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Takayasu Associated Myocarditis in a Young Female: A Case Report

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ABSTRACT

Takayasu arteritis (TA) is a rare granulomatous panarthritis, characterized by stenosis or obliteration of large and medium-sized arteries, such as the aorta. The onset of symptoms tends to lead to a delay in diagnosis that can range from months to years, during which time vascular disease may initiate and progress to become symptomatic. In this context, TA may present with various uncommon cardiovascular complications such as dilated cardiomyopathy, valvulitis, and myocarditis. Herein, we report on a 29-year-old female who initially presented with active myocarditis and was later incidentally diagnosed with TA.

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Introduction

Takayasu arteritis (TA) is an infrequent granulomatous vasculitis affecting large vessels, primarily the aorta and its principal branches (1). Moreover, TA has a higher incidence among young Asian females and is known to manifest various signs and symptoms such as chest pain, dyspnea, limb weakness, and vision loss (2). The presentation and severity of TA vary based on the involved artery and the degree of inflammation (2, 3). Additionally, TA can lead to life-threatening conditions, including heart failure, myocardial infarction, myocarditis, and aneurysm (4, 5). Given these diverse presentations, the timely diagnosis of TA can be challenging. Early diagnosis and management of TA are crucial to prevent such complications (6). Herein, we describe a case of TA in a young Asian female who presented with myocarditis.

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Case Presentation

A 29-year-old female patient visited our hospital with complaints of dyspnea (NYHA Class III), paroxysmal nocturnal dyspnea (PND), orthopnea, loss of appetite, fatigue, and weakness. Two weeks prior, she experienced chest pain and dyspnea (NYHA Class II). She had no significant medical history. Upon admission, her vital signs were as follows: right-hand blood pressure 110/70 mmHg, pulse rate 110 beats per minute, respiratory rate 18 breaths per minute, and temperature 37°C. Physical examination revealed a systolic murmur (3/6) at the left sternal border, clear lung auscultation, and +2 edema in the lower extremities. Additionally, she exhibited asymmetric pulses in her upper limbs (left radial pulse was not detectable). There were no signs of volume overload. The obtained electrocardiogram (ECG) is shown in Figure 1.

Laboratory tests yielded the following results: HIV Antibody: Negative, Hepatitis B Surface Antigen (HBS Ag): Negative, Hepatitis C Antibody: Negative, Rheumatoid Factor (RF): Negative, Thyroid Function Tests (TFT): Within Normal Limits (T3 = 0.76, T4 = 6.65, TSH = 2.5), Sodium: 136 mmol/L, Potassium: 3.3 mmol/L, Blood Urea Nitrogen (BUN): 12 mg/dL, Creatinine: 1.1 mg/dL, Calcium: 7.9 mg/dL, Phosphorus: 2.9 mg/dL, Magnesium: 2 mg/dL, C-reactive Protein (CRP): 28.49 mg/L, Erythrocyte Sedimentation Rate (ESR): 68 mm/h, Urinalysis (Proteinuria: Trace, White Blood Cells: 2-3/HPF, Red Blood Cells: 6-8/HPF, Few Bacteria), D Dimer: 44% (Negative), White Blood Cell Count: 8.9 x 10^3/µL, Hemoglobin: 10.7 g/dL, Platelet Count: 331,000/µL, Pro B-type Natriuretic Peptide (Pro BNP): 2023 pg/mL, Troponin I: 29.1 ng/mL (Positive), Reumatogist tests(Anti dsDNA,Anti ccp,ANA,Lupus Anticoagulant,B2, Glycoprotein, PANCA,CANCA):Negative, COVID-19 Polymerase Chain Reaction (PCR): Negative, Abdominopelvic Ultrasound: Normal.

Echocardiography indicated mild left ventricular enlargement, severe systolic dysfunction (ejection fraction [EF]: 35%), and global hypokinesia predominantly involving the apical segment. Other findings included severe left atrial enlargement, mild mitral valve leaflet systolic motion abnormalities, moderate functional mitral regurgitation (peak E velocity = 0.6 m/s), small circumferential pericardial effusion, and brief right atrial invagination. No significant hemodynamic effects were observed. Furthermore, there was significant internal thickening (4 mm) circumferentially in the thoracic descending aorta. A cardiac magnetic resonance imaging (CMR) was conducted and revealed normal left ventricular size, severely reduced systolic function (EF: 29%), mildly reduced right ventricular systolic function (EF: 29%), mild aortic insufficiency, mild tricuspid regurgitation, moderate mitral regurgitation, evidence of myocardial edema in the mid-to-apical left ventricular segments, elevated T2 value (54 ms), elevated T1 value (1091 ms), and moderate pericardial effusion (Figure2).

During hospitalization, the patient received treatment including eplerenone, sacubitril/valsartan, furosemide, empagliflozin (an SGLT2 inhibitor), vitamin B1, and folic acid.

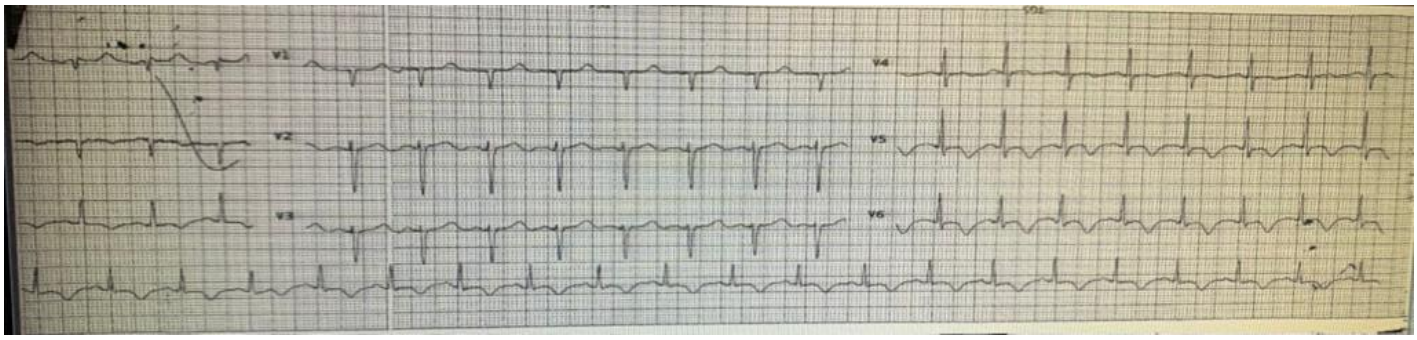
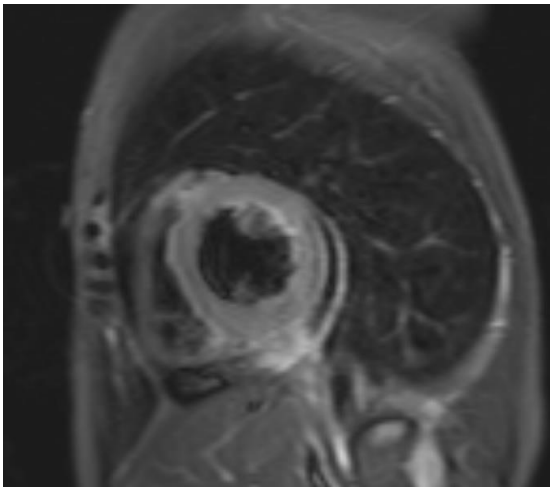
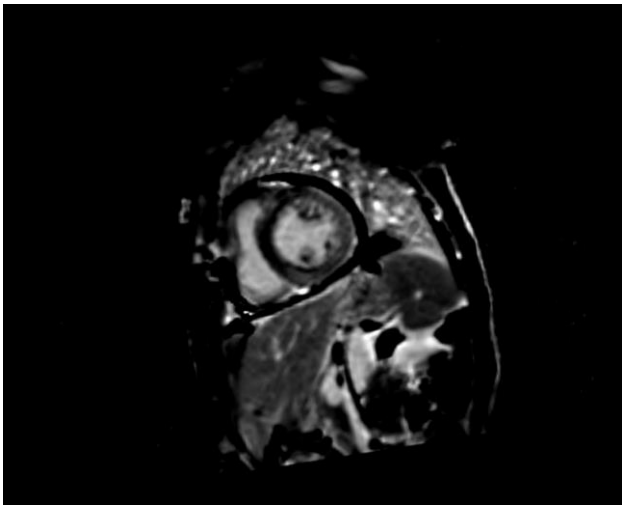


Figure 1. The patient ECG, normal sinus rhythm, standard axis deviation.

A contrast-enhanced computed tomography angiography (CTA) of the abdominal and thoracic aorta revealed circumferential wall thickening of the thoracic and abdominal aorta consistent with Takayasu disease (Figure 3). artery was found to be cut after the left internal mammary artery (LIMA) graft. Bilateral common iliac arteries showed mild stenosis, while the right vertebral artery showed moderate stenosis at its origin, and the left vertebral artery exhibited severe stenosis at its origin.



(A)



(B)

Figure 2. A) Cardiac Magnetic Resonance (CMR) STIR sequence short axis view shows high signal intensity in the mid postero lateral segment of LV wall .The pericardium shows high signal with pericardial effusion.B) Short axis Late Gadolinium Enhancement sequence demonstrates subepicardial hyperenhancement of posterolateral wall of LV and also pericardium.

Furthermore, circumferential wall thickening was noted in the supra-aortic branches. The right subclavian artery exhibited long-segment stenosis, and the left subclavian Circumferential wall thickening was also evident in the descending abdominal aorta, leading to mid-abdominal aorta stenosis, with the origin of the celiac artery, right renal artery, and aorta appearing normal.

The left renal artery was occluded, resulting in reduced enhancement of the left kidney. Following consultation with a rheumatology specialist, the patient was treated with a three-day course of methylprednisolone pulses, mycophenolate mofetil (CellCept), and osteophytes. Subsequent echocardiographic assessments during the follow-up period indicated normalization of left ventricular size and improvement in systolic function (EF: 50%), as well as the persistence of severe left atrial enlargement.

Discussion

TA is an idiopathic chronic vasculitis that predominantly affects young Asian females under the age of 40 (7). Although the precise etiology of TA remains unknown, the infiltration of inflammatory agents in an autoimmune process is understood to contribute to its pathogenesis (8). Furthermore, there are no specific diagnostic laboratory tests for TA. Therefore, the diagnosis is established through high clinical suspicion and imaging modalities such as Doppler ultrasonography and cardiovascular magnetic resonance (6, 8). In the current case report, we present a case of myocarditis in an individual with previously undiagnosed TA. Several key points warrant attention in this patient's presentation.

In our patient, the conducted CTA revealed circumferential wall thickening of the thoracic and abdominal aorta. As previously mentioned, the diagnosis of TA can be particularly challenging, and a timely diagnosis is essential to prevent further complications (9). Elevated erythrocyte sedimentation rate (ESR) and the absence of palpable pulses heightened clinical suspicion for TA. Moreover, the administration of corticosteroids and immunosuppressive agents, such as mycophenolate mofetil, led to

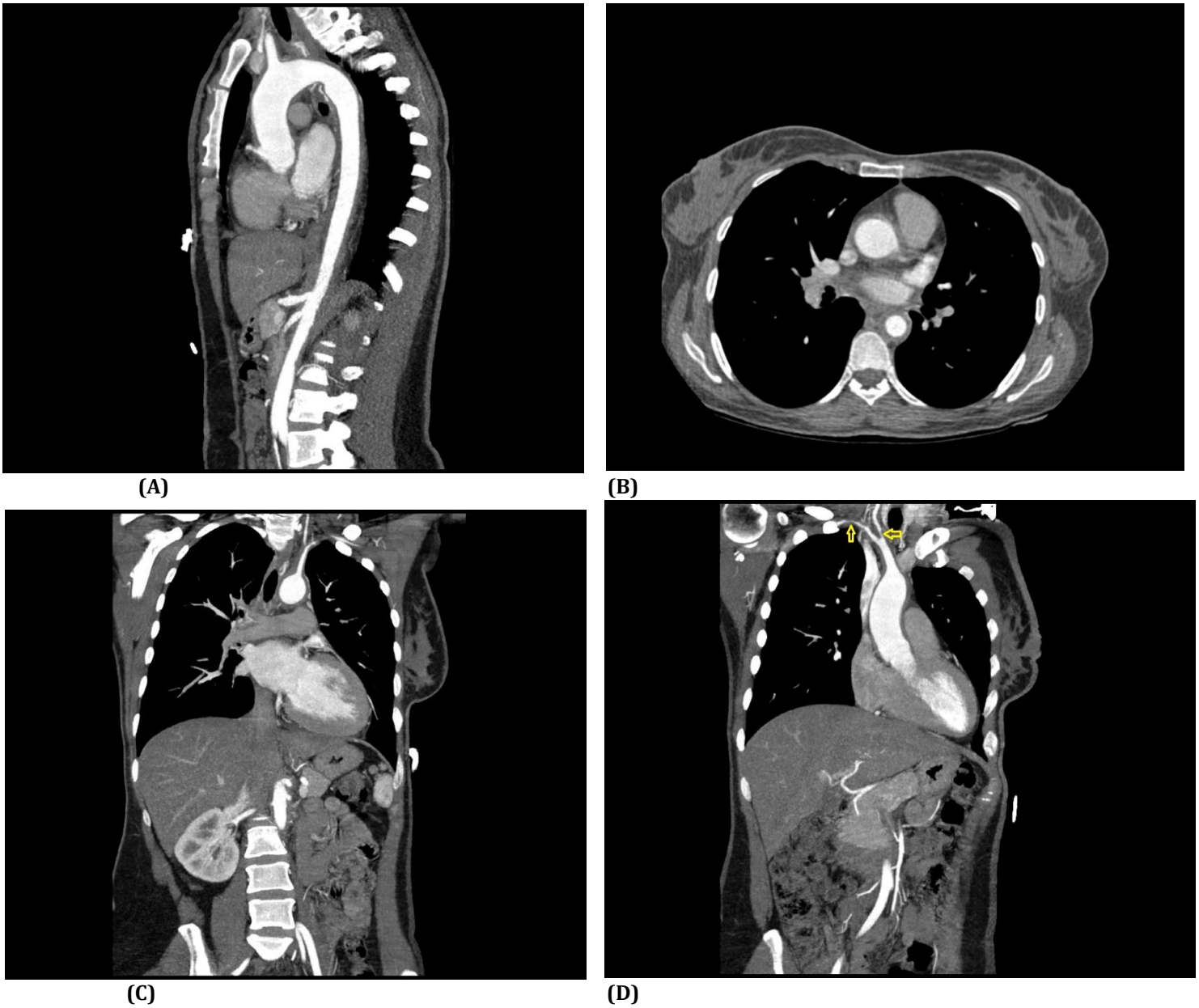


Figure 3. A, B) Multidetector cardiac CT angiography axial and sagittal view, demonstrates circumferential wall thickening throughout the aorta C) MIP oblique reconstruction of aorta shows significant stenosis of left subclavian artery (white arrow). D) MIP oblique reconstruction of aorta demonstrates significant stenosis of the right subclavian and vertebral artery (yellow arrow).

resolution of heart failure symptoms and an improvement in ejection fraction.

Conclusion

This case report underscores the significance of investigating potential vasculitis in young patients presenting with heart failure. Early diagnosis of vasculitis and the implementation of appropriate anti-inflammatory measures can prevent or mitigate the development of vasculitis-related complications in these patients.

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