

## IMAGING

### THE FOUR CORNERS: CLINICAL VIGNETTE CORNER

# Long-Standing Chronic Recurrent Pericarditis Managed With Advanced Diagnostic and Therapeutic Methods



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#### ABSTRACT

A 65-year-old woman with a history of idiopathic pericarditis presented with chronic recurrent pericarditis. Because of the inability to taper off anakinra without recurrent flares, she transitioned to rilonacept, which led to symptom abatement. Her positive response to rilonacept therapy correlated with an improvement in inflammatory changes noted on cardiac magnetic resonance imaging. (JACC Case Rep. 2024;29:102557) © 2024 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

A 65-year-old woman with a history of chronic recurrent idiopathic pericarditis complicated by a large pericardial effusion requiring a pericardial window and adhesiolysis presented for recurrent chest pain. Her pericarditis regimen initially included nonsteroidal anti-inflammatory drugs and colchicine and progressed to requiring prednisone, colchicine, and anakinra. The physical examination was pertinent for normal vital signs, no jugular venous distention, no Kussmaul sign, no pericardial rub or knock, regular heart rate and rhythm, and no peripheral edema.

On the initial diagnosis of pericarditis, her C-reactive protein level and erythrocyte sedimentation rate were elevated at 30.5 mg/dL (normal, <0.9 mg/dL) and 30 mm/h (normal, 0-20 mm/h, respectively). Transthoracic echocardiography (TTE) showed a moderate circumferential pericardial effusion. Cardiac magnetic resonance (CMR) showed circumferential increased pericardial signal intensity on T2 short tau inversion recovery imaging and moderate circumferential pericardial late gadolinium enhancement (LGE). Initially, she was treated with ibuprofen, colchicine, and prednisone, but she continued to experience pericarditis flares. Transitioning to anakinra resulted in improvement in both symptoms and decreased LGE on CMR. However, she was unable to taper off anakinra without pericarditis recurrences. Therefore, she transitioned to rilonacept in place of anakinra, which resulted in marked symptomatic improvement. Serial CMR showed improvement from moderate to trace LGE, and the patient was able to taper off colchicine after 1 month (**Figure 1, Supplemental Figure 1**). The patient discontinued rilonacept after 2.5 years and has not had any flares over the 4 months since discontinuation.

Recurrent pericarditis, a condition characterized by repeated episodes of pericardial inflammation, can be challenging to manage, especially when it is corticosteroid dependent and colchicine resistant. In such cases,

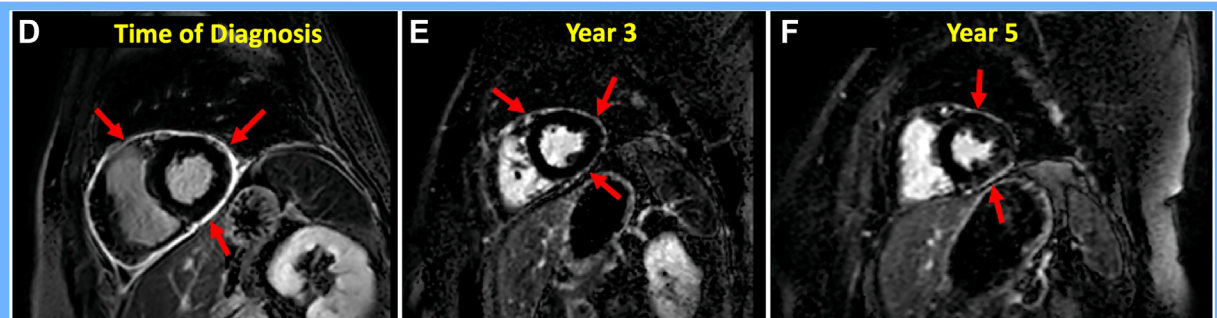
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**ABBREVIATIONS  
AND ACRONYMS****CMR** = cardiac magnetic resonance**LGE** = late gadolinium enhancement**TTE** = transthoracic echocardiography

interleukin (IL)-1 antagonists have emerged as a promising treatment option. IL-1 antagonists, such as anakinra and rilonacept, target the inflammatory cytokine IL-1 (including IL-1 $\alpha$  or IL-1 $\beta$ ), which plays a key role in the pathogenesis of recurrent pericarditis.<sup>1,2</sup> By inhibiting IL-1, these medications can effectively reduce inflammation and prevent recurrences in patients who are unable to discontinue corticosteroids and colchicine. Multimodality imaging, including both TTE and CMR, has become an essential tool in the diagnosis and management of pericarditis.<sup>3</sup> CMR provides detailed images of the heart and surrounding structures, thus allowing for accurate assessment of pericardial inflammation and fluid accumulation. In the context of recurrent pericarditis, CMR can be particularly valuable in monitoring the response to treatment. CMR-guided management strategies have been shown to lead to improved outcomes, including reduced rates of recurrence, decreased reliance on corticosteroids, and fewer pericardiocentesis procedures.<sup>3</sup> TTE is a valuable tool for assessing for pericardial effusions and guiding interventions in recurrent pericarditis. In this case, multimodality imaging was used to monitor the progression of disease and the response to advanced therapies (**Figure 1**, **Supplemental Figure 1**).

**FIGURE 1** Multimodality Imaging in Recurrent Pericarditis**Serial Transthoracic Echocardiograms****Serial Cardiac Magnetic Resonance**

- Ibuprofen + Colchicine + Prednisone
- CRP- 30.5 mg/dL, WSR- 30 mm/hr
- Anakinra and Colchicine were stopped
- CRP- 7.4 mg/dL, WSR- 41 mm/hr
- Rilonacept
- CRP- < 0.3 mg/dL, WSR- 5 mm/hr

(Top) Serial transthoracic echocardiograms demonstrating pericardial effusion (arrows), which initially resolved with initiation of anakinra, then recurred once anakinra was discontinued, and resolved again with initiation of rilonacept. (Bottom) Serial cardiac magnetic resonance imaging showing a severe late gadolinium enhancement correlating with inflammatory markers, including improvement in late gadolinium enhancement with initiation of anakinra, subsequent worsening with discontinuation of anakinra, and eventual improvement and resolution with initiation of rilonacept. CRP = C-reactive protein; WSR = Westergren method sedimentation rate.

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**APPENDIX** For a supplemental figure, please see the online version of this paper.

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**KEY WORDS** anakinra, cardiac magnetic resonance imaging, IL-1 blockers, recurrent pericarditis, rilonacept, transthoracic echocardiography