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CASE REPORT

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Unusual presentation of Takayasu Arteritis during pregnancy mimicking peripartum cardiomyopathy

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Abstract

Takayasu arteritis (TA) is a chronic inflammatory vasculitis affecting medium and large arteries predominantly in young women which can result in lethal complications. The pathology involves mononuclear cell infiltration and granulomatous inflammation in the arterial wall, arterial thickening, stenosis, occlusion and aneurysmal dilation. During the acute phase, patients may experience non-specific constitutional symptoms which can pose significant diagnostic dilemmas. Here we describe a 33-year-old woman who presented with generalised weakness, fatigue, and malaise who was treated for peripartum cardiomyopathy and subsequently suspected to have TA based on an incidental finding of circumferential wall thickening of the carotid arteries on an ultrasound scan of thyroid. An aortogram confirmed the characteristic features of TA. Prompt diagnosis and intervention are essential to prevent life-threatening complications of this relatively rare condition.

Keywords: Takayasu arteritis, medium and large vessel vasculitis, peripartum cardiomyopathy

Introduction

Takayasu arteritis (TA) is a rare vasculitis that affects large arteries, causing granulomatous inflammation, leading to complications such as stenosis, occlusion, and aneurysmal dilation. Advanced stages may result in life-threatening complications like cerebral thrombosis, haemorrhage, myocardial infarction, aneurysm rupture, pulmonary and systemic hypertension, or organ failure due to compromised blood flow.(1) The incidence of stenosis or occlusion and aneurysm formation has regional variation.(2,3) TA was first described by Mikito Takayasu in 1908.(4,5) A noteworthy correlation exists between retinal artery and large artery involvement in TA. Retinal microaneurysms serve as a prognostic indicator of disease severity.(6) Updated criteria by the American College of Rheumatology published in 2022 has improved the accuracy of diagnosis.(7) Magnetic resonance arteriography(MRA) is now considered to be the preferred non-invasive imaging technique, with computed tomography angiography(CTA) and ultrasound considered as alternatives.(8) Fluorodeoxyglucose positron emission tomography (FDG-PET) helps to measure vascular inflammation and assess disease activity.(9)

Biopsies reveal adventitial inflammation, elastic tissue destruction in media, neovascularisation of intima and media.(10) Macrophages play a crucial role in inflammation and remodeling.(11) First-line treatments include steroids and methotrexate.(14) For severe cases, Tumor necrosis factor (TNF) inhibitors and Janus Kinase (JAK) inhibitors are recommended. Disease activity is assessed using clinical indices and biomarkers.(15)

Case presentation

A 33-year-old woman diagnosed to have dilated cardiomyopathy, diabetes mellitus and bronchial asthma who has undergone evaluation for

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persistently elevated inflammatory markers since 2019 presented with generalized malaise and weakness for two weeks. She did not have loss of weight or appetite, night sweats or fever. There were no features suggestive of connective tissue diseases or autoimmune disorders. She had no contact history of tuberculosis or history suggestive of high-risk behavior. An intermittent cough was noted and treated as bronchial asthma since 2019. She had undergone elective caesarean section in 2020 at 32+4 weeks of gestation due to severe left ventricular dysfunction with pulmonary hypertension. There was no significant allergic history or family history noted.

Physical examination during the initial evaluation was unremarkable except for bilateral thyroid nodules with elevated blood pressure. In this presentation, her body temperature was 38.2°C, and her heart rate was 96 beats per minute. Examinations of the chest and precordium were unremarkable

Table 1 summarises baseline investigations performed during the course of the illness. Again the investigations revealed very high inflammatory markers with anaemia. Further investigations were performed to identify a cause. ECG showed T inversions in anterolateral chest leads. Trans-thoracic echocardiography demonstrated global hypokinesia with severe left ventricular dysfunction with an ejection fraction of 35% which could not be attributed to an underlying cause.

Blood picture showed anaemia of chronic disease and serum protein electrophoresis consistent with chronic inflammation. Blood cultures, venereal disease screening and autoimmune serological tests and chronic granulomatous disease screenings were unrewarding. Her renal, thyroid and liver profiles were normal except for a reversed albumin-globulin ratio.

Ultrasound scan of the neck showed small TR3 nodules in both thyroid lobes with an incidental finding of circumferential wall thickening of carotid arteries which raised the possibility of an underlying large vessel vasculitis. Carotid duplex showed bilateral diffuse circumferential wall thickening of common carotid arteries with approximately 50% stenosis.

Patient was reexamined based on the ultrasound scan findings which revealed a striking difference in blood pressures between the right and left arms. Her systolic blood pressure in the left arm was between 90-100 mmHg while diastolic blood pressure was 80 mmHg. In her right arm, the systolic blood pressure was 140 and the diastolic blood pressure was 70 mmHg. The brachial and radial pulses on her left were feeble but normal in character. Carotid and subclavian artery bruits were present on the left side.

The CT Aortogram carried out showed circumferential wall thickening of the aorta involving the aortic arch up to the bifurcation, right brachiocephalic trunk, bilateral common carotid, proximal internal carotid and subclavian arteries.

Table 1 - Summary of investigations

Investigation	Reference range	05/04 2019	19/09 2020	23/02 2024	27/06 2024	11/09 2024	21/09 2024
WBC (10^9/L)	4-10	9.04	9.14	9.30	9.03	9.15	9.92
Neutrophil (%)	50-70	64	60	48	60	62	64
Lymphocyte (%)	20-40	33	34	36	38	32	33
Hemoglobin (g/dL)	11-15	9.4	11.3	10.8	9.4	10.5	11.8
MCV (fL)	80-100	77	76.6	81.2	77.4	76.9	75.3
MCH (pg)	27-34	28.6	26.2	28.6	27.4	24.3	24.6
RDW (%)	11-16	14	16.4	17.2	16	18.2	19.3
Platelets (10^9/L)	150-450	532	447	494	532	489	488
ESR (mm/1st hour)	<20	140	139	117	140	80	20
CRP (mg/L)	0-3	182	160	145	147	127.4	10.2

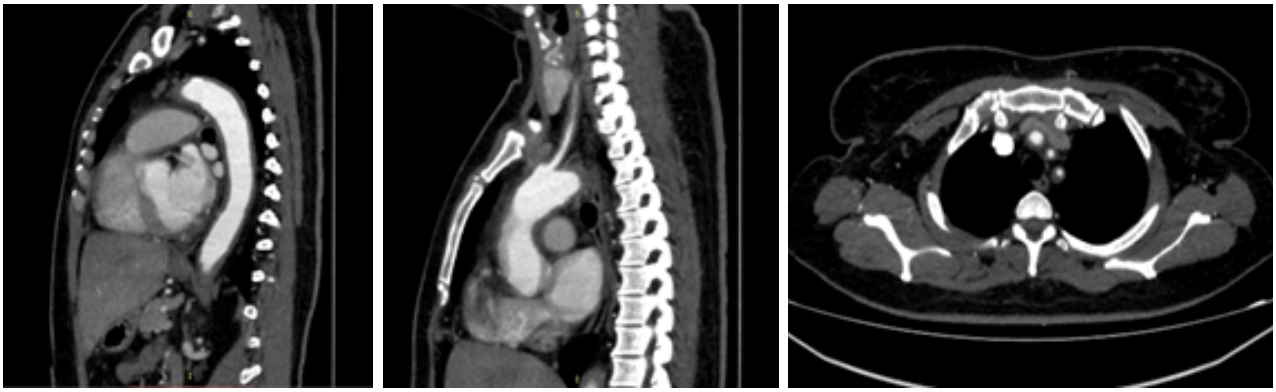
Mild wall thickening is observed in the bilateral common iliac, proximal superior mesenteric, bilateral renal arteries. There is moderate luminal narrowing of the proximal coeliac trunk at the origin (figure 1).

She was treated with a tapering regimen of oral prednisolone which started at 60 mg mane, azathioprine 100 mg daily and with other supportive measures after which she became asymptomatic and her inflammatory markers became normal gradually over three weeks. Oral prednisolone was tapered at an interval of every two weeks. She remained symptom-free following discharge. A repeat 2D echocardiogram showed an ejection fraction of 65%. A coronary angiogram was not performed since the patient refused.

Figure 2 outlines the chronology of evaluation since 2019.

Discussion

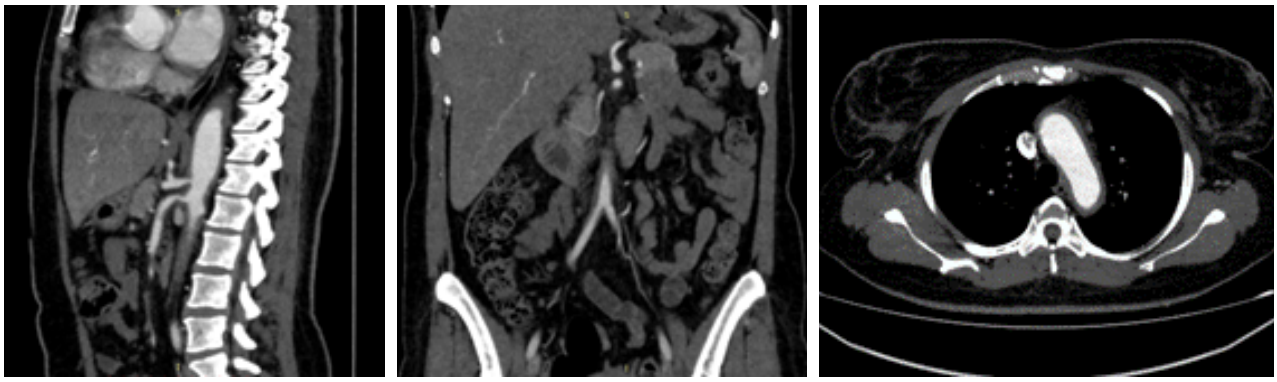
Diagnosis of TA is challenging and involves ruling out similar aetiopathology such as atherosclerosis, fibromuscular dysplasia, tuberculosis, syphilis, systemic lupus erythematosus, rheumatoid arthritis, sarcoidosis, Marfan syndrome and giant cell arteritis. (12,13) The gold standard diagnostic tool is angiography, although non-invasive techniques like Doppler ultrasound and magnetic resonance angiography (MRA) are also highly effective.(5,8) Criteria by American College of Rheumatology (ACR)



Descending thoracic aorta (sagittal)

Left common carotid artery (sagittal)

Branches of aortic arch (axial)



Coeliac trunk and SMA origin from the abdominal aorta (sagittal)

Aortic bifurcation (coronal)

Arch of aorta (axial)

Figure 1 - CT aortogram demonstrating luminal wall thickening

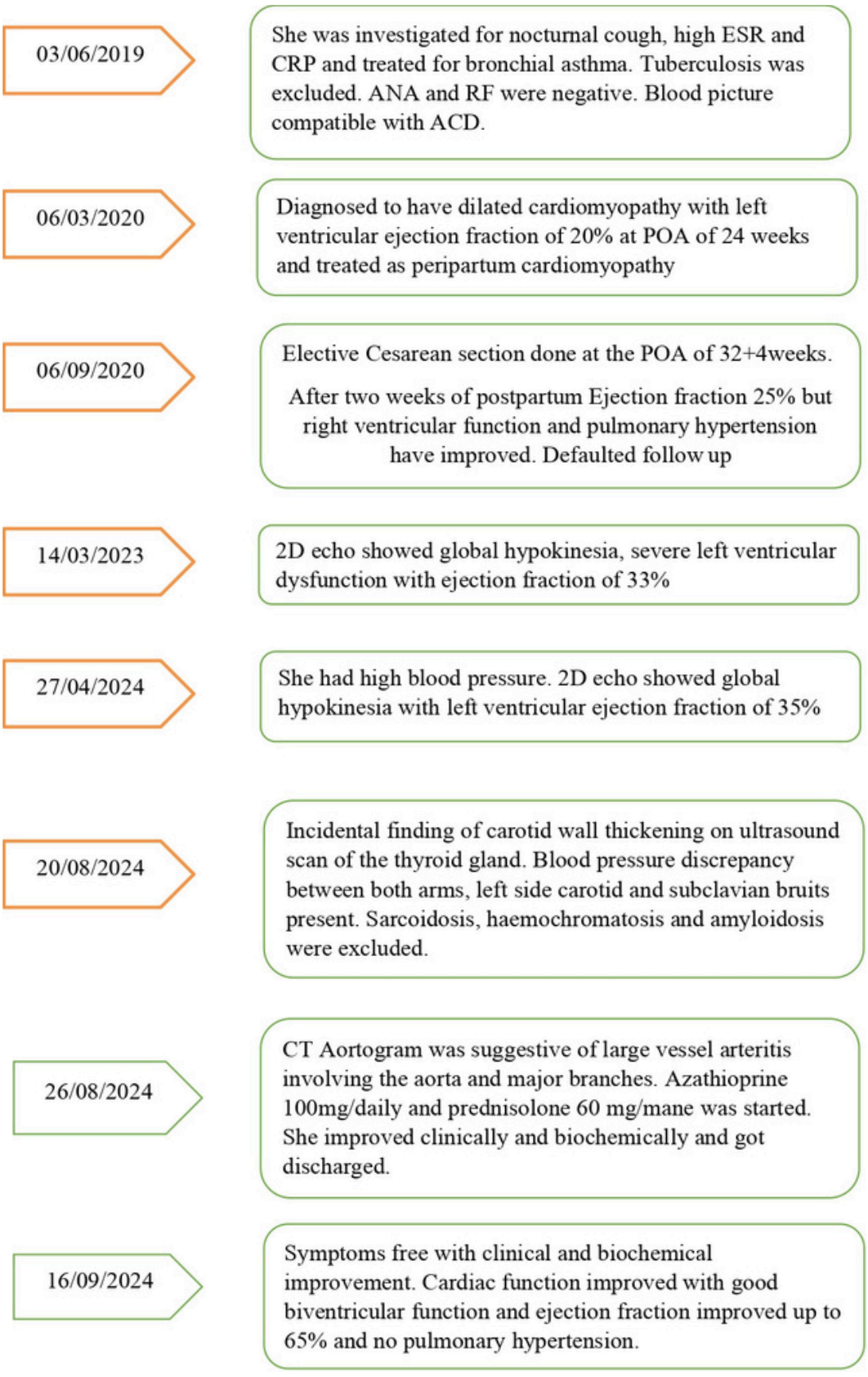


Figure 2 - Chronology of evaluation since 2019

for Takayasu arteritis show a sensitivity of 90.5% and a specificity of 97.8%. Additionally, elevated acute phase reactants (ESR,CRP) are often present, supporting the diagnosis.(6)

This case highlights the importance of challenges encountered in early detection of TA when it presents with non-specific symptoms. In this case, the differential diagnoses included other primary vasculitides, which do not present with a discrepancy in blood pressures between the arms, which is a key feature in TA. This specific physical sign would be easily missed unless looked for in particular. TA primarily affects large vessels and their branches, with symptoms corresponding to the specific vessels involved. In this case there were no significant occlusive symptoms except for the discrepancy in blood pressures between the arms.

The cornerstone of management focuses on minimising narrowing of vessels rendered with immunosuppression and systemic corticosteroids. (14) Surgical or endovascular interventions may be considered in severe cases to restore blood flow and prevent further complications.(16)

Conclusion

This case report illustrates the challenges in diagnosing vasculitis when a patient presents with nonspecific manifestations that mimic other systemic illnesses. TA is one such condition that could pose a challenge in diagnosis as it is predominant in young females where other autoimmune diseases and pregnancy related conditions are common. Non-invasive imaging studies are proven to be beneficial in picking up vital pathology like TA even in low resource settings, and should be sorted whenever possible.

Declarations

Conflicts of interest
The authors declare that they have no conflicts of interest

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