Case Report

A rare case of Systemic Lupus Erythematosus presenting as acute myopericarditis with bilateral pleural effusions.

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Abstract

Acute myopericarditis is a rare presentation in clinical practice with multiple aetiologies. Eventhough cardiac manifestations are known to be present in up to 50% of Systemic Lupus Erythematosus (SLE) patients, acute myopericarditis is an uncommon presentation, occurring in up to 1% of patients. Here we report a patient who presented with fever and pleuritic type chest pain and was managed as acute myopericarditis with bilateral exudative pleural effusions and later diagnosed to have SLE. The patients were initially treated with nonsteroidal anti-inflammatory drugs and later with steroids and hydroxychloroquine. Early diagnosis of myopericarditis and identification of the aetiology is essential to halt the progression of disease. Pericarditis due to Tuberculosis need to be excluded before starting steroids in the Sri Lankan setting.

Keywords: Acute myopericarditis, Systemic lupus erythematosus, Bilateral pleural effusions

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Introduction

Acute myopericarditis is a rare presentation in clinical practice with multiple aetiologies including infectious causes, malignancy and autoimmune causes such as Systemic Lupus Erythematosus (SLE). Even though studies show that 20% to 50% of patients with SLE will develop cardiac manifestations [1], acute pericarditis is an uncommon presentation, occurring in up to 1% of patients [2].

Here we report a patient who presented with fever and pleuritic type chest pain and was managed as acute myopericarditis and later found to have SLE.

Case Report

A 22-year-old male from Sri Lanka presented with fever and pleuritic type left sided chest pain for one day duration. Three months back he had developed right elbow joint pain for two weeks. The pain was constant throughout the day (no early morning stiffness). There was no swelling, warmth, or tenderness of the elbow joint. He had a history of chills at night for two months but there was no documented fever. There were no urinary or respiratory symptoms.

There was no significant past medical, surgical or allergy history. There was no history to suggest

exposure to leptospirosis and there was no history of intravenous drug abuse or sexual promiscuity.

On examination the patient was febrile with a blood pressure of 110/80 mmHg and a pulse rate of 120 beats per minute. His abdomen was soft, and lungs were clear. There were no murmurs nor pericardial friction rub. Pulse oximetry showed an oxygen saturation of 98 % on room air. The ECG taken on admission showed concave shaped ST Elevations V1-V5 with sinus tachycardia and the high sensitive troponin I was negative.

On day two of admission a pericardial rub could be heard and the high sensitive troponin I was positive (Table 1). Two-dimensional echocardiography showed a pericardial effusion which was mainly extending posteriorly and laterally, but minimal anteriorly and not significant enough for aspiration. On day three of admission the patient ingested 200 ml of "Detol" (Chloroxylenol). On further questioning third person auditory hallucinations and persecutory delusions were elicited which resolved completely 12 hours later. Chest Xray and ultrasound scan of chest showed bilateral pleural effusion. Pleural fluid analysis revealed an exudative effusion (Table 1). The two-dimensional echocardiogram was repeated on day four of admission and showed a pericardial effusion which was minimal anteriorly, 17mm laterally, 12mm inferiorly and 14 mm posteriorly. There was no right atrium nor right ventricular collapse and the ejection fraction was more than 60 %. The patient was started on ibuprofen 200 mg three times daily for symptomatic relief.

The serum antinuclear antibody (ANA) was positive (>1:80 titer) but double-stranded DNA was negative. Since financial restraints prevented the autoimmune panel from being done immediately, a multidisciplinary meeting was held. The multidisciplinary team's main concern was to exclude tuberculosis. They decided to go ahead with a pericardial biopsy with pericardial fluid aspiration. The pericardial biopsy was to be sent for histology, tuberculosis gene expert – ultra sensitive, and tuberculosis liquid culture. The pericardial fluid was to be sent for full report and tuberculosis gene expert.

However before sending the patient for the pericardial biopsy, the consultant cardiologist decided to repeat the echocardiogram, and the repeat echocardiogram showed that the pericardial effusion was no longer present. This was an unexpected finding. This echocardiogram was done ten days after starting the patient on ibuprofen. There are no studies or case reports of pericardial effusions in SLE being treated with only non-steroidal anti-inflammatory medication, though it is known that a majority resolve quickly with steroids and anti-rheumatologic drugs. However the tachycardia persisted for 2 weeks (after starting prednisolone), which could indicate that the cardiac involvement was not completely resolved.

As the pericardial effusion was no longer present, the decision was taken to withhold the pericardial biopsy. Later in the day the autoimmune panel results were available and showed a strongly positive anti-smith(sm) antibody titer, a strongly positive Ribonucleoprotein (RNP)/Sm antibody titer and strongly positive Ro-52 antibody titer (Table 2). A diagnosis of SLE was made under the 2019 European League Against Rheumatism (EULAR) and the American College of Rheumatology (ACR) classification criteria for SLE as there was fever, acute pericarditis, positive anti smith antibody titer, and a positive ANA titer of > 1:80.

Discussion

This case describes a patient presenting with acute myopericarditis and bilateral pleural effusions, and later diagnosed to have SLE. Acute myopericarditis is a rare presentation in clinical practice and an uncommon presentation of SLE. In fact, there are only a few case reports of SLE presenting as myopericarditis worldwide. In this case, the clinicians were faced with the additional difficulty of diagnosing SLE as the patient didn't fit the prototypical patient picture and financial constraints delayed the testing for autoimmune antibodies.

The term myopericarditis is used when there is cardiac involvement determined by abnormal cardiac enzymes and imaging demonstrating myocardial involvement, change in baseline cardiac function, or wall motion abnormalities [3] . In clinical practice both pericarditis and myocarditis coexist because they share common etiologic agents, mainly cardiotropic viruses and up to 15% of cases of acute pericarditis have a significant myocardial involvement [3,4] .In myopericarditis pericardial effusion is less common than in acute pericarditis [4].

The use of anti-double stranded DNA and anti-Smith antibodies is highly specific for SLE and seen in approximately 70% and 30% of patients, respectively [5].In a multicenter, cross-sectional study with a study population of 593 subjects, it was reported that a positive ANA in combination with anti-dsDNA resulted in a sensitivity of 84.5% and specificity of 90.7% in diagnosing SLE from other rheumatic diseases [6] .

Table 1 : The biochemical parameters of the patient during hospital stay.

Test	Normal Range	04/02/23	09/02/23	18/02/23	27/02/23
Full Blood Count					
WBC ($\times10^9$/ml)	4.0-10.0	14.57	13.01	10	5.67
Neutrophils ($\times10^9$/ml)	2.0-7.0	12.11	10.76	7.59	3.4
Lymphocytes ($\times10^9$/ml)	1.0-3.0	0.73	1.18	1.09	1.21
Neutrophils %	50--70	83.2	82.6	75.8	60
Lymphocytes%	20-40	5	9.1	10.9	21
Monocytes ($\times10^9$/ml)	0.2-1.0	1.7	1.0		1.01
Eosinophils ($\times10^9$/ml)	0.02-0.5	0.01	0.05		0.02
Basophils ($\times10^9$/ml)	0.02-0.1	0.02	0.02		0.03
Monocytes %	3.0-12.0	11.7	7.7		17.9
Eosinophils %	0.5-5.0	0.0	0.4		0.3
Basophils %	0.0-1.0	0.1	0.2		0.5
RBC ($\times10^{12}$/ml)	4.5-5.5	4.12	4.22	3.92	3.63
Haemoglobin (g/dl)	12.0-16.0	12.5	12.3	11.5	10.6
HCT (%)	37-54	38.9	12.3	36.6	29.7
MCV (fL)	83-101	95.5	91.2		81.8
MCH (pg)	27-32	30.3	29		29.2
MCHC (g/dl)	31-34	32.1	31.9		35.7
RDW- SD (fl)	11.6-14.0	46	45.1		44
RDW CV (%)	39.0-46.0	13	13.5		13,2
Platelets ($\times10^9$)	150-400	286	444	679	485
Renal Function Tests					
Sodium (mmol/L)	136-146	129		136	135
Potassium (mmol/L)	3.5-5.1	4		3.1	3.3
Urea (mmol/L)	2.8-7.2	3.6		3.3	2.3
Creatinine (µmol/L)	74-110	96		91	74.8
Albumin (g/L)	35-52		38.7	33.7	
Corrected calcium (mmol/L)	2.062-2.60		2.26		
Liver Enzymes and Liver Function Tests					
AST (U/L)	<50	12		20	15
ALT (U/L)	<50	9.6		21	19.2
Total Protein (g/L)	66-83				
Albumin (g/L)	35-52		24.6	26.9	
Globulin (g/L)	25-35		44.52	28.11	
Total Bilirubin (µmol/L)	5.0-21	15.47		7.1	5.6
Direct Bilirubin (µmol/L)	0.0-3.4	6.7			2.8
Direct Antiglobulin Test (DAT)		Negative			
Inflammatory markers					
CRP		128	310	28	10
ESR		60	70	70	50
RBS		102			
HS Trop I	<0.0342	0.0342	1.82	<0.001	
Creatinine Kinase	<171				
TSH	0.5-4.7		1.39		
Blood Culture		Negative	Negative	+Pseudomonas	
Urine Culture		Negative	Negative	Negative	
Pleural fluid analysis					
Colour	Straw coloured				
Protein (mg/L)	51 (Serum: 77)				
ADA (35)(U/L)	34.9				
Tb Gene Expert	Negative				
Other Tests to exclude tuberculosis					
Mantoux	Negative (0)				
Sputum AFB x 3	Negative				
Sputum Gene Expert	Negative				

WBC: White Blood Cells, RBC : Red Blood Cells, HCT: Haemoatocrit, MCV: Mean Corpuscular Volume, MCHC : Mean Corpuscular Haemoglobin Concentration, AST : Aspartate aminotransferase, ALT: Alanine aminotransferase, INR : International Normalization Ratio, APTT : Activated partial thromboplastin time, CRP : C Reactive Protein, ESR: Erythrocyte Sedimentary Rate, RBS: Random Blood Sugar, HS Trop I : High Sensitive Troponin , ADA : Adenosine deaminase, Tb : Tuberculosis.

In the presence of a positive ANA titer and acute pericarditis, it is important to consider other differentials such as Sjogren's syndrome [7] and malignancy such as cardiac lymphoma [8] . Another important differential that must be excluded is tuberculosis which is responsible for approximately 70% of cases of large pericardial effusion and most cases of constrictive pericarditis in developing countries. A definite or proven diagnosis is based on demonstration of tubercle bacilli in pericardial fluid or on histologic section of the pericardium. A probable diagnosis is based on proof of tuberculosis elsewhere in a patient with otherwise unexplained pericarditis, a lymphocytic pericardial exudate with elevated biomarkers of tuberculous infection, and/or appropriate response to a trial of antituberculosis chemotherapy [9]. Exclusion of tuberculosis is especially crucial before starting steroids and immunosuppressants for autoimmune diseases. Studies have shown that Anti-Ro -52 antibodies are associated with long QT syndrome and congenital heart block (due to transplacental passage of the antibodies from mother to foetus) [10]. However, our patient had no arrhythmias and there is no proven associated between pericarditis and Anti-Ro -52 antibodies.

Nonsteroidal anti-inflammatory drugs are the primary choice in the management of pericarditis. However, SLE treatment includes disease-modifying anti-rheumatologic drugs, glucocorticoids, and even immunomodulators [11] .In our case the patient was started on oral prednisolone 30 mg mane and oral hydroxychloroquine 200 mg nocte. Ivabradine 5 mg nocte was given to control the heart rate. The patient's chest pain resolved within 2 days and heart rate reduced to 100 beats per minute. The chest x- ray and ultrasound scan of chest done after one month showed that the pleural effusions were minimal and the 2-dimensional echo was normal. Ivabradine was tailed off as his pulse rate dropped to 76 beats per minute and the ST elevations of the ECG were completely resolved. The prednisolone dose was gradually reduced and at the end of 3 months post admission, he was on prednisolone 5mg mane.

Conclusion

This case describes a patient who presented with acute myopericarditis with bilateral exudative pleural effusions who was later diagnosed with SLE. Though cardiac manifestations of the disease are commonly reported in SLE patients, it is rare on presentation. In fact, SLE presenting with myopericarditis is yet to be reported in Sri Lanka. Another interesting fact is that the pericardial effusion disappeared after ten days of Ibuprofen and before the introduction of steroids.

While acute pericarditis is diagnosed in the acute setting and treated with anti-inflammatory agents, further investigations are warranted as it is crucial to diagnose the aeteology. Pericarditis due to tuberculosis need to be excluded before starting steroids in the Sri Lankan setting.

Table 2 : Autoimmune pannel and other serum investigations

Autoimmune	
ANA -Nuclear pattern	Positive > 1: 80
ANA Cytoplasmic pattern	Negative
ANA Mitotic pattern	Negative
Double Stranded DNA	Negative
Anti Smith Antibody	Strongly Positive
RNP/Sm Antibody	Strongly Positive
SS-A/Ro Antibody	Negative
Ro-52 Antibody	Strongly Positive
SSB/La Antibody	Negative
Scl 70 Antibody	Negative
Anti mitochondrial (m2) Antibody	Borderline
C3	Normal
C4	Normal
Rheumatoid Factor	Positive
ASOT	Negative

Serum	
VDRL	Negative
HIV I,II Antibody	Negative
Hepatitis C Antibody	Negative
EBV IgG	Positive
EBV IgM	Negative
Zika IgM Elisa	Negative
Chickungunya IgM(ICT)	Negative
Dengue RT-PCT	Negative

VDRL: Venerology Disease Research Laboratory, HIV : Human Immunodeficiency Virus, ANA : Antinuclear Antibody, ANCA : Antineutrophilic Cytoplasmic antibody.

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Conflict of interest

The Authors declare that there is no conflict of interest.

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