

Images in Rheumatology

Acute Perimyocarditis in a Case of Multisystem Inflammatory Syndrome in Adults

Jens T. Van Praet¹, MD, PhD, Department of Nephrology and Infectious Diseases, AZ Sint-Jan Brugge-Oostende AV, Bruges, and Department of Internal Medicine and Pediatrics, Ghent University; Pascale De Paepe, MD, Department of Pathology, AZ Sint-Jan Brugge-Oostende AV, Bruges; Levi Hoste², MD, Filomeen Haerynck², MD, PhD, Primary Immunodeficiency Research Lab, Jeffrey Modell Diagnosis and Research Center, Ghent University Hospital, and Department of Internal Medicine and Pediatrics, Ghent University, Ghent, Belgium. Address correspondence to Dr. J.T. Van Praet, Division of Nephrology and Infectious Diseases, AZ Sint-Jan Brugge-Oostende AV, Ruddershove 10, 8000 Bruges, Belgium. The authors declare no conflicts of interest relevant to this article. Written informed consent was obtained from the patient. Ethics committee approval was not requested since all elements of diagnosis and treatment belonged to standard care.

Multisystem inflammatory syndrome in adults is a rare postinfectious complication, initially reported in children developing features of Kawasaki disease and toxic shock syndrome after a SARS-CoV-2 infection. Subsequently, the clinical spectrum of the condition was recognized to be broader, defined as an inappropriate systemic inflammatory response with multi-organ dysfunction involving the skin, mucous membranes, and the heart, among other organ systems.

A 31-year-old woman without notable medical history presented at our emergency ward with fever, cough, and rapidly progressive shortness of breath. One week earlier, she had experienced an episode of cramping abdominal pain and diarrhea. On examination, tachycardia and bibasal crepitations were noted. Her electrocardiogram showed sinus tachycardia. Laboratory tests revealed markedly elevated acute-phase response (C-reactive protein 381 mg/dL and erythrocyte sedimentation rate > 120 mm/h), neutrophilia, lymphopenia, thrombocytopenia, and elevated cardiac biomarker levels (high-sensitivity troponin T [hs-TnT] 151 ng/L, and N-terminal pro B-type natriuretic peptide 25,386 pg/mL). Echocardiography documented severely depressed left ventricular function due to moderate to severe hypokinesia, suggestive of myocarditis. Minimal pericardial effusion and contrast enhancement of the pericardium but not the myocardium were seen on magnetic resonance imaging scan (Figure 1). A myocardial biopsy documented mononuclear infiltration of the myocardium, predominantly with T lymphocytes and macrophages (Figure 2).

Real-time PCR on a nasopharyngeal swab for SARS-CoV-2 was negative, but antibodies against the nucleocapsid protein of SARS-CoV-2 in the serum were detected, indicative of recent SARS-CoV-2 infection and consistent with a diagnosis of multisystem inflammatory syndrome in adults.¹ Two months earlier, the patient reported to have tested positive for SARS-CoV-2, with mild symptoms of fatigue and a common cold at that time. A multiplex cytokine panel showed very high CXCL9 and CXCL10 levels as well as signs of inflammasome activation.² Treatment with subcutaneous anakinra (100 mg daily), an interleukin-1 receptor antagonist, was initiated on day 2 of admission, with subsequent fast recovery of cardiac

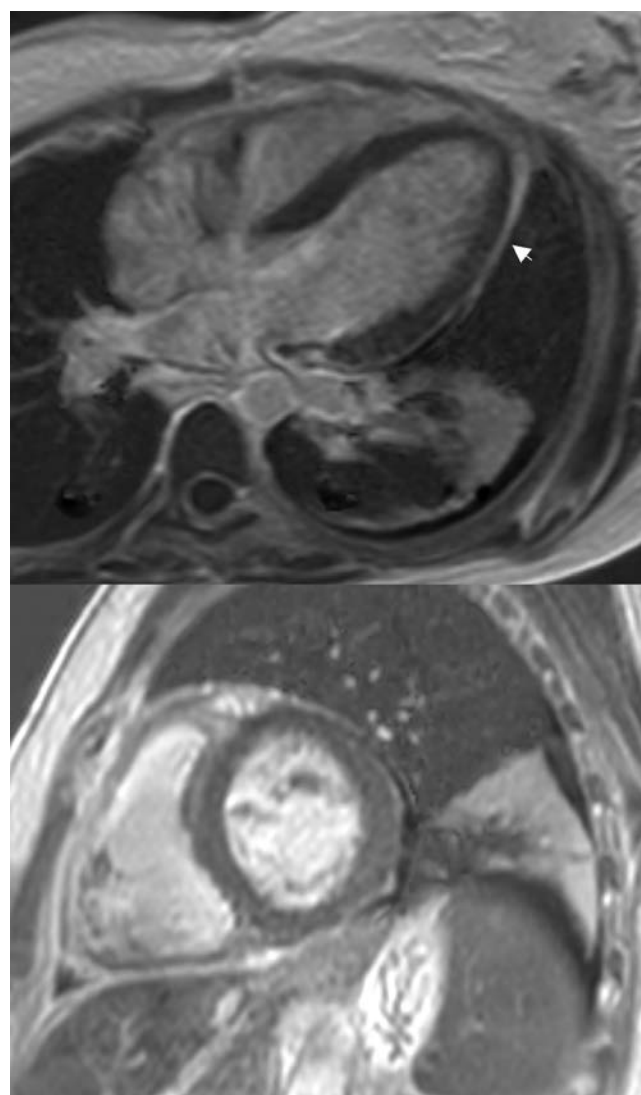


Figure 1. MRI scan of the heart shows minimal pericardial effusion and enhancement of the pericardium up to the left free ventricular wall (arrow). The upper panel is the horizontal long axis view and the lower panel the short axis view. MRI: magnetic resonance imaging.

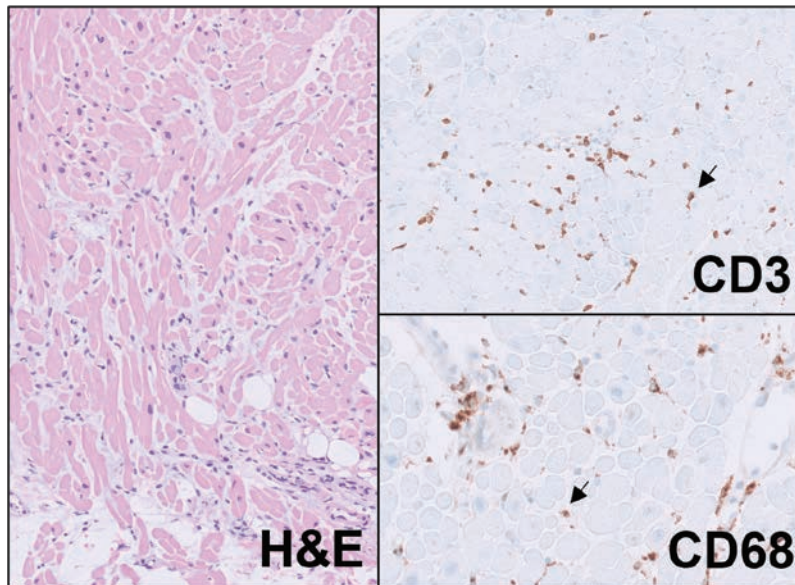


Figure 2. Myocardial biopsy shows mononuclear infiltration of the myocardium, with CD3-positive lymphocytes (arrow) and CD68-positive macrophages (arrow), without formation of granulomas (H&E stain and immunochemistry [CD3 and CD68]).

function and resolution of systemic inflammation and hs-TnT elevation.^{3,4}

ACKNOWLEDGMENT

We thank Dr. P. Debonnaire and Dr. J. Delanote for their help with the MRI images.

REFERENCES

1. Morris SB, Schwartz NG, Patel P, et al. Case series of multisystem inflammatory syndrome in adults associated with SARS-CoV-2 infection — United Kingdom and United States, March–August 2020. *MMWR Morb Mortal Wkly Rep* 2020;69:1450-6.
2. Kleiner G, Marcuzzi A, Zanin V, Monasta L, Zauli G. Cytokine levels in the serum of healthy subjects. *Mediators Inflamm* 2013;2013:434010.
3. Cattaneo P, Volpe A, Cardellino CS, et al. Multisystem inflammatory syndrome in an adult (MIS-A) successfully treated with anakinra and glucocorticoids. *Microorganisms* 2021;9:1393.
4. Bemtgen X, Klingel K, Hufnagel M, et al. Case report: lymphohistiocytic myocarditis with severe cardiogenic shock requiring mechanical cardiocirculatory support in multisystem inflammatory syndrome following SARS-CoV-2 infection. *Front Cardiovasc Med* 2021;8:716198.