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Case Report

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Multidisciplinary Management of Type IIa Takayasu Arteritis with Mechanical Aortic Valve and Subclavian Artery Occlusion: A Complex Case Report

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ABSTRACT

Takayasu arteritis (TA) is a rare, chronic large-vessel vasculitis characterized by granulomatous inflammation of the aorta and its major branches. Diagnosis and management are challenging due to its insidious presentation, variable vascular involvement, and frequent relapsing course. We report the case of a 52-year-old European woman with Type IIa TA complicated by bilateral subclavian artery disease, prior Dacron graft repair for an ascending aortic aneurysm, and mechanical aortic valve replacement for severe regurgitation. The patient presented with metrorrhagia requiring gynecological intervention, raising significant perioperative concerns due to her chronic anticoagulation and high cardiovascular risk profile. On admission, she demonstrated absent left upper limb pulses, anemia secondary to ongoing bleeding, and an elevated INR. Anticoagulation was carefully bridged with low molecular weight heparin, in accordance with international guidelines for mechanical prosthetic valves, allowing safe biopsy and polypectomy with concurrent endocarditis prophylaxis. During hospitalization, she experienced hypotensive episodes and brief atrial fibrillation, necessitating optimization of antihypertensive therapy and rhythm monitoring. Ongoing rheumatologic follow-up was arranged, with continued low-dose methotrexate and corticosteroid therapy for disease control. This case highlights the intricate balance required in managing TA patients with complex cardiovascular comorbidities, emphasizing the interplay between autoimmune inflammation, vascular complications, anticoagulation strategies, and procedural safety. Effective outcomes relied on a multidisciplinary approach involving cardiology, rheumatology, gynecology, and hematology. Our report underscores the importance of individualized treatment strategies in TA, especially when complicated by prosthetic heart valves, grafts, and severe large-vessel disease.

Keywords: Takayasu Arteritis, Type IIa, Subclavian Artery Occlusion, Mechanical Aortic Valve, Anticoagulation Bridging, Multidisciplinary Management, Case Report

Introduction

Takayasu arteritis (TA) is a rare, chronic granulomatous vasculitis that primarily targets large arteries, especially the aorta and its major branches. Most frequently affecting young women under the age of 40, particularly of Asian descent, though it can occur globally and in older individuals, it remains a challenging diagnosis due to its insidious onset and nonspecific early symptoms [1]. TA progresses in two phases: an early systemic phase characterized by constitutional symptoms such as fatigue, fever and arthralgia, and a later occlusive phase marked by ischemic symptoms to progressive arterial stenosis or aneurysm formation.

The Numano classification system divides TA into six types based on angiographic findings (see table 1 below). Type IIa (in our case) involves the ascending aorta, aortic arch and its main branches, but spares the descending and abdominal aorta [2]. This subtype most often compromises blood flow to the upper limbs and brain, posing risks such as limb claudication, syncope and stroke.

The underlying mechanism of Takayasu arteritis is thought to involve an autoimmune process, where activated T-cells and the release of pro-inflammatory cytokines especially interleukin-6 (IL-6) and tumor necrosis factor-alpha (TNF-α) lead to progressive damage of the vascular walls [3]. Diagnostic evaluation and disease monitoring heavily rely on imaging techniques with MRI and CT angiography offering high sensitivity for detecting vessel wall thickening, inflammation, stenotic lesions and aneurysmal changes [4].

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Standard treatment begins with high dose corticosteroids. Immunosuppressants like methotrexate or azathioprine are used for maintenance or steroid sparing effects. In refractory or relapsing cases, biologics such as tocilizumab or TNF-α inhibitors may be employed [5]. Adjunct antiplatelet therapy is often recommended to reduce thrombotic risk.

Table 1: Numano Classification of Takayasu Arteritis

Type	Arterial Involvement	Description	
Type I	Branches of the aortic arch only	This includes the brachiocephalic trunk, carotid arteries and subclavian arteries. This type is sometimes referred to as "occlusive cerebrovascular disease" due to its higher association with neurological symptoms.	[2]
Type IIa	Ascending aorta, aortic arch and its branches	Often involves bilateral subclavian or carotid disease and may present with arm claudication, pulse deficits and blood pressure asymmetry.	[2]
Type IIb	Ascending aorta, aortic arch, its branches and thoracic descending aorta	Involves both the aortic arch and descending thoracic aorta. This may present with more widespread vascular symptoms including limb ischemia, hypertension and even aortic aneurysm.	[2]
Type III	Thoracic descending aorta, abdominal aorta and/or renal arteries	Sparing the aortic arch and its branches. This type is commonly associated with renovascular hypertension, mesenteric ischemia and lower limb claudication.	[2]
Type IV	Abdominal aorta and/or renal arteries only	Mostly confined to the abdominal vessels, particularly the renal arteries and may mimic fibromuscular dysplasia or other causes of secondary hypertension.	[2]
Type V	Combined involvement of the aortic arch, thoracic descending aorta and abdominal aorta (i.e., I + III or IIb + IV)	Represents widespread disease. This type has the most systemic impact and is associated with higher rates of complications like aortic regurgitation, aneurysms and stroke.	[2]

In this report, we present a complex case of a 52-year-old woman with Type IIa Takayasu arteritis, complicated by bilateral subclavian artery disease, aortic valve mechanical prosthesis and previous Dacron graft placement for ascending aortic aneurysm. The coexistence of severe hypertension, iron deficiency anemia and a need for gynecological intervention required multidisciplinary coordination. Her case exemplifies the intricate interplay of vascular, autoimmune and procedural risk in managing TA patients, especially those with prosthetic heart valves and altered vascular anatomy.

Case Presentation

A 52 y/o European female presented to our hospital with a complex cardiovascular and autoimmune profile, including a known history of Takayasu arteritis type IIa, as well as mechanical aortic valve replacement due to severe aortic insufficiency, Dacron graft placement for an ascending aortic aneurysm and significant subclavian artery disease with complete occlusion of the left and stenosis of the right subclavian artery. These vascular changes are commonly consistent with Takayasu arteritis which predominantly affects large vessels such as the aorta and its primary branches.

The patient also suffers from grade 3 hypertension with very high cardiovascular risk, iron deficiency anemia, chronic gastritis, fatty liver disease and spondyloarthrosis with nerve root involvement, reflecting the multi system burden of both inflammatory and degenerative disease processes.

She was admitted for pre operative evaluation and anticoagulant management, in preparation for an endometrial biopsy due to one month of metrorrhagia. At admission she was stable with a blood pressure of 150/85 mmHg, a regular pulse of 68 bpm and oxygen saturation of 97%. Upon clinical examination a loud mechanical murmur was heard in the aortic area, consistent with her prosthetic valve, while the peripheral pulse on the left upper limb was absent, aligning with her known subclavian occlusion.

Laboratory tests revealed a case of normocytic normochromic anemia with reticulocytosis suggesting ongoing blood loss, likely gynecological in origin. The inflammatory markers showed mild elevation with ESR being 37 mm/hr and CRP at 0.72 mg/dL. Importantly her INR was elevated at 3.25, necessitating cessation of acenocoumarol and transition to low molecular weight heparin (LMWH); Fraxiparine 0.8 mL BID for safer perioperative anticoagulation which is a standard bridging strategy in patients with mechanical heart valves [6].

A transthoracic echocardiogram confirmed appropriate function of the mechanical aortic valve, left ventricular hypertrophy and mildly enlarged left atrium. Mitral and tricuspid regurgitation were minimal, with preserved global systolic and diastolic function. Mild pulmonary hypertension was suspected based on tricuspid regurgitation velocity.

On April 13, the patient reported worsening bleeding. A follow up complete blood count showed a further drop in hemoglobin by 1 g/dL over 24 hours, prompting transfusion of one unit of red blood cells. After stabilization and anticoagulation adjustment, a gynecological evaluation was performed on April 15, including removal of an endocervical polyp and endometrial sampling using a Novak curette. Given the mechanical valve, prophylactic antibiotics; ampicillin 2 g IV were administered to reduce the risk of infective endocarditis, in accordance with current ESC guidelines [7].

During hospitalization, the patient experienced a brief syncope and recurrent presyncopal episodes. A 24-hour blood pressure

Holter monitor revealed a tendency toward hypotension around midday, leading to an adjustment in antihypertensive therapy specifically, discontinuation of indapamide and halving of perindopril to 5 mg daily.

An ECG Holter showed mostly normal findings, with occasional supraventricular and ventricular ectopics and a short 7-second episode of atrial fibrillation. Although brief, such arrhythmias warrant close monitoring in a patient with prosthetic valves and prior aortic surgery due to elevated thromboembolic risk. Notably, after adjusting therapy, no further arrhythmic symptoms occurred.

Given the underlying autoimmune vasculitis, a rheumatology consult was also requested. The patient remained on low dose methotrexate about 2.5 mg weekly and was advised to follow up in one month with updated inflammatory markers. Folic acid and vitamin D supplementation were continued.

At discharge, the patient was stable and advised to maintain regular follow up. Her anticoagulation regimen was resumed with adjusted acenocoumarol dosing, targeting an INR of 2.5 and the patient was educated on switching back to Fraxiparine during any future dental or surgical procedures, along with prophylactic antibiotics as needed.

Discussion

This case illustrates the intricate challenges faced in managing a patient with type IIa Takayasu arteritis (TA), who additionally has a mechanical aortic valve, ascending aortic Dacron graft and subclavian artery disease, complicated by metrorrhagia necessitating an invasive procedure.

Vascular Complications and Ischemic Symptoms

Type IIa TA involves the ascending aorta and primary branches, including the subclavian and carotid arteries, consistent with our patient's bilateral subclavian disease, leading to absent pulses and syncope. These findings align with well documented ischemic manifestations such as subclavian steal syndrome, where significant proximal subclavian stenosis can lead to retrograde vertebral flow and neurologic symptoms like dizziness and syncope [8,9].

Perioperative Anticoagulation in Mechanical Valve Carriers

Managing anticoagulation in the context of a mechanical heart valve is particularly precarious. Guidelines recommend bridging with low molecular weight heparin (LMWH) during periods of vitamin K antagonist (VKA) interruption to mitigate the high thromboembolic risk, especially for non cardiac procedures [10,11].

In this case, the elevated INR on admission warranted transition to LMWH, which proved essential for balancing bleeding risk during biopsy with the need to prevent valve thrombosis. Current evidence supports stopping VKA approximately five days pre procedure with resumption 12 to 24 hours post procedure when hemostasis is assured [10].

Immunosuppressive and Biologic Therapy in Takayasu Arteritis

First line therapy for TA typically involves high-dose glucocorticoids, often followed by steroid sparing agents such as methotrexate for long-term disease control. In refractory or relapsing disease, biologic therapies, especially IL-6 inhibitors like tocilizumab and TNF-α antagonists are increasingly supported by evidence [12].

Notably, the TAKT (Takayasu arteritis Treated with Tocilizumab) phase 3 trial demonstrated that weekly subcutaneous tocilizumab reduced relapse risk compared to placebo, with better outcomes in the per protocol analysis [13]. Additionally, long-term open label extension data confirmed a steroid sparing effect and stable or improved disease on imaging up to 96 weeks, with favorable safety profiles [14]. Regional real-world data, like the Japanese

ACT-Bridge study, further support the sustained effectiveness and tolerability of tocilizumab in diverse TA populations [15].

Our patient remains on low dose methotrexate and steroids, with plans for reevaluation in one month to assess disease activity and steroid tapering aligning well with contemporary treatment approaches.

Holistic Multi-Disciplinary Management

This case underscores the indispensable role of interdisciplinary coordination among cardiology, rheumatology, gynecology and hematology. For example:

- Gynecological Intervention was carefully timed post-LMWH bridging and included endocarditis prophylaxis; ampicillin 2g IV, following ESC recommendations for prosthetic valve carriers undergoing invasive procedures [7].
- Antihypertensive Optimization, particularly reducing perindopril and discontinuing indapamide in response to midday hypotension, illustrates individualized blood pressure management to prevent further syncope or arrhythmia exacerbation.
- Holter Monitoring revealed a brief episode of atrial fibrillation, reinforcing the need for close rhythm surveillance and potentially influencing long-term anticoagulation strategies.
- Patient Education, particularly around anticoagulation bridging protocols for future procedures, was integral to her discharge plan.

Conclusion

This case highlights the multifaceted challenges in managing patients with Takayasu arteritis who also carry significant cardiovascular comorbidities. Our patient, with a history of type IIa disease complicated by severe aortic insufficiency requiring mechanical valve replacement, prosthetic grafting of the ascending aorta and critical subclavian artery disease, presented with gynecological bleeding in the setting of chronic anticoagulation. Her clinical course illustrates the delicate balance required between preventing thromboembolic events, controlling systemic inflammation and minimizing hemorrhagic complications.

Takayasu arteritis remains a chronic relapsing remitting condition and its management extends beyond vascular interventions to long-term immunosuppressive therapy aimed at suppressing autoimmune mediated vascular inflammation. In this case, methotrexate and corticosteroids were continued, with rheumatology input for treatment adjustment.

Importantly, her peri procedural care demonstrates the role of careful anticoagulation bridging strategies in patients with mechanical prostheses, as well as the importance of multidisciplinary coordination between cardiology, rheumatology and gynecology.

This case underscores the need for individualized management guided by disease activity, comorbidities and procedural risks. It also reflects current understanding that optimal outcomes in Takayasu arteritis depend not only on controlling inflammation with immunosuppressive agents but also on addressing cardiovascular risk factors, monitoring vascular complications through imaging and adapting therapy to evolving clinical needs.

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