Case Report

Nepalese Heart Journal 2024; Vol 21(1), 51-54

Acute Coronary Syndrome in a Patient with Sarcoidosis: A Case Report

Rohit Murarka[1], Asraf Hussain[1], Shyam Raj Regmi1, Bishnu Mani Dhital[1], Amir Khan[1], Sagar Thapa[1], Ram Gobind Chaudhary[1], Indresh Yadav[1], Roshan Shrestha[1]

[1] Department of Cardiology, Chitwan Medical College, Bharatpur, Nepal

Corresponding Author: Asraf Hussain
Email: asrafjeevanjyoti2060@gmail.com

Cite this article as: Murarka, R. ., Hussain, A. ., Regmi, S. R., Dhital, B. M., Khan, A., Thapa, S., Chaudhary, R. G., Yadav, I., & Shrestha, R. . Acute Coronary Syndrome in a Patient with Sarcoidosis: A Case Report. Nepalese Heart Journal 2024; 21(1), 51-54

Submission date: 12th july 2023
Accepted Date: 6 April 2024

Abstract

Sarcoidosis is a complex systemic disorder characterized by an increased cellular immune response and the development of non-caseating granulomas. Sarcoidosis can affect any other organ and cardiac involvement has been reported in a substantial percentage of individuals. A sound clinical evaluation is needed for rapid recognition and implementation of appropriate treatment alternatives. Chest pain is a common presenting symptom in sarcoidosis. We present here an interesting case of a 39-year-old female presenting with ST elevation myocardial infarction who was managed by primary percutaneous intervention.

Key words: Acute coronary syndrome, Coronary Angiography, Coronary Intervention, Sarcoidosis

DOI: https://doi.org/10.3126/nhj.v21i1.65653

Introduction

Sarcoidosis is a complex systemic disorder characterized by an increased cellular immune response and the development of non-caseating granulomas. Because it can affect practically every organ in the body, it can induce a wide range of clinical symptoms.[1] The etiology of sarcoidosis is unknown, and because it can mimic other common clinical illnesses, diagnosing it can be difficult. Although the prognosis can be improved, early discovery and vigorous treatment are required.[1]

While sarcoidosis can affect many other organs, cardiac involvement has been reported in a substantial percentage of individuals. The first recorded case of cardiac sarcoidosis occurred in1929, and subsequent autopsy studies have showed that up to 76% of sarcoidosis patients have cardiac involvement.[2] Despite its prevalence, cardiac sarcoidosis is commonly misdiagnosed because it resembles other heart illnesses. A sound clinical evaluation is needed for rapid recognition and implementation of appropriate treatment alternatives.[3]

Studies suggest that sarcoidosis results from the combination of immune dysregulation, environmental factors, and genetic predisposition.[6]

Sarcoidosis can present clinically in a variety of ways depending on the organs involved. Typical symptoms include fatigue, weight loss, fever, and general malaise. Pulmonary involvement is the most common sarcoidosis symptom, and patients usually complain of chest pain, shortness of breath, and coughing. Sarcoidosis, on the other hand, can affect the skin, eyes, liver, spleen, lymph nodes, and neurological system causing a variety of symptoms and issues.[7]

Chest pain is a common presenting symptom in sarcoidosis. Perfusion defects indicative of myocardial ischemia was detected in almost 50% cases in a study focusing on the importance of angina in sarcoidosis patients.[4] It is important to note that, despite the discovery of myocardial involvement, epicardial coronary involvement was not pathologically established in explanted hearts untilrecently.[5]

Sarcoidosis must be diagnosed using clinical evaluation, imaging studies, laboratory tests, and histological analysis. The first evaluation includes a detailed medical history and physical examination to search for any possible organ involvement symptoms. Laboratory tests may reveal abnormalities such as hypercalcemia, liver dysfunction, or elevated angiotensin-converting enzyme (ACE) levels. These findings, however, can be discovered in a variety of disorders, not just sarcoidosis.

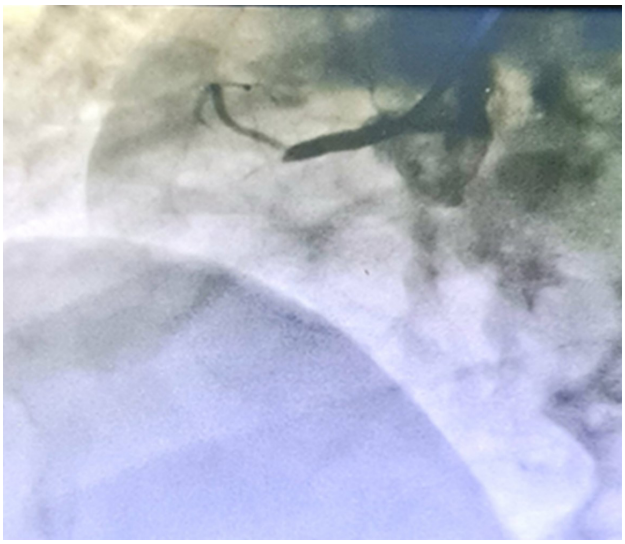

Figure 1: Coronary angiogram showing total occlusion in Proximal RCA.

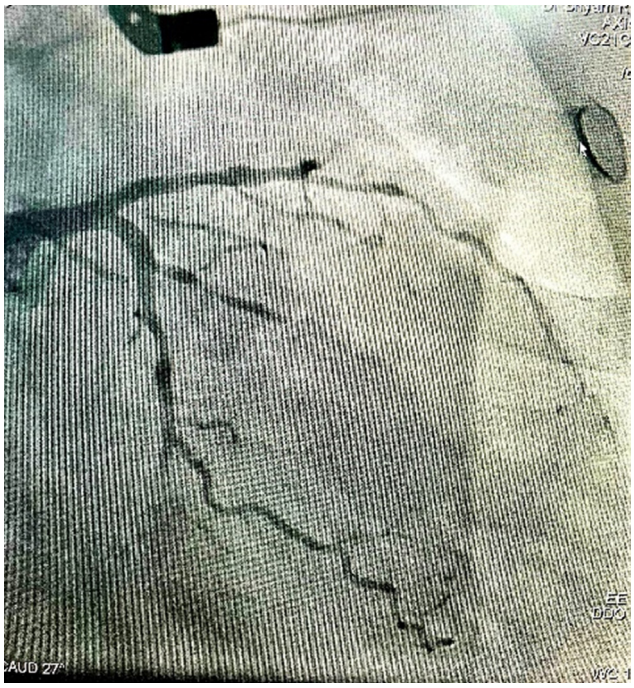

Figure 2: Coronary angiogram showing 100% stenosis in mid LAD and diffused disease in LCX and OMI

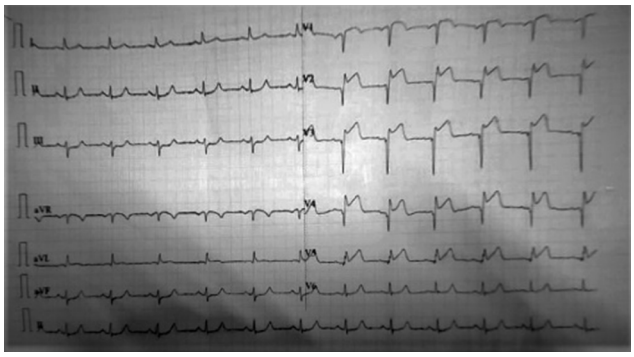

Figure 3: ECG revealed ST elevation in V1-V4 with reciprocal ST depression in II, III and aVF.

Imaging investigations, such as chest X- rays, CT scans, or magnetic resonance imaging (MRI), are critical for establishing organ involvement and assessing disease severity. Sarcoidosis is distinguished by pulmonary infiltrates and bilateral hilar lymphadenopathy. Other imaging modalities, including as positron emission tomography (PET) scans, can help determine extra pulmonary involvement and disease activity.

For a definitive diagnosis of sarcoidosis, histopathological evidence of non-caseating granulomas is required. A biopsy of the affected organ or tissue is typically performed to do this. A heart biopsy in patients with cardiac sarcoidosis may be difficult to acquire because to its invasive nature. However, in some cases where there is a high clinical suspicion, cardiac samples or even explanted heart testing may be required to establish the diagnosis.[9]

When a diagnosis of sarcoidosis is made, the severity and amount of organ involvement decide how the disease should be managed. Sarcoidosis may occasionally resolve on its own or require only supportive care. However, when organ function is severely damaged or the disease is progressing, medication is essential to control inflammation and avert the consequences.[10]

Case Presentation

A 39-year-old woman, presented with chest pain, and shortness of breath that had been persistent for five days. She was a known case of Sarcoidosis for five years. She was diagnosed to have hypertension and type2diabetes for two years. She was treated for pulmonary tuberculosis on clinical grounds. No improvement in symptoms even after completion of anti-tubercular therapy warranted further evaluation.

During work up her angiotensin-converting enzyme (ACE) level was elevated the value was more than 300 U/L; normal value 12-68 U/L. On USG abdomen and pelvis, she had hepatomegaly and numerous intraabdominal lymph nodes. A true cut biopsy of the liver was done and reported as many lymphoid and fibrous tissue-encircled non-caseating epithelioid cell granulomas. The findings of Granulomatous inflammation on liver biopsy pointed to liver involvement in sarcoidosis.

After diagnosis of Sarcoidosis was made, she was being treated with azathioprine 50mg and prednisolone as and when required. She was doing well one day prior to presentation at our center.

On evaluation at our center the ECG revealed ST elevation in V1-V4 with reciprocal ST depression in II, III and AVF. Her Qualitative Troponin done outside center was positive. Diagnosis of anteroseptal STEMI was made. Echocardiography revealed hypokinesia of Left anterior descending artery (LAD) territory and LVEF of 35%. Coronary angiogram was performed and diagnosis of triple vessel disease involving LAD, Right coronary artery (RCA) and Left circumflex artery (LCX) was made. PCI was performed on LAD and RCA.

Her condition was stable at the time of discharge. She was discharged on Dual antiplatelet therapy (DAPT), Sodium Glucose Transporter 2 (SGLT2) inhibitors, Mineralocorticoid receptor antagonist (MRA), Angiotensin receptor blocker (ARB), Diuretics and azathioprine. Prednisolone therapy was avoided keeping in consideration the acute myocardial infarction.

On follow-up she did not have complaints of chest pain or chest heaviness. Her vitals were stable with LVEF of 45% on 2D echo.

Discussion:

A systemic inflammatory illness called sarcoidosis is characterized by the development of non-caseating granulomas in the organs that are afflicted. Heart failure, conduction problems, arrhythmias, and sudden cardiac death are just a few of the manifestations that can result from cardiac involvement in sarcoidosis, often known as cardiac sarcoidosis (CS). The connection between sarcoidosis and ACS is, however, not very common.[11] However, our patient had cardiac involvement in terms of heart failure and coronary artery disease which was very less commonly reported.

Uncertainty exists regarding the precise pathophysiological mechanisms behind the ACS development in sarcoidosis. The infiltration of coronary arteries by sarcoid granulomas is thought to cause luminal constriction and ensuing myocardial ischemia. The development of atherosclerosis and plaque instability may be aided by inflammation and fibrosis within the artery walls, raising the risk ofACS.[12]

It was unexpected, nonetheless, for ACS to develop alongside sarcoidosis.[13] Our diagnosis was supported with initial findings of

non-caseating granulomas in the liver biopsy and increase level of ACE. In our case indicated that the patient, had a known history of sarcoidosis.

Patient was diagnosed with diabetes and hypertension for short duration of two years. There was absence of any other traditional risk factor for coronary artery disease in this young lady. At the time of diagnosis of sarcoidosis there was involvement of liver and lungs. The nature of presentation in this case supported the causal role of disease in development of acute MI.

Due to the necessity to combine antiplatelet and anticoagulant medications with the danger of aggravating granulomatous inflammation, the management of ACS in patients with sarcoidosis can be difficult. Due to the amount and severity of coronary artery disease in our situation, revascularization was advised.[14] This strategy was designed to increase the myocardial blood flow and reduce symptoms. The treatments also included antiplatelet medications, statins, beta-blockers, and angiotensin receptor blockers.[15]

The management of her illness was made difficult by the existence of numerous comorbidities, including hypertension, type 2 diabetes, and her coronary artery disease most likely developed and progressed as a result of these comorbidities. As prolonged inflammation and the development of granulomas can have resulted in vascular dysfunction and a prothrombotic condition, sarcoidosis itself may have contributed to the emergence of ACS.[16]

In order to effectively manage patients with sarcoidosis and ACS over the long term, medical therapy must be optimized with frequent follow-up visits. Ventricular arrhythmias and sudden cardiac death are likely to occur in ACS patients, hence, implanted cardioverter-defibrillator (ICD)insertion may be considered in certain circumstances.[17]

Conclusion:

The necessity of taking sarcoidosis into account as a possible underlying cause in patients who come with ACS is highlighted by this case, especially in those who have a history of sarcoidosis or suspicious clinical characteristics. For these patients' results to be maximized, early identification and adequate therapy are essential. Unanswered questions that whether the pathophysiology and behavior of the coronary artery disease remains different in this subset of patients' needs to be addressed. Alternative therapies which have less deleterious effect on long term basis needs to be sought. More investigation is required to comprehend the pathophysiological pathways that connect sarcoidosis and ACS and to create specialized treatment plans for this particular population.

References

1. Sève P, Pacheco Y, Durupt F, Jamilloux Y, Gerfaud-Valentin M, Isaac S, et al. Sarcoidosis: A Clinical Overview from Symptoms to Diagnosis. Cells. 2021 Mar 31;10(4):766. doi : 10.3390/cells10040766
2. Sekhri V, Sanal S, DeLorenzo LJ, Aronow WS, Maguire GP. Cardiac sarcoidosis: a comprehensive review. Archives of Medical Science : AMS [Internet]. 2011 Aug 1 [cited 2021 Nov 4];7(4):546–54. doi: 10.5114/aoms.2011.24118
3. Nunes H, Bouvry D, Soler P, Valeyre D. Sarcoidosis. Orphanet Journal of Rare Diseases [Internet]. 2007;2(1):46. Available from: https://ojrd.biomedcentral.com/articles/10.1186/1750-1172-2-46 doi: 10.1186/1750-1172-2-46
4. Truong, Justina, and John Ashurst. "A Case Report of Pulmonary Sarcoidosis: An Uncommon Cause of Chest Pain." Clinical Practice and Cases in Emergency Medicine, vol. 4, no. 4, 9 Sept. 2020, pp. 645–648, www.ncbi.nlm.nih.gov/pmc/articles/PMC7676790/ . doi: 10.5811/cpcem.2020.7.48310
5. Uthurralt, N, et al. "Thallium-201 Scintigraphy for Diagnosis of Old Myocardial Infarction: Comparison with Electrocardiographic, Ventriculographic, and Coronary Arteriographic Findings." Heart, vol. 43, no. 5, 1 May 1980, pp. 527–534. doi: 10.1136/hrt.43.5.527
6. Jain, Rashi, et al. "Sarcoidosis: Causes, Diagnosis, Clinical Features, and Treatments." Journal of Clinical Medicine, vol. 9, no. 4, 10 Apr. 2020, p. 1081, https://doi.org/10.3390/jcm9041081. Doi: 10.3390/jcm9041081
7. Nunes, Hilario, et al. "Sarcoidosis." Orphanet Journal of Rare Diseases, vol. 2, no. 1, 2007, p. 46, ojrd.biomedcentral.com/articles/10.1186/1750-1172-2-46, https://doi.org/10.1186/1750-1172-2-46. Doi: 10.1186/1750-1172-2-46
8. Crouser, Elliott D., et al. "Diagnosis and Detection of Sarcoidosis. An Official American Thoracic Society Clinical Practice Guideline." American Journal of Respiratory and Critical Care Medicine, vol. 201, no. 8, 15 Apr. 2020, pp. e26–e51, https://doi.org/10.1164/rccm.202002-0251st.
9. Melani, Andrea S., et al. "A Comprehensive Review of Sarcoidosis Diagnosis and Monitoring for the Pulmonologist." Pulmonary Therapy, 6 June 2021, link.springer.com/article/10.1007%2Fs41030-021-00161-w, https://doi.org/10.1007/s41030-021-00161-w
10. Ungprasert, Patompong, et al. "Clinical Manifestations, Diagnosis, and Treatment of Sarcoidosis." Mayo Clinic Proceedings: Innovations, Quality & Outcomes, vol. 3, no. 3, Sept. 2019, pp. 358–375, https://doi.org/10.1016/j.mayocpiqo.2019.04.006.
11. Lehtonen, Jukka, et al. "Cardiac Sarcoidosis: Phenotypes, Diagnosis, Treatment, and Prognosis." European Heart Journal, vol. 44, no. 17, 16 Mar. 2023, p. ehad067, pubmed.ncbi.nlm.nih.gov/36924191/, https://doi.org/10.1093/eurheartj/ehad067.
12. Gonen, Tal, et al. "The Association between Sarcoidosis and Ischemic Heart Disease—a Healthcare Analysis of a Large Israeli Population." Journal of Clinical Medicine, vol. 10, no. 21, 29 Oct. 2021, p. 5067, www.ncbi.nlm.nih.gov/pmc/articles/PMC8584952/, https://doi.org/10.3390/jcm10215067. Accessed 8 Mar. 2023.
13. "Hepatic Sarcoidosis." Journal of Clinical and Translational Hepatology, vol. 1, no. 2, 1 Dec. 2016, https://doi.org/10.14218/jcth.2013.00016. .
14. Sopek Merkaš, Ivana. "Antiplatelet Therapy after Coronary Artery Bypass Graft Surgery – Unevenness of Daily Clinical Practice." Acta Clinica Croatica, 2021, https://doi.org/10.20471/acc.2021.60.03.26.
15. Kumar, Amit, and Christopher P Cannon. "Acute Coronary Syndromes: Diagnosis and Management, Part I." Mayo Clinic Proceedings, vol. 84, no. 10, 2009, pp. 917–38, www.ncbi.nlm.nih.gov/pmc/articles/PMC2755812/, https://doi.org/10.1016/S0025-6196(11)60509-0.

16. Leon, Benjamin M, and Thomas M Maddox. “Diabetes and Cardiovascular Disease: Epidemiology, Biological Mechanisms, Treatment Recommendations and Future Research.” World Journal of Diabetes, vol. 6, no. 13, 10 Oct. 2015, p. 1246, www.ncbi.nlm.nih.gov/pmc/articles/PMC4600176/, https://doi.org/10.4239/wjd.v6.i13.1246.

17. Skowasch, Dirk, et al. “Management of Sudden Cardiac Death in Cardiac Sarcoidosis Using the Wearable Cardioverter Defibrillator.” PLOS ONE, vol. 13, no. 3, 22 Mar. 2018, p. e0194496, https://doi.org/10.1371/journal.pone.0194496.

.