Case report: A rare presentation of cardiac sarcoidosis with recurrent large pericardial effusion and stress-induced cardiomyopathy

Ayman Naguib[1], Riyadh Qasim[1], Khalid Alkatout[2], and Fatema Qaddoura[1]

[1]King Fahd Military Medical Complex
[2]Imam Abdulrahman Bin Faisal University College of Medicine

April 16, 2024

Abstract

A 65-year-old female known to have type 2 diabetes mellitus, with no significant past medical history, had presented to another hospital with progressive dyspnea, orthopnea, lower limb edema in the two weeks preceding the reporting of this case. Her echocardiography revealed large pericardial effusion with echocardiographic signs of increased intrapericardial pressure. An emergency pericardiocentesis was performed. One week afterwards, she was admitted to the authors' hospital because of re-accumulation of her large pericardial effusion, which mandated another pericardiocentesis. A CT of the chest revealed enlarged mediastinal lymph nodes (LNs). Video-assisted thoracic surgery (VATS) was done to obtain a pericardial window and a biopsy from the mediastinal LN, which revealed non-caseating granuloma, highly suggestive of sarcoidosis. A few days after surgery she experienced a neurogenic bladder, acute renal shutdown, and metabolic acidosis, during which she developed stress-induced cardiomyopathy that improved a few days later. The patient's symptoms improved after receiving corticosteroids.

Introduction:

Sarcoidosis is a granulomatous disease of unknown etiology, the pathological hallmark of which is noncaseating granulomas. It can affect various organs including the heart.[1] Cardiac involvement by sarcoidosis (CS) can affect any portion of the heart, including the pericardium, atria, ventricles, papillary muscles, and valves. Clinical presentation of CS varies widely, from being asymptomatic, to decompensated heart failure, heart block, and malignant arrhythmias.[2] CS has various phenotypes in cardiac imaging with either ventricular septum thinning, especially in the basal portion, ventricular wall thickening, dilatation, regional wall motion abnormalities, valvular dysfunction or, rarely, pericardial effusion. This makes the diagnosis of CS challenging in many clinical instances.[3]

History of presentation:

A 65-year-old female known to have type 2 diabetes mellitus, on oral hypoglycemic medications with no other significant past medical history, presented with two weeks' history of gradual onset and progressive course dyspnea, orthopnea, dry cough, and lower limb edema, as well as reduced appetite and lethargy. She had no other cardiac complaints and was admitted to another hospital. Clinically, the patient had signs of cardiac tamponade and echocardiography revealed large pericardial effusion with echocardiographic signs of increased intrapericardial pressure. Emergency pericardiocentesis was done and she was discharged after three days following improvement of her symptoms and follow-up echocardiography which revealed minimal pericardial effusion. One week later, she presented to the authors' hospital with recurrence of her initial symptoms. Her physical examination was remarkable for a low body weight female lying in a semi-seated position, with respiratory distress, tachypnea, tachycardia (HR: 120 b/m), hypotensive (BP 80/40 mmHg),

and engorged jugular veins. Local cardiac examination revealed distant heart sounds and decreased breath sounds at both lung bases. Urgent echocardiography revealed large circumferential echocardiography with echocardiographic signs of cardiac tamponade. Urgent pericardiocentesis with a drain of approximately 1.5 L was done.

Investigations:

ECG showed sinus tachycardia, left axis deviation, low voltage, and poor R wave progression on precordial leads. **Initial echocardiography** showed normal LV systolic function, large pericardial effusion with signs of increased intra-pericardial pressure (Videos 1–2, Figures 1–2). **Pericardial fluid analysis** showed clear serous fluid, where cells were mainly lymphocytes, remarkable exudative effusion, with no malignant cells, and negative for AFB and TB PCR. The**CT coronary angiogram** showed non-significant coronary artery disease. **CT of the chest** showed bilateral symmetrical hilar mediastinal lymphadenopathy and bilateral pleural effusion. **LN biopsy** showed a picture of non-necrotising granulomatous lymphadenitis.**CMR** was done after three months from acute illness, due to unavailability at time of hospitalisation as a result of the COVID-19 pandemic. This showed diffuse patchy epicardial late gadolinium enhancement at basal to mid inferior, inferolateral, and lateral myocardial walls with patchy mesocardium at basal septum.

Differential diagnosis:

Myopericarditis, Cardiac sarcoidosis, tuberculosis, lymphoma

Management:

After the second pericardiocentesis and after reviewing the findings of the chest CT with the thoracic surgeon in a multidisciplinary team discussion, decision was made to undertake VATS. A biopsy was taken from the mediastinal LN and from the pericardium; a pericardial window was also performed. The patient was then started on oral corticosteroid therapy 25 mg OD, NSAID, colchicine and was discharged home to be followed up as an outpatient.

However, five days after discharge, she presented with anuria and shortness of breath. Her initial laboratory work-up revealed severe metabolic acidosis and acute renal injury. She was readmitted to ICU. A urogenital ultrasound did not reveal any obstruction. The urology team's assessment was that the underlying cause was acute urine retention, secondary to urogenic autonomic bladder. A urinary catheter was placed and her condition improved.

During the ICU admission because of acute kidney injury :

Echocardiography revealed mild left ventricular systolic dysfunction and new regional wall motion abnormalities in the form of hypokinesia and ballooning of the apical segments, other segments were hyperkinetic (Figure 3, Videos 3–5).

Few days after resolution of metabolic acidosis and normalisation of kidney function tests , the echo was repeated and revealed normal left ventricular systolic function and normal regional wall motion (Videos 6–7).

Discussion:

This report is presenting a rare presentation of cardiac sarcoidosis with recurrent large pericardial effusion causing tamponade. Echo played an important role in establishing this diagnosis of large pericardial effusion; the significant respiratory variations in PWD over tricuspid and mitral valves provided an indication of increased intrapericardial pressure and, in the clinical context of distress and hemodynamic instability, mandated the rapid action of pericardiocentesis. The recurrence of the large pericardial effusion one week afterwards is also a rare finding, reported in very few case reports.[4,5] The search for the specific aetiology of pericardial effusion in our patient was negative, according to laboratory results. During the patient's clinical course, she developed acute renal shut down and severe metabolic acidosis; echocardiography showed a picture of stress-induced cardiomyopathy with transient ballooning of the apical segments, which improved

to normal two days afterwards. This is rare and, to our knowledge, has not before been reported in such a clinical context. In our case, the presence of mediastinal lymphadenopathy and histopathology of the LN biopsy was a major factor in establishing the diagnosis. The absence of significant coronary artery disease by cardiac CT helped us to rule out underlying coronary artery disease. The CMR findings were non-specific; this could be attributed to the lateness (after three months), or due to the nature of the disease and heterogenicity of the LGE affection pattern.[6,7]

Follow-up:

During her clinical follow-up two months later, the patient showed good improvement of her symptoms and complete recovery of acute renal failure. She was on steroids for two months, which was gradually reduced over four weeks, and she remained symptom-free. She also showed elevated blood glucose levels, for which she was started on insulin.

Conclusion:

Pericardial effusion is a rare presentation of cardiac sarcoidosis and can be not only large enough to cause cardiac tamponade, but also recurrent. Stress-induced cardiomyopathy occurring in such a clinical context has not previously been reported, to the best of our knowledge.

Learning objectives:

1. To recognise recurrent pericardial effusion as a rare presentation of cardiac sarcoidosis.
2. To report the occurrence of stress-induced cardiomyopathy, which occurred in a complex clinical situation.
3. To highlight the demonstration of non-caseating granuloma on biopsy, either cardiac or extra-cardiac, as an important diagnostic step of cardiac sarcoidosis.

References:

1. Birnie DH, Nery PB, Ha AC, Beanlands RS. Cardiac sarcoidosis. Journal of the American College of Cardiology. 2016 Jul 26;68(4):411-21. https://doi.org/10.1016/j.jacc.2016.03.605
2. Birnie DH, Sauer WH, Bogun F, et al. HRS expert consensus statement on the diagnosis and management of arrhythmias associated with cardiac sarcoidosis. Heart rhythm. 2014 Jul 1;11(7):1304-23. https://doi.org/10.1016/j.hrthm.2014.03.043
3. Terasaki F, Azuma A, Anzai T, et al. JCS 2016 Guideline on diagnosis and treatment of cardiac sarcoidosis–Digest version–. Circulation Journal. 2019 Oct 25;83(11):2329-88. https://doi.org/10.1253/circj.CJ-19-0508
4. Whiteside H, Jyothidasan A, Sorrentino R, Patel V, Sharma G. Cardiac sarcoidosis: an unusual cause of recurrent large pericardial effusion. Journal of the American College of Cardiology. 2016 Apr 5;67(13S):1073-. https://www.jacc.org/doi/full/10.1016/S0735-1097%2816%2931074-9
5. Navaneethan, SD, Venkatesh S, Shrivastava R, Mehta J, and Israel R. "Recurrent pleural and pericardial effusions due to sarcoidosis." *PLoS Med 2* , no. 3 (2005): e63. https://doi.org/10.1371/journal.pmed.0020063
6. Okada DR, Bravo PE, Vita T, et al. Isolated cardiac sarcoidosis: a focused review of an under-recognized entity. Journal of Nuclear Cardiology. 2018 Aug;25(4):1136-46. https://doi.org/10.1007/s12350-016-0658-1

7- Smedema JP, Van Kroonenburgh MJ, Snoep G, Backes W, Gorgels AP. Images in cardiovascular medicine. Cardiac sarcoidosis in a patient with hypertrophic cardiomyopathy demonstrated by magnetic resonance imaging and single photon emission computed tomography dual-isotope scintigraphy. Circulation. 2004 Dec 1;110(24):e529-31. https://doi.org/10.1161/01.cir.0000149749.95902.a4

Figures/Video legend:

Figure 1: PWD over TV showing significant respiratory variations.

Figure 2: PWD over MV showing significant respiratory variations.

Figure 3: Apical four-chamber view [A: end diastolic frame, B: End systolic frame] showing mild LV systolic dysfunction by modified Simpson's method.

Video 1: Subcostal view showing large pericardial effusion with RA and RV collapse.

Video 2: Modified apical view showing large pericardial effusion with RA and RV collapse.

Video 3: Apical four-chamber view showing hypokinesia and ballooning of the apical segments.

Video 4: Zoomed apical four-chamber view showing hypokinesia and ballooning of the apical segments.

Video 5: Modified parasternal view showing hypokinesia and ballooning of the apical segments.

Video 6: Apical four-chamber view showing improvement of LV systolic function, hypokinesia, and ballooning of the apical segments.

Video 7: Zoomed apical four-chamber view showing improvement of LV systolic function, hypokinesia, and ballooning of the apical segments.