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HEART FAILURE AND CARDIOMYOPATHIES

CASE REPORT: CLINICAL CASE

Myocardial Recovery After Corticosteroid Therapy in a Rheumatic Myopericarditis Pediatric Patient

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ABSTRACT

In an endemic region for acute rheumatic fever, the suspicion of myocarditis origin had also to be directed into rheumatic etiology. We present a case of a 10-year-old patient with subacute fever and myocardial systolic dysfunction (ejection fraction: 25%). One week after treatment, recovery of systolic function was observed. (JACC Case Rep. 2024;29:102694)

HISTORY OF PRESENTATION

A 10-year-old patient had a fever for 14 days, with nonspecific joint pain. The patient had slight difficulty breathing for 2 days before admission. His previous medical history was unremarkable. On physical examination, his body weight was 32 kg, blood pressure 148/105 mm Hg, pulse 100 beats/min, regular beat, and SpO2 98 % in room air. Heart and lung examination results were within normal limits

TAKE-HOME MESSAGES

- This case highlights the importance of considering rheumatic etiology as differential diagnosis of myocarditis causes, especially in endemic regions.
- The importance of early echocardiographic examination and prompt management to increase probability of myocardial reversibility is stressed.

PAST MEDICAL HISTORY

There was no remarkable medical condition.

INVESTIGATIONS

Electrocardiogram (ECG) showed sinus rhythm with normal axis with QRS rate 90, PR interval 200 ms, QRS duration 80 ms with deep T-wave inversion at precordial lead (**Figure 1**).

Blood test examination showed positive antistreptolysin O (ASO) antibody, increased erythrocyte sedimentation rate (ESR), qualitative C- reactive protein (CRP) (+), and increased troponin I and lactate dehydrogenase (LDH) level which showed myocardial tissue injury (**Table 1**). Urinalysis examination revealed hematuria and proteinuria (**Table 2**).

Echocardiography finding was left ventricle (LV) dilation (left ventricular end-diastolic volume [LVEDV] 91 mL) with reduced LV systolic function, whereas left ventricular ejection fraction (LVEF) 25% (Simpson's method), as well as LV global hypokinesia,

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ABBREVIATIONS AND ACRONYMS

ASO = antistreptolysin O

CRP = C-reactive protein

DCM = dilated cardiomyopathy

ECG = electrocardiography

ESR = erythrocyte sedimentation rate

LDH = lactate dehydrogenase

LVEF = left ventricle ejection fraction

PMP = phenoxymethylpenicillin

TAPSE = tricuspid annular plane systolic excursion

reduced right ventricular (RV) systolic function (tricuspid annular plane systolic excursion [TAPSE] 11 mm), and minimal pericardial effusion. There was no sign of valvular abnormality (**Figure 2**, Videos 1 and 2).

MANAGEMENT

This patient fulfills the revised Jones criteria for acute rheumatic fever, which underlie the carditis and glomerulonephritis complication. Patient was diagnosed with acute rheumatic fever (ARF) with myopericarditis and acute glomerulonephritis. At hospital admission, the patient was administered intravenous furosemide 2 × 30 mg, ramipril 2.5 mg once daily, bisoprolol 1.25 mg once daily, spironolactone 6.25 mg once daily for antiheart failure therapy, and prednisone 2 mg/kg body weight, divided in 4 doses a day for an anti-inflammatory regimen, as well as antibiotic therapy phenoxymethylpenicillin (PMP) 250 mg twice daily. Patient also underwent total bed rest during admission. In the subsequent day, patient symptoms were resolved. The patient was discharged after the 11th admission day, with stable hemodynamics. We continued oral medication for heart failure, prednisone, and oral PMP antibiotic therapy as well. The prednisone dose was down-titrated gradually, according to guidelines.

OUTCOME AND FOLLOW-UP

In outpatient clinic visits, after a week postdischarge evaluation, we found improvement in clinical status with LV systolic function recovery, which showed in improvement of LVEF to 62% (Simpson's method) and RV systolic function (TAPSE 18 mm), and abatement of pericardial effusion (**Figure 3**, Videos 3 and 4). We continued the heart failure and PMP therapy because of its rheumatic etiology of myocarditis with down-titrated prednisone dose.

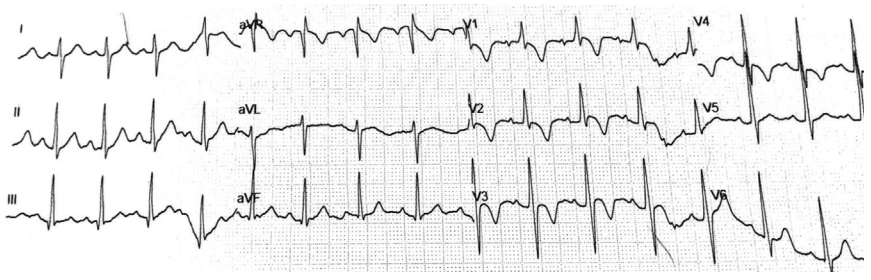
DISCUSSION

The most common cause of pediatric myocarditis is viral myocarditis. However, other etiologies including autoimmune, medication-related, hypersensitivity reactions, and toxins are also established.[1] Moreover, in an ARF endemic region such as Indonesia, it should be considered for an etiology in patients with myocarditis clinical presentation. In 2015, Indonesia placed fourth for the world's largest estimated number of cases of rheumatic heart disease (1.18 million), below India (13.17 million), China (7.07 million), and Pakistan (2.25 million).[2]

Carditis, as one of the major criteria for ARF, has become an important aspect for evaluation[3,4] and comprises primarily valvulitis, which involves endocardium and valvular tissue, but in rare situations myocarditis and pericarditis could ensue.[5] These phenomena could raise suspicion among clinicians for patients with subacute fevers and cardiac specific symptoms such as dyspnea, chest pain, or hypertension.[5,6] Our case showed myopericardial involvement, which was shown by echocardiography by reduced ventricular systolic function and the presence of pericardial effusion. In addition, increased serum troponin and LDH levels describe significant myocardial injury.

Echocardiography is a first-line imaging modality to detect cardiac involvement in any suspicious subacute fever with dyspnea and hypertension.[5-7] The purpose is to assess left ventricular structure, wall-motion abnormalities, regional or global ventricular dysfunction, valvular insufficiency, and pericardial effusion, as these abnormalities are often present in acute myocardial inflammation,[6,7] and also for the evaluation aspect as well.[1]

FIGURE 1 Patient's 12-Lead Electrocardiography



Echocardiography criteria for cardiac involvement of ARF is well described in several guidelines including mitral and aortic valve changes.[3,4] Our patient met the Jones criteria for ARF from the qualitative ASO assay; major criterion is carditis and minor criteria are increase in erythrocyte sedimentation rate, positive CRP, and fever. The patient also showed signs of glomerulonephritis on urinalysis. From these greater pictures, it seems that, in addition to the myopericarditis complication of ARF, the patient also had acute glomerulonephritis, which, on one hand, worsened the prognosis but, on the other hand, provided additional evidence of rheumatic process as myopericarditis etiology.

Echocardiography also has an evaluation role as well. As we can see in our patient, after the 1-week postdischarge evaluation, echocardiography showed significant recovery of LV and RV systolic function. These phenomena describe attenuation of myocardial inflammation and the reversibility of myocardial systolic function. Myocardial recovery in myocarditis depicts a good prognosis.[1,6]

The conflicting data concerning ARF carditis criteria in our patient is about the involved tissue that affected whereas only myocardium and pericardium that is affected without endocardium or valvulitis involvement. It is widely known that carditis of ARF has been considered to be pancarditis and can involve the endocardium, myocardium, and pericardium tissue; however, valvulitis is by far the most specific feature of ARF, and isolated pericarditis or myocarditis should rarely, if ever, be considered rheumatic as an etiology.[5] In this case, myopericardial involvement can be considered as rare carditis manifestation in ARF, or it could be a concomitant infectious myocarditis from viral etiology.

A study including 222 pediatric patients with dilated cardiomyopathy (DCM) caused by myocarditis revealed a 5-year rate of freedom from transplantation of 81% and an estimated survival of 92% at 1 year and 90% at 2 and 5 years.[7] According to a Pediatric Cardiomyopathy Registry study, most patients showed recovery of ventricular size and systolic function within several months.[8] After myocardial inflammation subsides, heart recovery will take place. However, signs of persistent inflammation can lead to continuous adverse myocardial remodeling and DCM. Depressed LVEF is associated with worse outcomes and increased risk of death.[1,9] Our case showed recovery of ventricular systolic function at 1-week follow-up, which showed a good prognosis.

The myocardial inflammation could lead to myocardial fibrosis and culminate in DCM.[1] Considering that rheumatic heart disease is an important preventable cause of cardiovascular death and disability, this warrants prompt diagnosis and treatment to prevent the progression to DCM, the condition that is associated with worse prognoses.[1,2,7]

TABLE 1 Laboratory Blood Test Examination Result

Blood Test	Result	Reference Range
Erythrocyte sedimentation rate (ESR) mm/h	104[a]	<15
Qualitative C-reactive protein	Positive[a]	Negative
Troponin I level (ng/mL)	0.30[a]	<0.02
Qualitative antistreptolysin antibody (ASO)	Positive[a]	Negative
Serum creatinine (mg/dL)	0.9	0.3-0.9
Blood urea nitrogen (BUN) (mg/dL)	20	6-24
SGOT (U/L)	15	5-40
SGPT (U/L)	5	7-56
Lactate dehydrogenase (LDH) U/L	439[a]	143-370

[a]Abnormal result compared to reference range.

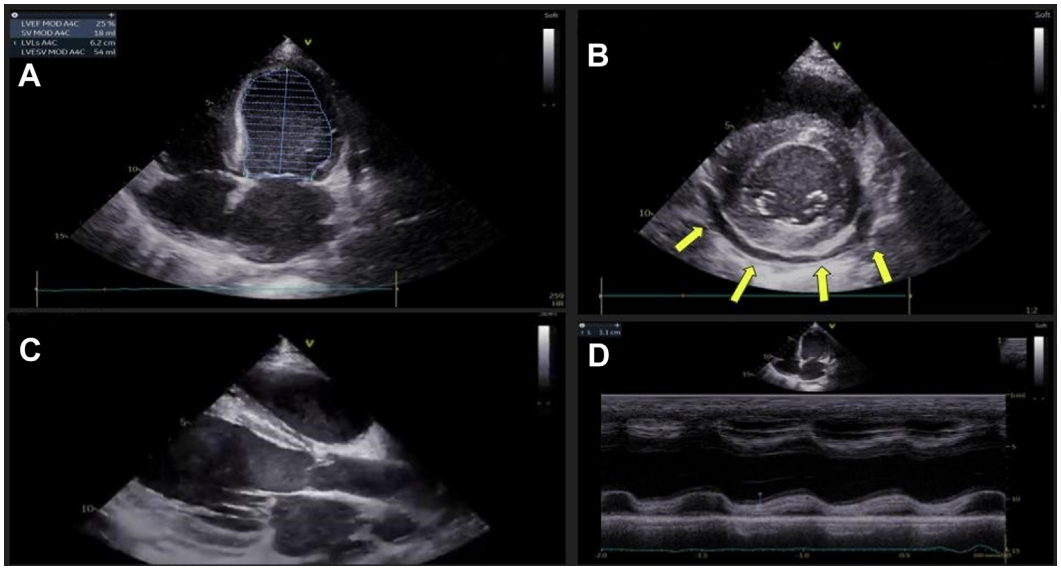
Treatment of myocarditis varies depending on the clinical severity and stage of illness.[1,7] Corticosteroids have potent anti-inflammatory effects.[6] This therapy should be commenced as soon as possible in the subacute process or autoimmune phase to attenuate further myocardial injury, to increase the possibility of recovery, and to prevent deterioration into DCM.[7] Studies in the adult population by Mason et al[10] did not show improvements in echocardiography or survival. However, Frustaci et al[11] showed LV systolic function recovery in 38 of 43 patients treated with prednisone and azathioprine compared with 0 of 42 in the placebo group in biopsy-proven myocarditis without persistence of myocardial viral genome. These results support that immunosuppression is effective for the autoimmune phase of myocarditis without active viral infection in adults.[6] In the recent case, we showed that corticosteroid therapy in the autoimmune phase is effective to improve ventricular systolic function significantly.

Therapy after absence of both infectious and autoimmune stages is primarily targeted on prevention and reversal of the myocardial remodeling process and reduction of hemodynamic stress by using

TABLE 2 Urinalysis Examination Result

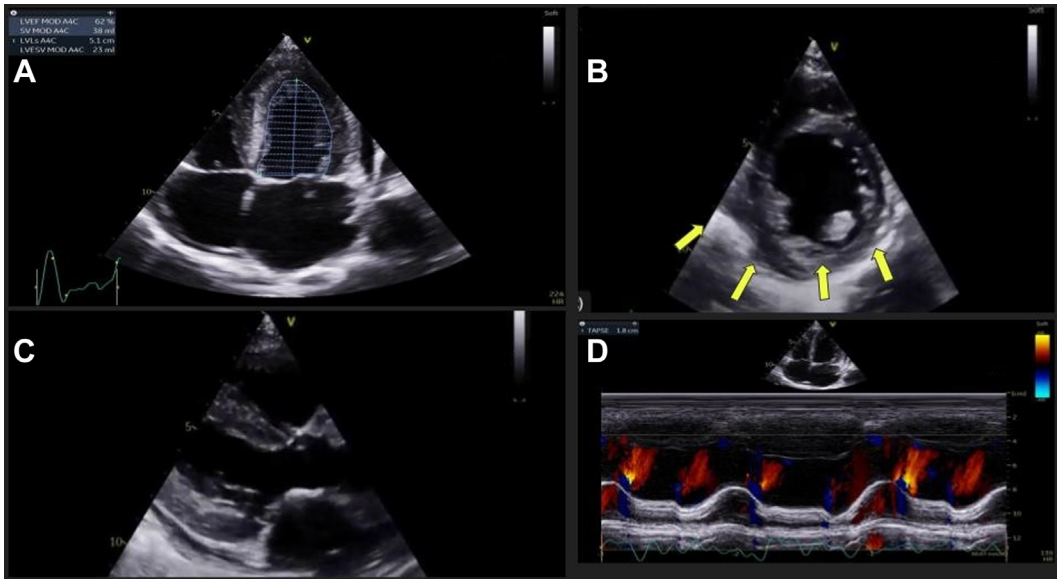
	Result	Reference Range
pH	5.50	4.6-8.5
Blood	3+	Negative
Protein	3+	Negative
Leukocyte esterase	3+	Negative
Bilirubin	Negative	Negative
Urobilinogen	Normal	Normal
Leukocyte	4+	1+
Erythrocyte	4+	1+

FIGURE 2 **Patient's Baseline Echocardiography Examination**



(A) LVEF assessment by Simpson's 4-chamber method. (B) Parasternal short axis view in mid LV level shown pericardial involvement with pericardial fluid (yellow arrow). (C) Parasternal long axis view. (D) TAPSE assessment. LVEF = left ventricular ejection fraction; LV = left ventricle; TAPSE = tricuspid annular plane systolic excursion.

FIGURE 3 **Patient's Echocardiographic Evaluation**



(A) LVEF assessment by Simpson's 4-chamber method. (B) Parasternal short axis view in mid-LV level shown resolution of pericardial fluid (yellow arrow). (C) Parasternal long axis view. (D) TAPSE assessment. Abbreviations as in Figure 2.

angiotensin converting enzyme (ACE) inhibitors, beta blockers, and spironolactone.[1,7] Finally, considering the rheumatic origin for myocarditis etiology, we administered PMP for pediatric therapeutic dose followed by long-term prophylactic doses to prevent recurrent rheumatic activation, according to guidelines.[3,4,12]

CONCLUSIONS

We presented a 10-year-old patient with a case of myopericarditis from rheumatic origin as an etiology. After a period of corticosteroid therapy, the patient underwent significant recovery of myocardial systolic function based on echocardiographic evaluation.

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KEY WORDS corticosteroid therapy, echocardiography, pericarditis, rheumatic myocarditis

APPENDIX For supplemental videos, please see the online version of this paper.