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IMAGES IN RHEUMATOLOGY

Rare coexistence of large vessel vasculitis and nonbacterial thrombotic endocarditis: a case report

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INTRODUCTION

Large-vessel vasculitis (LVV) involves inflammation of the large arteries, often leading to vessel occlusion or aneurysm formation. Nonbacterial thrombotic endocarditis (NBTE) is a rare condition associated with hypercoagulable states and malignant diseases, characterized by sterile vegetations on previously undamaged cardiac valves. Here, we present a patient diagnosed with LVV, who also had echocardiographic vegetations on the mitral and aortic valves, suggestive of NBTE.

CASE REPORT

A 61-year-old woman presented to the emergency department with a one-week history of episodes of syncope preceded by weakness, sweating, and dizziness upon standing. Over the past three months, she reported fatigue on minimal exertion, unintentional weight loss (6 kg; 8% of body weight), intermittent claudication of the lower limbs and tinnitus in the frontal region. She denied fever, headache, visual symptoms, jaw claudication, cutaneous manifestations such as digital ischemia or purpura, and arthralgia. Her background history included arterial hypertension and an ischemic stroke in the prior three months with minor sequelae (left hemiparesis). On physical examination, temperature was 38°C, blood pressure 167/67 mmHg on the right and 188/67 mmHg on the left arm, and cardiac auscultation revealed rhythmic S1 and S2 sounds with a diastolic murmur at the aortic focus (grade III/VI) and a left basal systolic murmur (grade III/VI) radiating to the axilla. Laboratory tests showed microcytic/hypochromic anemia (Hb 12.9 g/dL), minor leukocytosis (11.60x10^9/L), elevated ESR (84 mm/h) and CRP (16.6 mg/dL), and cholestasis (GGT 236 U/L, alkaline phosphatase 347 U/L) with normal bilirubin levels (0.70 mg/dL), likely driven by the systemic inflammatory response, as imaging ruled out hepatic vascular thrombosis and Budd-Chiari syndrome. Anti-nuclear and anti-neutrophil cytoplasmic antibodies were negative. Blood cultures, including for HACEK bacteria, and serologic tests for HIV, hepatitis B, hepatitis C, and syphilis were negative. Coagulation studies revealed an elevated fibrinogen level (785 mg/dL), an INR of 1.31, and an APTT ratio of 0.99, while antiphospholipid an-

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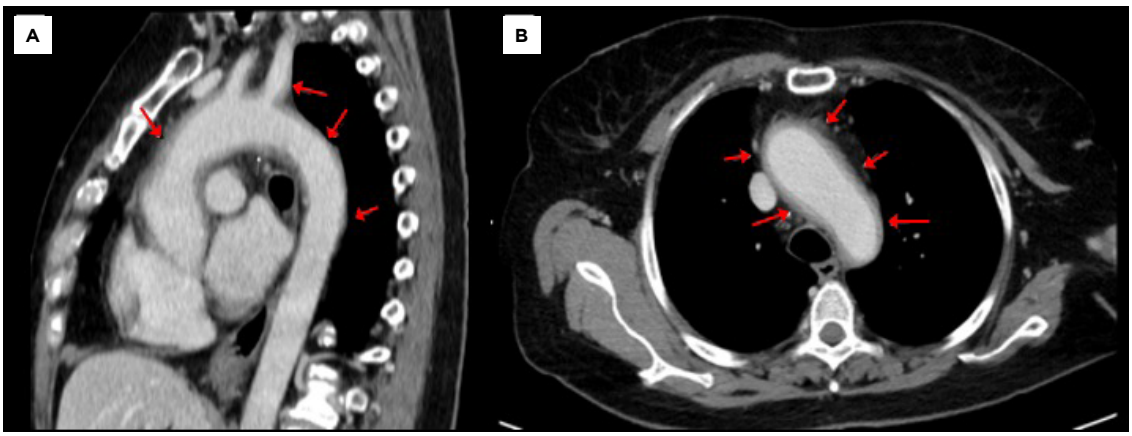

Figure 1. Thoracic contrast-enhanced CT images in axial (A) and sagittal (B) planes show diffuse circumferential wall thickening (arrows) of the ascendent thoracic aorta, aortic arch, proximal descending thoracic aorta and supra-aortic vessels, compatible with vasculitis.

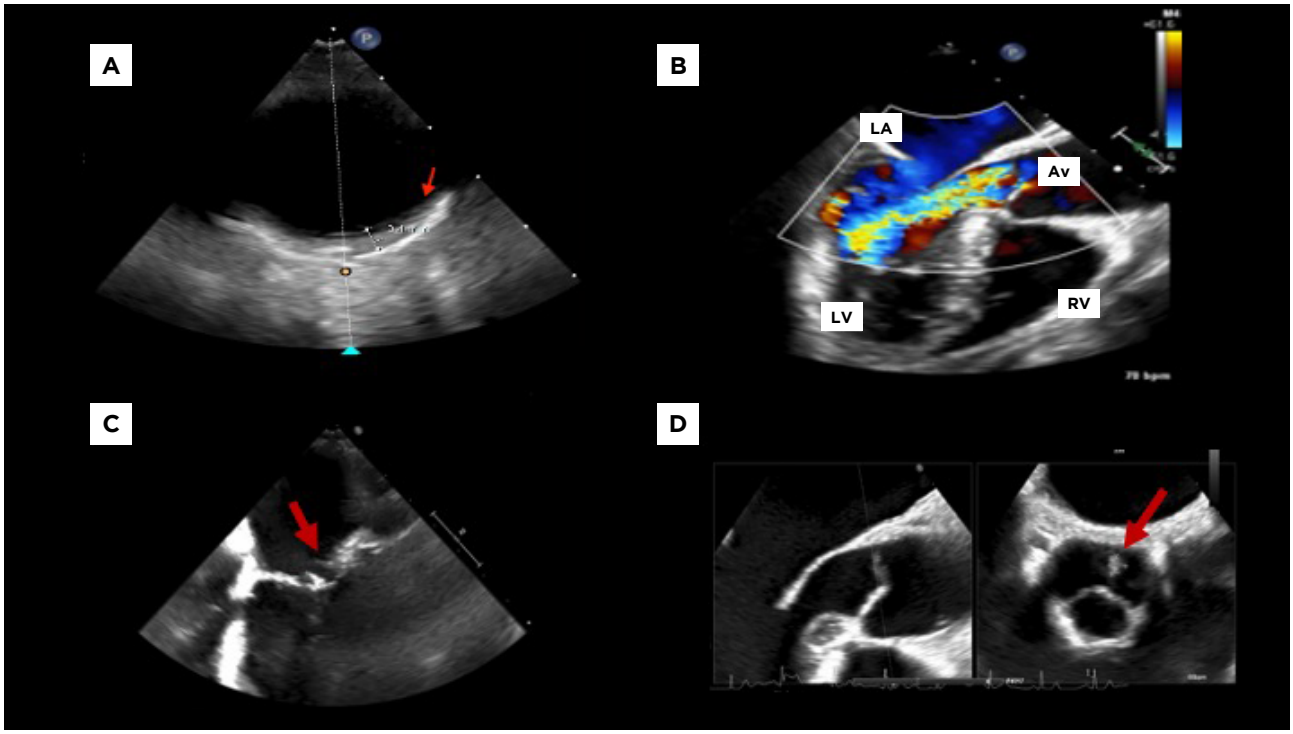

Figure 2. **A)** Transoesophageal echocardiogram images showing thickening of the aortic wall (arrow), suggesting large-vessel vasculitis. **B)** Transoesophageal echocardiogram showing severe aortic regurgitation. **C) and D)** Transoesophageal echocardiographic views of the vegetation at the level of the aortic valve (arrows). RV (right ventricle); LA (left atrium); LV (left ventricle); Av (aortic valve).

tibodies were negative. Ultrasound of the temporal and axillary arteries showed no halo sign. Thoracic-abdominal-pelvic CT revealed wall-thickening of the ascending aorta, aortic arch, proximal descending thoracic aorta, carotid and vertebral arteries, indicative of vasculitis, with no space-occupying lesions suggestive of underlying malignancy. Transthoracic and transoesophageal echocardiogram revealed multiple hypoechoic masses attached to the aortic and mitral valves, with severe aortic and minimal mitral regurgitation. The diagnosis of LVV with NBTE was established. The patient was started on anticoagulation therapy with enoxaparin 80 mg/day and methylprednisolone pulses (1g/day for three days), followed by 60 mg/day of oral prednisolone (with gradual tapering) and methotrexate 12.5 mg/weekly. Her symptoms resolved rapidly, and after five months of follow-up, the echocardiogram showed mild aortic regurgitation and no evidence of valve vegetation. Treatment with tocilizumab 162 mg/weekly was initiated after nine months due to glucocorticoid-related adverse events. Anticoagulation therapy has been maintained to date. Presently, at two years of follow-up, the patient is in clinical remission, experiencing only occasional mild asthenia and a reduction in lower limb claudication symptoms, managed with prednisolone 5mg/day, methotrexate and tocilizumab.

DISCUSSION

NBTE has been linked with various immune-mediated conditions, such as systemic lupus erythematosus, but its association with LVV is exceedingly rare. Although the precise mechanisms linking LVV with NBTE require further investigation, it is believed that the systemic inflammation characteristic of LVV may contribute to a hypercoagulable state, with chronic endothelial activation, elevated inflammatory cytokines, and vascular injury promoting thrombus formation on cardiac valves.[1] To the best of our knowledge, only four cases involving these concurrent conditions have been reported to date, all in patients with giant cell arteritis and older than our patient.[2-5] Additionally, this report marks the first case of NBTE with extensive large vessel vasculitic involvement documented through imaging. Our findings highlight the need for heightened awareness and personalized management strategies in these complex clinical scenarios.

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