

CLINICAL VIGNETTE

Successful Management of Recurrent Pericarditis with Riloncept in a Young Adult

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Introduction

Recurrent pericarditis presents a diagnostic and therapeutic challenge due to its varied etiologies and unpredictable clinical course.¹ This 26-year-old male with recurrent pericarditis achieved sustained remission with the interleukin-1 inhibitor riloncept after failing conventional treatment.

Case Presentation

A 26-year-old previously healthy male presented with severe pleuritic chest pain since April 2020, accompanied by neck muscle tightness, night sweats, chills, and fatigue. Recurrent episodes occurred every two to three months, with exacerbations in July 2021, October 2021, January 2022, and March 2022. These were initially misdiagnosed as costochondritis and treated with short courses of Ibuprofen during the attacks. Episodes lasted for one to two weeks, and he felt back to normal between the attacks. In April 2022, he was admitted with severe pleuritic chest pain with low grade fever and extreme fatigue. Echocardiogram revealed a moderate pericardial effusion without tamponade. Treatment with prednisone, colchicine, and ibuprofen resulted in symptom improvement, but attempts to taper prednisone led to recurrence of pericarditis.

The patient also had a documented COVID-19 infection in July 2022, raising suspicion for viral pericarditis as a potential etiology. However, the onset of his pericarditis preceded the COVID-19 infection.

During subsequent admissions in October and November 2022, the patient experienced severe chest pain necessitating IV analgesics and escalated steroid doses. Additional investigations, including echocardiograms, lung CT angiograms, CT abdomen and pelvis, and laboratory tests, were unremarkable except for elevated inflammatory markers. Extensive evaluation was performed, including:

1. Electrocardiograms (EKG): Showed normal sinus rhythm with ST elevation, raising suspicion for pericarditis or early repolarization; and PR depression. (Figure 1)
2. Infection testing: COVID-19, RSV (Respiratory Syncytial Virus), influenza, EBV (Epstein Barr Virus), CMV, coxsackie, HIV (Human Immunodeficiency Virus), Coccidioides, parvovirus B19, QuantiFERON-TB Gold, hepatitis B, and hepatitis C - all negative.

3. Autoimmune rheumatologic testing: ANA (antinuclear antibody), extended ANA subtypes, vasculitis panel: -ANCA (antineutrophil cytoplasmic autoantibody), cryocrit, and sarcoidosis serologies are all negative. Familial Mediterranean fever MEFV genetics was negative.

Despite comprehensive evaluation, the exact etiology remained uncertain.

Treatment and Outcome

Anakinra, an interleukin-1 inhibitor, was initiated as his recurrent pericarditis was refractory to systemic oral steroids, colchicine and NSAIDs (Nonsteroidal anti-inflammatory drugs). He noted significant symptom improvement, and was subsequently switched to riloncept, a longer-acting interleukin-1 antagonist, due to its prolonged duration of action and ease of administration. Since initiating riloncept, the patient has experienced sustained symptom relief and successfully tapered off prednisone, colchicine, and ibuprofen. There has been no evidence of recurrent pericarditis for over a year.

Discussion

Recurrent pericarditis poses challenges in diagnosis and management, often requiring comprehensive evaluation and targeted therapy.² While infections, autoimmune and auto-inflammatory conditions, and malignancies are common etiologies, definitive diagnosis remains elusive in many cases, as illustrated by our patient. Acute pericarditis is caused by active inflammation of the pericardium, with 80% of cases idiopathic.³ In developing countries such as sub-Saharan Africa, infectious agent such as Mycobacterium Tuberculosis are responsible for most cases.³

Treatment typically involves NSAIDs, colchicine, and corticosteroids.³ However, steroid dependence and intolerance necessitate alternative therapies. Anakinra and riloncept, interleukin-1 antagonists, have emerged as promising options for recurrent pericarditis.

The AIRTRIP randomized clinical trial reported the efficacy and safety of anakinra in patients with recurrent pericarditis, with significant reductions in recurrence rates and symptom improvement compared to placebo.⁴ Similarly, the RHAPSODY clinical trial evaluated riloncept in patients with

acute pericarditis and showed a reduction in recurrent pericarditis episodes and inflammatory markers compared to placebo.⁵ Rilonacept, a dimeric fusion protein consisting of the extracellular domains of the interleukin-1 receptor components, inhibits interleukin-1 α and interleukin-1 β signaling, disrupting the inflammatory cascade implicated in pericarditis pathogenesis.⁶

Conclusion

This case report highlights the diagnostic and therapeutic challenges associated with recurrent pericarditis and the potential role of interleukin-1 inhibitors, such as anakinra and rilonacept, as effective treatment strategies in steroid-refractory or steroid-dependent cases. Rilonacept provided sustained remission and allowed for steroid tapering in this patient, underscoring its potential as a long-term therapy for challenging cases of recurrent pericarditis.

Figures

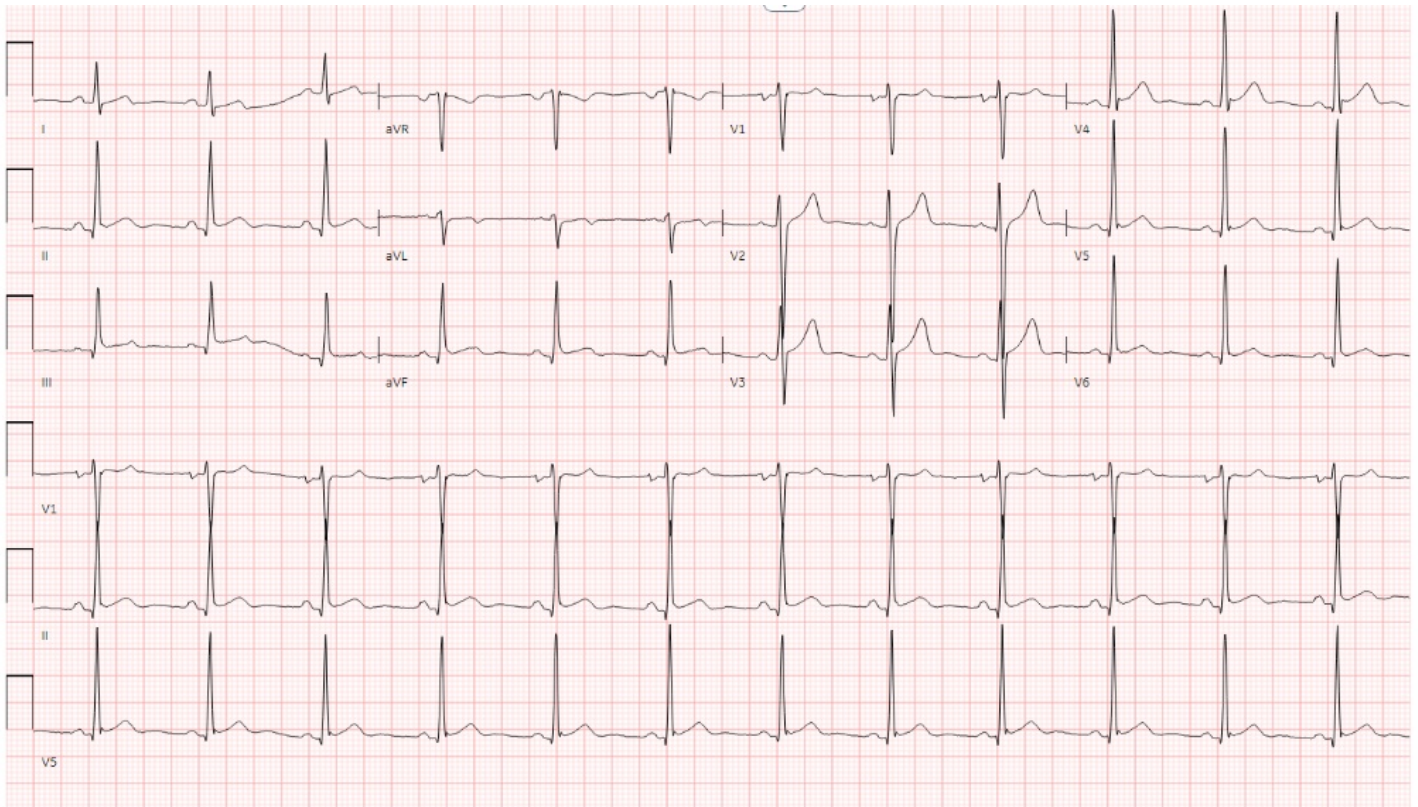


Figure 1. 12-lead EKG, Widespread concave ST elevation and PR depression throughout the limb leads (I, II, III, aVL, aVF) and precordial leads (V2-6).

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