

Quest for a Unifying Diagnosis: Felty Syndrome?

Biondi NL , Pico M  and Das M*

Division of Cardiovascular Disease, Department of Internal Medicine, University of Cincinnati, Ohio, USA

***Correspondence:** Manisha Das, Division of Cardiovascular Disease, Department of Internal Medicine, University of Cincinnati, Ohio, USA

Received on 17 April 2023; Accepted on 11 May 2023; Published on 18 May 2023

Copyright © 2023 Biondi NL, et al. This is an open-access article and is distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

Keywords: Felty syndrome, rheumatoid arthritis, neutropenia, infective endocarditis

Abbreviations: RA: rheumatoid arthritis; DMARDs: disease-modifying antirheumatic drugs

Introduction

Felty syndrome is a rare seropositive rheumatoid arthritis (RA), splenomegaly, and absolute neutropenia [1] occurring in less than 1% of patients with RA [2]. As the disease itself is asymptomatic, patients usually present with acute infection superimposed on a long-standing, erosive RA diagnosis [2].

Infective endocarditis is one of the most common life-threatening infectious syndromes [2]. We describe a rare case of infective endocarditis in a patient with a diagnosis of Felty syndrome.

Case Presentation

A 65-year-old female with a history of membranous ventricular septal defect and long-standing RA on steroids presented with pancytopenia, unexplained splenomegaly, rapidly progressive crescentic glomerulonephritis, stroke, and pulmonary embolism. During her hospital course, she developed *Streptococcus sanguinis* bacteremia, prompting an echocardiography including a transesophageal echocardiogram which revealed large vegetations on the atrial and ventricular sides of the tricuspid valve.

Her thrombocytopenia continued to worsen, eventually reaching 11,000/microliter. Due to her comorbid conditions and thrombocytopenia, the patient was not considered to be a candidate for cardiac surgery. As an alternate procedure, AngioVac thrombectomy was then performed and all vegetations were removed.

The pathology report subsequently confirmed infective endocarditis. The patient made a medical recovery following vegetation removal.

Discussion

This patient had a longstanding history of RA complicated by an acute episode of infective endocarditis with unexplained splenomegaly. Though her VSD is a risk factor, it is essential to consider an underlying disease process predisposing the patient to infective endocarditis.

Due to recurrent septic emboli to the pulmonary and systemic circulations and the persistence of the vegetation despite appropriate antibiotics, an AngioVac thrombectomy was performed [3]. The patient made a medical recovery following vegetation removal, which included weaning from renal replacement therapy with improved post-streptococcal glomerulonephritis. The patient had moderate neutropenia; neutropenia was thought to be masked in the presence of acute infection and chronic corticosteroid use. Literature supports the use of disease-modifying antirheumatic drugs (DMARDs), especially methotrexate, in the treatment of Felty syndrome [2]. The patient was treated with methotrexate which was later discontinued shortly before her bacteremia developed.

There is a lack of significant data evaluating various treatment options for this rare clinical condition. However, the therapeutics used have focused mainly on resolving neutropenia. First-line treatments most often include DMARDs, usually methotrexate, followed by granulocyte colony-stimulating factor (G-CSF) and splenectomy. Compared to uncomplicated RA, the prognosis is much poorer for these patients due to recurrent infections [2]. Infectious endocarditis portends increased morbidity and mortality as one of the most common life-threatening infectious syndromes [3].

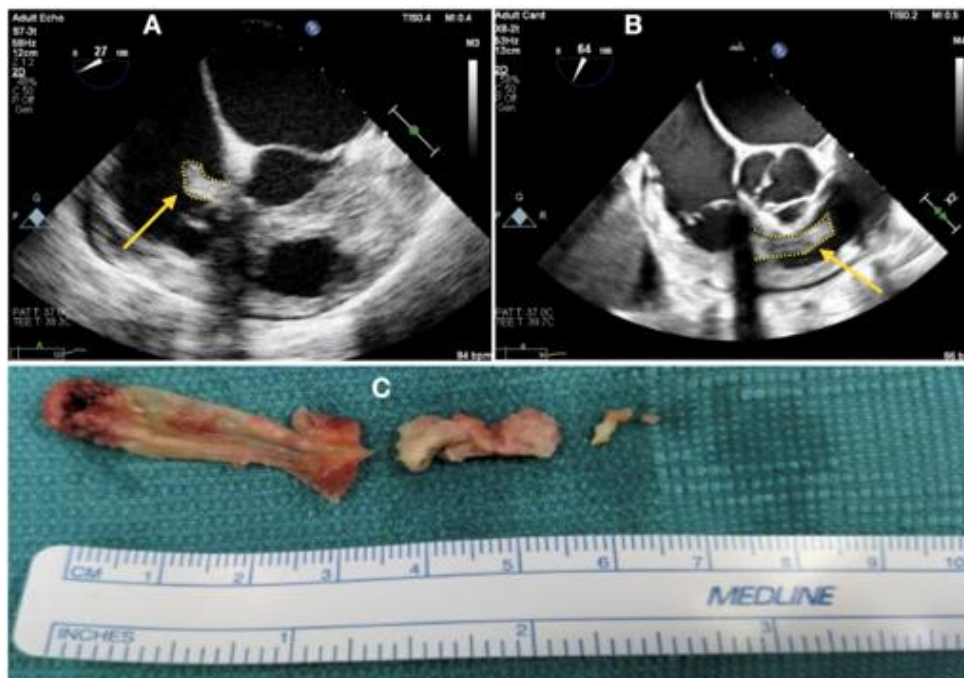


Figure 1: A: Transesophageal echocardiogram in the mid-esophageal window. The yellow arrow and dotted outline identifying infective endocarditis vegetation on the atrial aspect of the septal tricuspid leaflet. B: Transesophageal echocardiogram in the mid-esophageal window. The yellow arrow and dotted outline identifying infective endocarditis vegetation on the ventricular aspect of the septal tricuspid leaflet sub-valvular apparatus. C: Gross anatomic specimen of infective endocarditis vegetation post AngioVac removal.

Conclusion

For patients with the diagnosis of infective endocarditis, identifying an underlying infectious susceptibility can help overall patient management decisions. This is a unique case of infective endocarditis and persistent vegetation refractory to antibiotics and treated successfully by non-surgical intervention.

References

1. Owlia MB, Newman K, Akhtari M. Felty's Syndrome, Insights and Updates. *Open Rheumatol J.* 2014;8:129-36.
2. Baddour LM, Wilson WR, Bayer AS, et al. Infective Endocarditis in Adults: Diagnosis, Antimicrobial Therapy, and Management of Complications: A Scientific Statement for Healthcare Professionals From the American Heart Association. *Circulation.* 2015;132(15):1435-486.
3. Wang A, Fosbøl EL. Current recommendations and uncertainties for surgical treatment of infective endocarditis: a comparison of American and European cardiovascular guidelines. *Eur Heart J.* 2022;43(17):1617-625.