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Case Report

Complex Management of Takayasu Arteritis with Multiorgan Involvement: A 4-Year Follow-up Case Report

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ABSTRACT

Takayasu Arteritis (TA) is a rare vasculitis characterised by inflammation primarily affecting the aorta and its branches, predominantly observed in young Asian females. This case report documents the clinical journey of a 28-year-old Asian male diagnosed with TA over a four-year period. Initially presenting with claudication and chest pain, the patient was found to have extensive coronary, pulmonary and renal artery involvement. Recurrent hospitalisations ensued due to complications including anaemia, accelerated hypertension, renal failure and sepsis. Management challenges arose, complicated by adverse drug reactions and the necessity for haemodialysis. Effective management of TA mandates early diagnosis, judicious employment of immunosuppressive therapy and vigilant monitoring. This case underscores the essential role of a multidisciplinary approach and personalised care in navigating the complex challenges posed by TA. It emphasises the importance of clinician awareness of diagnostic criteria, facilitating timely intervention and optimising patient outcomes. By elucidating the intricate nuances of TA management, this report contributes to a deeper understanding of this rare disease and advocates for tailored therapeutic strategies addressing the complexities of individual patient presentations.

Keywords: Takayasu arteritis, Renal failure, Pulmonary arterial hypertension, Coronary artery disease, Myocardial infarction, Cardiomyopathy, Acute coronary syndrome, Congestive heart failure, Dialysis.

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INTRODUCTION

Takayasu Arteritis (TA) is a rare, chronic and progressive condition that is characterized by inflammation, primarily affecting aorta and its branches.[1] The disease commonly involves large vessels such as the ascending aorta, aortic arch, descending thoracic and abdominal aorta and its branches, usually the renal arteries.[2] The incidence rate is 1-2 cases per million globally.[3] TA is more prevalent in young Asian females but can also occur in young males.[4] Mortality often results from comorbidities namely, congestive heart failure, myocardial infarction and renal failure.[5] We describe the 4-year course of a patient with the diagnosis of TA with extensive coronary, pulmonary and renal artery involvement.

CASE REPORT

A 28-year-old Asian male presented with claudication in lower extremities and recurrent episodes of chest pain in December 2018. His Heart Rate (HR) was at 102 beats/min, Blood Pressure (BP) was 170/90 mm Hg, Respiratory Rate (RR) was 18 breaths/min. The jugular venous pulse was not elevated. S1 and S2 heart sounds were heard but not S3 and S4. Laboratory evaluation showed normal hematological, biochemical and urine analyses. Electrocardiography revealed sinus rhythm at 75 beats per min, QRS 80 milliseconds, PR 120 milliseconds and ST segment elevation in leads V1-V4. Echocardiogram revealed fair Left Ventricular (LV) systolic function with no Pulmonary Arterial Hypertension (PAH). Coronary angioplasty was performed using Percutaneous Old Balloon Angioplasty (POBA) and thrombus aspiration to Left Main Coronary Artery (LMCA). The procedure showed ostial LMCA and Left Anterior Descending (LAD) artery disease (Figure 1). The patient was diagnosed with Coronary Artery Disease (CAD)-Acute Coronary Syndrome (ACS)-extensive anterior wall myocardial infarction. After 7 months, the patient underwent a coronary angiogram and aortic angiogram, which showed moderate stenosis of left main artery. Thoracic

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aortogram revealed infrarenal abdominal aortic complete occlusion (Figure 2) from internal mammary artery, superior mesenteric artery, intercostals, and lumbar arteries.

The patient presented with a complaint of epistaxis in early 2021. His HR was 92 beats/min, BP was 160/100 mm Hg. Electrocardiography revealed anteroseptal MI, hypertrophic cardiomyopathy and left ventricular hypertrophy (Figure 3). After 3 months, he presented with complaints of abdominal pain, and tenderness. Abdominal X-ray revealed intestinal obstruction. The symptoms improved with the administration of lactulose.

In mid 2022, he presented with chief complaints of cough, dyspnea and abdominal discomfort. His HR was 88 beats per min, BP was 200/110 mm Hg, oxygen saturation was 97%, Hemoglobin (Hb) was low and creatinine was slightly high (1.7 mg/dL). 2D ECHO was performed which showed dilated LV global hypokinesia, hypokinetic LAD territory, moderate to severe PAH and severe LV systolic dysfunction. Renal Doppler reports were suggestive of renal ischemia. He was diagnosed with accelerated hypertension, mild renal failure, infra-renal aorta total occlusion resulting in Peripheral Arterial Disease (PAD) and TA. The patient also underwent POBA to treat blockage in LAD. The patient was

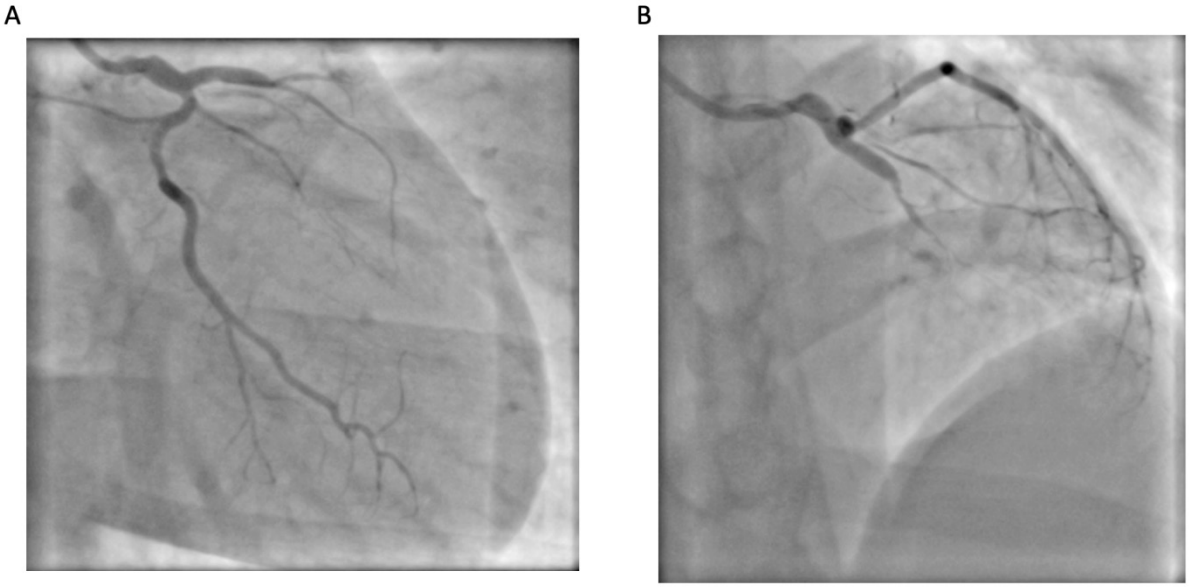

Figure 1: A-B. Coronary angiogram showing occlusion of ostial left main coronary artery and left anterior descending artery.

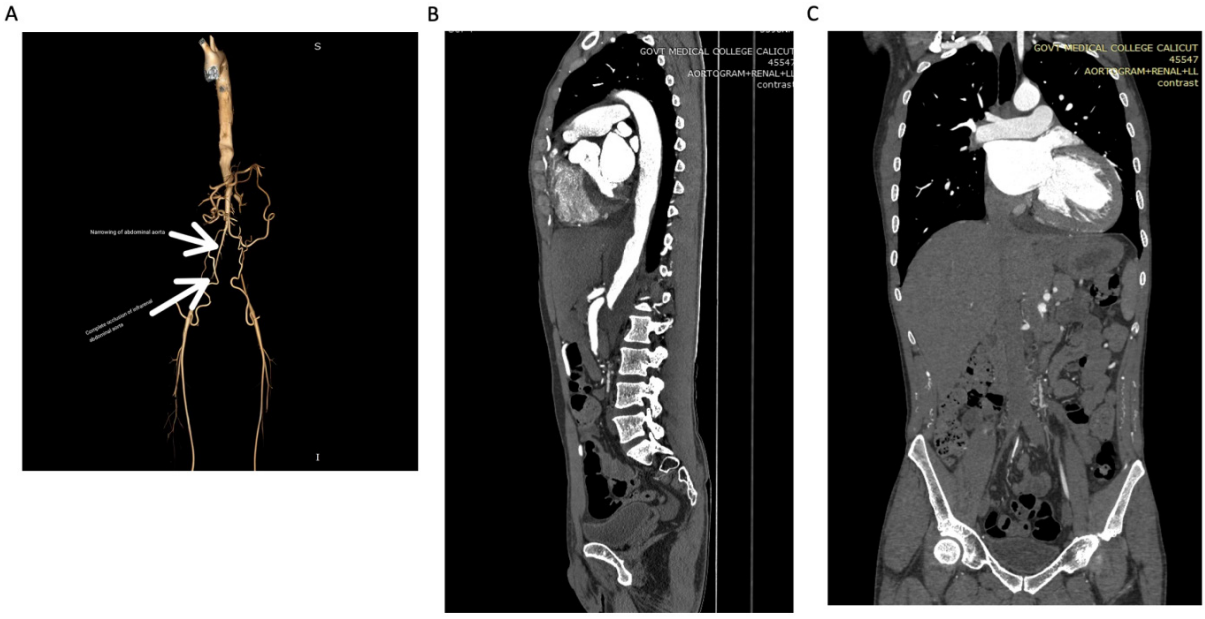

Figure 2: A. CT Aortogram showing narrowing of abdominal aorta and complete occlusion of infrarenal abdominal aorta (white arrows). Aortogram with renal and lower limb contrast showing B. Sagittal view of abdominal aorta occlusion and C. Coronal view of renal occlusion.

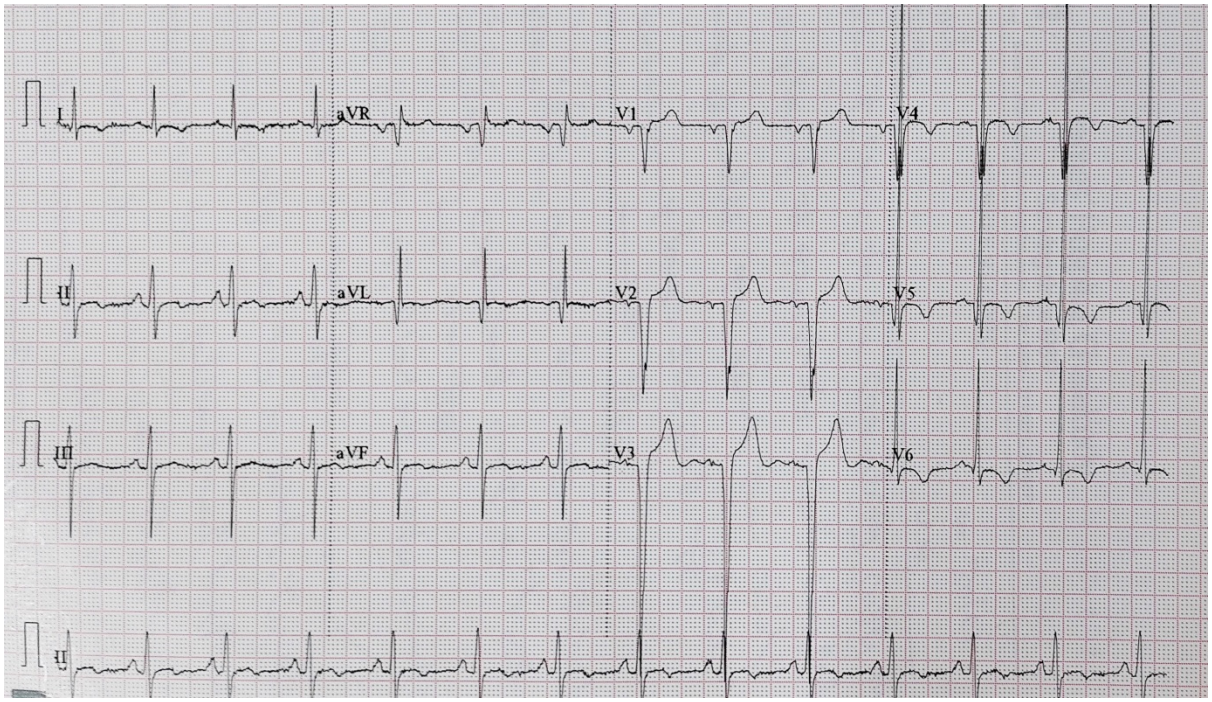

Figure 3: Electrocardiogram showing abnormal Q waves (V1, V2, V3, V4, V5), subsequent T wave abnormality, anteroseptal myocardial infarction, hypertrophic cardiomyopathy and left ventricular hypertrophy.

managed with antihypertensives, IV antibiotics and symptomatic measures. He improved with treatment and was discharged after three days. Following discharge, the patient presented after 2 months with complaints of cough, throat discomfort and bilateral pedal edema for more than a week. His HR was 68 beats/min, BP was 140/80 mm Hg, Hb was 8.7 g/dL, serum creatinine (3.7 mg/dL) and blood urea (227 mg/dL) levels were high. He was also diagnosed with anemia after 2 months when Hb levels were between 4.7 and 6.8 g/dL. His creatinine levels were 2.3 mg/dL and he was diagnosed with Chronic Kidney Disease (CKD). After 1 month, the patient presented with complaints of cough and fever. His BP was 140/80 mm Hg, HR was 68 beats/min, RR was 20 breaths/min. Hemodialysis was also performed in view of desaturation and elevated creatinine levels. The patient also complained of itching and watering eyes for which treatment was given. He was diagnosed with congestive heart failure and lower respiratory tract infection. The patient presented with symptoms of increased fluid retention 2 weeks after discharge. He also experienced transient thrombocytopenia and an aspirate culture taken from his dialysis catheter showed positive results. His blood pressure was 160/100 mm Hg, heart rate was 92 beats/min and oxygen saturation was 95%. The patient was advised for hemodialysis on alternate days.

At the beginning of 2023, an arteriovenous fistula was created in the patient. His Hb levels were still low and creatinine levels were high. He also experienced restless leg syndrome, one of the complications in hemodialysis patients.

DISCUSSION

We present the case of a 28-year-old male with rapidly progressive TA and multiple comorbidities over 4 years. TA is a chronic, relapsing and difficult-to-diagnose condition, as the clinical features are non-specific.[6] There is significant variation in the long-term prognosis of patients with TA.[7] Though unclear, the etiopathogenesis involves a combination of genetic factors, supported by the association with HLA-B52, autoimmune-mediated processes, vascular remodeling and endothelial dysfunction.[8] It may also be triggered by viral or bacterial (*Mycobacterium tuberculosis*) infections.[9] Management of TA involves early diagnosis, effective therapy and continued monitoring to improve patient outcomes.[10] In this case, the patient had multiple comorbidities that deteriorated his clinical course and various complications that required repeated hospitalizations. While the physicians did not employ diagnostic criteria to confirm the diagnosis, the initial presentation of claudication of extremities is one of the criteria under the Modified Ishikawa Criteria for diagnosing TA.[11] With imaging evidence of vasculitis and age under 60 years, the patient also meets the 'ACR/EULAR classification criteria for TA'.[12] Encouraging physicians to include diagnostic criteria for TA while managing patients with vasculitis may promote early diagnosis and improve outcomes.

Other clinical features vary depending on the stage of TA and the region involved. Under 20% of TA, patients show a classic triphasic pattern of the pre-pulseless stage with constitutional symptoms,[13] the pulseless stage with vascular inflammation and ischemia and the fibrotic stage with vascular damage.

Additionally, coronary artery involvement is seen in 7% to 9% of cases presenting a therapeutic challenge.[14] Chest pain, in this case, was attributed to ACS and subsequent angiography showed moderate stenosis of the left main artery. The patient underwent POBA to treat the LAD blockage. However, after seven months, further angiography showed infrarenal abdominal aortic complete occlusion, leading to PAD. This is a rare presentation in a young patient that required urgent intervention. TA can also contribute to the development of aortic stenosis and PAD in the patient. Additionally, hypertrophic cardiomyopathy can also cause LV hypertrophy and increase the risk of cardiovascular events, such as ACS. PAH in patients with TA is a rare condition.[15] This patient was diagnosed with PAH in mid-2022 without confirmation using Right Heart Catheterization (RHC) which is the gold standard for PAH diagnosis.[16] The mechanisms that contribute to PAH associated with TA are not well known and the prognosis in patients with PA involvement with PAH is worse than in those without it. It is unclear whether PAH-specific therapies could be effective in the treatment of PAH associated with TA.

The patient also had CKD, which is a known risk factor for cardiovascular disease and is associated with an increased risk of mortality. This comorbidity may have contributed to the patient's accelerated hypertension and renal failure. The patient's renal dysfunction required hemodialysis, which caused complications such as sepsis and restless leg syndrome. In addition, the patient had fluid retention, which may have been due to renal dysfunction or heart failure. The patient had anemia, which may have been due to CKD, requiring blood transfusions for management of anemia.

The primary treatment for TA involves high-dose glucocorticoids such as prednisolone and immunosuppressive agents like azathioprine.[17] Glucocorticoid therapy alleviates symptoms but does not improve survival.[18] This patient experienced azathioprine-induced pancytopenia and febrile neutropenia, and the drug was withheld. Severe pancytopenia and febrile neutropenia while receiving azathioprine is a serious but uncommon complication.[19,20] Cell counts should be frequently monitored after initiating azathioprine. Such adverse drug reactions emphasize the need for drug monitoring and individualized treatment approaches in patients with TA.

CONCLUSION

In summary, TA is a rare disease with no curative therapy. This case highlights the importance for a multidisciplinary approach and comprehensive care in the management of patients with TA. The multiorgan involvement of the case presents a unique diagnostic and therapeutic challenge. The patient required multiple hospitalizations for various complications, including anemia, accelerated hypertension, renal failure and sepsis. The multiorgan involvement of the case presents a unique diagnostic and therapeutic challenge. This case emphasizes the need for personalized care in the management of TA patients with complex medical histories.

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

Ethics approval was not required for this single case report as per institutional guidelines. Written informed consent was obtained from the patient for both publication and participation in this case report.

ABBREVIATIONS

TA: Takayasu arteritis; **HR:** Heart rate; **BP:** Blood pressure; **RR:** Respiratory rate; **PH:** Pulmonary hypertension; **PAH:** Pulmonary arterial hypertension; **POBA:** Percutaneous old balloon angioplasty; **LMCA:** Left main coronary artery; **LAD:** Left anterior descending; **CAD:** Coronary artery disease; **ACS:** Acute coronary syndrome; **Hb:** Hemoglobin; **PAD:** Peripheral arterial disease; **CKD:** Chronic kidney disease.

SUMMARY

This case report details the 4-year course of a 28-year-old Asian male with TA, a rare vasculitis. Initially presenting with claudication and chest pain, he underwent coronary angioplasty for ACS. Over time, he developed multi-organ complications, including PAH, CKD and hypertrophic cardiomyopathy, necessitating hemodialysis and anaemia management. Despite treatment challenges, including adverse reactions to Azathioprine, this case underscores the need for comprehensive, personalized care in managing TA. The patient's complex medical history highlights the diagnostic and therapeutic challenges associated with TA, emphasizing the importance of a multidisciplinary approach to optimizing patient outcomes.

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