude some mimics, such as neoplasm and infections. In this case, we report one patient whose final diagnosis was cardiac myxoma but with mimicked clinical presentations of vasculitis.

Case Report

A 42-year-old woman was admitted to our hospital because of decreased vision of left eye and delirium for 1 year, and erythema at limbs for 6 months. One year before admission, her left eye visual acuity decreased abruptly. Fundus examination showed retinal vasculitis. Meanwhile, she had attacks of dizziness and loss of consciousness once every 1–2 months without any precipitating cause. Magnetic resonance imaging (MRI) of her head revealed multiple ischemia and infarctions involving bilateral pontine, cerebellum, infratemporal parieto-occipital cortex, and bilateral paraventricular area. Aspirin was given but there was no improvement. Six months ago, painful erythema and rash appeared repeatedly in her fingers and toes, accompanied by cyanosis. She had no fever, oral ulcer, swollen joints. Her blood routine, biochemical tests, blood lipid

profile, coagulation function, and urine routine test were all in normal range. Her blood tests for rheumatoid factor (RF), antinuclear antibody (ANA), antineutrophil cytoplasmic antibody (ANCA), antiphospholipid antibodies (APLs), cancer biomarkers, hepatitis B and human immunodeficiency virus (HIV) infections were negative. Erythrocyte sedimentation rate (ESR) was 35 mm/h, and C-reactive protein (CRP) was 12 mg/L. She was diagnosed as having systemic vasculitis. Prednisone 60 mg/day and cyclophosphamide (CYC) 50 mg/day were given orally. Then, her steroid was gradually tapered to 20 mg/day and CYC was continued. During this course, her fingers were still blue and purple (Figure 1), and dizziness and vertigo were still intermittently occurring as before. Therefore, head MRI was repeated and new local ischemia and infarction were found, but no abnormality could be found in her brain MRA. So, she was admitted to our hospital to clarify the diagnosis.

During the physical examination at admission, several painful erythema were found on her legs and arms. Ischemia of fingers and toes were discovered. Lung and heart examination were normal. No heart murmur was heard during auscultation.

After admission, her white blood cell count was $13.4 \times 10^9/L$, neutrophil count was $11.9 \times 10^9/L$, hemoglobin and platelet count were normal. Her urine test, liver and kidney function tests were normal. Her ESR was 18 mm/h and CRP was

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5.6 mg/L. Blood tests for autoantibodies, including ANA, anti-extractable nuclear antigen (ENA) antibody, ANCA, and APLs, were all negative. Echocardiography showed left atrium enlargement, and a neoplasm-like mass with middle-to-high echo, uneven density, and clear boundary was found. The mass was 45 mm × 22 mm in size and swung with blood flow and fell into left ventricle during diastole. The mass was most probably an atrial myxoma (Figure 2). So, her oral steroid was tapered and CYC was discontinued. Then, she was transferred to cardiac surgery unit for operation. The pathological examination of the removed neoplasma confirmed the diagnosis of myxoma. She recovered completely after operation and was discharged.

Discussion

Cardiac myxoma (CM) is by far the most common primary benign cardiac tumor,^[1] accounting for about 50% of heart neoplasma. The peak age of disease is at the 4th–6th decades, and the majority (60%–70%) affected by the disease are female.^[2,3] The myxoma can occur in any atrium or ventricle of the heart but are most commonly found in left atrium (80%). If the mass is very small, the myxoma causes no clinical symptoms. When the tumor becomes large in size, it can cause hemodynamic changes and peripheral vascular embolism. The most common clinical symptom of left atrial myxoma is palpitation and shortness of breath caused by obstruction of atrioventricular blood flow, which is similar to rheumatic mitral valve disease. But not all patients with CM can produce murmur that could be discovered in auscultation,^[3] which makes the diagnosis more difficult. The earliest clinical manifestation of CM is peripheral arterial embolism, even cerebral infarction and stroke.^[4,5] Meanwhile, some patients may present constitutional manifestations, such as fever, anorexia, weight loss, arthralgia, and anemia. Sometimes, elevated ESR and increased serum globulin could be found. The mechanism of these symptoms may be related to the immune response to tumor and the release of vascular endothelial growth factor (VEGF), interleukin (IL)-6 and other cytokines.^[6] When small gel fragments fall off from the myxoma body, they can lead to multiple arterial embolisms. All these mimic the clinical presentations of systemic vasculitis.^[7]

In this case, a middle-aged female patient had eye, central nervous system, and skin involvement consecutively. She



Figure 1: Ischemic changes of the patient's hands



Figure 2: Echocardiography of the patient (arrow shows the mass)

was in a mild inflammatory condition but negative for autoantibodies and there was no evidence of tumor and infection. Therefore, she was diagnosed with systemic vasculitis. But the subsequent treatment with steroid and immunosuppressants did not show relief of her symptoms and signs. Repeated echocardiography revealed a mass in her left atrium, which made the diagnosis clear. Operation confirmed the final diagnosis of atrial myxoma.

The diagnosis of systemic vasculitis should be cautious, particularly when there is no pathological evidence of vasculitis. Mimics should be carefully excluded, including neoplasm and infection.

Informed Consent

Informed consents have been obtained.

Conflict of Interest

None declared.

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