

Case Report

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Recurrent Anemia and Fever in a Young Male with Sickle Cell Disease and Bioprosthetic Valves: A Diagnostic Challenge in a Resource-Limited Setting

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Abstract

We report a challenging case of a 25-year-old male with homozygous sickle cell disease (SCD) and a history of bioprosthetic aortic and mitral valve replacement for rheumatic heart disease (RHD), performed 12 years ago. The patient presented to Tanga Regional Referral Hospital with recurrent anemia, exertional dyspnea, and low-grade fever, without jaundice. Notably, he had never been on anticoagulation post-surgery. The facility lacked both transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE), and a direct Coombs test was not available. He was managed empirically with ceftriaxonesulbactam and supportive care. This case emphasizes the complexities of managing overlapping chronic diseases and prosthetic valve complications in low-resource settings where standard diagnostic modalities are not accessible.

Keywords: sickle cell disease; bioprosthetic valves; rheumatic heart disease; anemia; echocardiography unavailable; resource-limited setting; ceftriaxone-sulbactam

Introduction

Sickle cell disease (SCD) and rheumatic heart disease (RHD) are both endemic in sub-Saharan Africa and may coexist in young individuals, compounding morbidity [1,2]. Bioprosthetic valves are often preferred over mechanical valves in patients unable to comply with lifelong anticoagulation or where monitoring is unavailable [3]. However, they are subject to earlier degeneration in younger patients, especially in the setting of chronic inflammation, such as that seen in SCD [4,5]. The utility of regular echocardiographic follow-up in such patients is undisputed [6], but many institutions, including Tanga Regional Referral Hospital, lack both transthoracic (TTE) and transesophageal (TEE) echocardiography. This significantly limits diagnostic capability, particularly in evaluating potential structural valve degeneration, thrombosis, or infective endocarditis [7].

Case Presentation

A 25-year-old male with known homozygous SCD presented with fatigue, worsening exertional dyspnea, and low-grade fever for three weeks. He denied any history of jaundice, hematuria, bleeding, weight loss, or respiratory symptoms.

Past Medical History

- Diagnosed with SCD at age 2; frequent vaso-occlusive crises in childhood.
- Bioprosthetic aortic and mitral valve replacement at age 13 for RHD.
- No anticoagulation since surgery.
- No cardiac or hematologic follow-up since surgery.

Clinical Examination

- General: Pale, afebrile, no jaundice
- Vitals: BP 110/68 mmHg, HR 96 bpm, RR 20/min, SpO₂ 94% RA
- Cardiac: Soft systolic murmur at the apex, no prosthetic clicks
- Respiratory: Clear auscultation
- Abdominal: Mild splenomegaly, no tenderness or organomegaly
- Neurological & Skin: Unremarkable

Investigations

- Hemoglobin: 6.3 g/dL
- MCV: 85 fL
- Reticulocyte Count: 5.6%
- WBC: 12,100/mm
- Platelets: 430,000/mm ESR: 65 mm/hr
- CRP: 26 mg/L
- LDH: 620 U/L
- Total Bilirubin: 1.1 mg/dL (Indirect: 0.4)
- Ferritin: 430 ng/mL Coombs Test: Not available HIV/Hepatitis B/C: Negative
- Blood Cultures: No growth after 5 days Chest X-ray: Cardiomegaly, clear lung fields TTE / TEE: Not available at our facility

Management

- 2 units of packed red blood cell transfusion
- Intravenous ceftriaxonesulbactam for 5 days empirically, then stopped after afebrile and cultures were negative
- Folic acid, hydration, and pain management
- Hydroxyurea optimization

- Initiated low-dose aspirin (75 mg daily)
- Deferred anticoagulation pending cardiac evaluation

Discussion

This case exemplifies the diagnostic uncertainty inherent in managing complex chronic disease in low-resource environments.

Bioprosthetic Valve Dysfunction

Bioprosthetic valves are more prone to degeneration in young, metabolically active patients and those with chronic inflammatory states like SCD (4,5). Without TTE or TEE, we could not assess for stenosis, regurgitation, or vegetation, thus clinical judgment was crucial.

Lack of Anticoagulation and Surveillance

Although bioprosthetic valves typically do not require long-term anticoagulation, in SCD patients with increased hypercoagulability, thrombotic complications have been reported (8). The absence of any follow-up echocardiography for over a decade significantly increases the likelihood of undiagnosed valve dysfunction (6).

Recurrent Anemia and Fever in SCD

Anemia in SCD is usually multifactorial: chronic hemolysis, inflammation, nutritional deficiency, or hypersplenism (9). Absence of jaundice and normal bilirubin suggested anemia of inflammation. The raised ESR/CRP with negative cultures and resolution of fever with antibiotics suggests subclinical infection, possibly involving the valve.

Imaging Limitations

The inability to perform both TTE and TEE at Tanga Regional Referral Hospital reflects a systemic gap in regional diagnostic capacity. Even basic surveillance tools were unavailable, severely compromising care (10).

Conclusion

This case emphasizes the vulnerability of SCD patients with prosthetic heart valves in settings without echocardiographic access. The absence of anticoagulation and lack of imaging over 12 years pose serious risks of undetected valve degeneration or endocarditis. It highlights the urgent need to strengthen diagnostic capacity and establish standardized follow-up protocols in

regional hospitals across Africa.

Patient Perspective

The patient reported that he believed valve surgery had "cured" his heart condition and was unaware of the need for lifelong monitoring. He expressed appreciation for the renewed attention and willingness to attend future cardiology follow-ups.

Informed Consent

Written informed consent was obtained from the patient for anonymous case publication.

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