Acute Pericarditis as a Rare Presentation of Cardiac Involvement in Sarcoidosis

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Introduction

Sarcoidosis is a chronic systemic disease without any identified etiology. In general, it affects the lungs and lymph nodes, but can implicate any other organ. Cardiac involvement in patients with systemic sarcoidosis is usually the cause of death.1 Usually, patients present with complete heart block, bundle branch block, ventricular tachycardia (VT), congestive heart failure, and/or sudden death.2 The myocardium and, to a lesser extent, the endocardium are usually involved. Pericardial involvement often presents as moderate to large pericardial effusion detected in echocardiography.3 We here present a case of symptomatic acute pericarditis, which is a rare manifestation of cardiac involvement in sarcoidosis.1,4,5

Case Report

A 65-year-old male patient arrived at our emergency department with dull oppressive retrosternal chest pain without other accompanying symptoms. The pain was exacerbated with inspiration, and had started 14 hours before. The patient had a medical history of pulmonary sarcoidosis diagnosed 10 years before (for which he was on a regimen of low doses of oral prednisolone), arterial hypertension and a hemorrhagic stroke 18 years ago. At initial presentation, blood pressure was 130/80 mmHg, heart rate was 90 beats/min, oxygen saturation 95% and temperature was 36.8°C. A pericardial friction rub was audible without any further pathological sounds on cardiac or respiratory auscultation. Chest X-Ray showed a normal cardiothoracic ratio and hilar enlargement, and the 12-lead electrocardiogram (ECG) showed widespread concave ST-segment elevation and PR-segment depression throughout most of the limb leads and precordial leads (Figure 1). Transthoracic echocardiography showed a normal myocardial appearance, LV diastolic dysfunction, and the presence of only minimal (6 mm-thick) pericardial effusion. Laboratory studies on admission revealed elevated white blood cell count (WBC: 12,680/μL) and elevated serum C-reactive protein (CRP) level (7.69 mg/dL); high-sensitive troponin-I and creatine kinase levels were normal (4.0 pg/ml and 101 IU/L respectively). Other diagnostic evaluations for viruses and thyroid function were negative.

The patient was hospitalized and started on intravenous prednisolone 75 mg/d and oral colchicine 1 mg/d. Three days later, he was discharged symptom-free on oral colchicine 1 mg/d and oral prednisolone 10 mg/d gradually tapered over the following weeks.

At the 10th day of follow-up, laboratory tests had been normalized (WBC: 9,200/μL and CRP: 0.12 mg/dL), ST-segment elevations had largely resolved and there was no pericardial fluid in the echocardiogram.

The patient was referred for cardiac magnetic resonance (CMR) study with gadolinium, which offers the advantage of high sensitivity and spatial resolution, in addition to no radiation exposure.7 Since this imaging technique was not available in our hospital, the patient underwent CMR in a tertiary center at a later stage, after six weeks under treatment, which ruled out granulomatous infiltration of the myocardium (Figure 2). Furthermore, the small pericardial effusion had resolved completely, which was in accordance with the normalized inflammation markers and the patient’s asymptomatic condition.

Keywords

Pericarditis; Sarcoidosis, Pulmonary; Hypertension; Stroke.
Figure 1 – Panel A: ECG from 1 year ago. Panel B: ECG at admission. Concave ST-segment elevations and PR-segment depressions were recorded throughout most of the limb leads (II, III, aVF) and precordial leads (V2-6), with reciprocal PR elevation and ST depression in aVR. Panel C: ECG at 10-days follow-up.

Discussion

Acute pericarditis with or without pericardial effusion is an inflammatory pericardial disease considered idiopathic in approximately 80% of cases. Secondary causes include several systemic diseases, such as systemic lupus erythematosus, Sjogren’s syndrome, rheumatoid arthritis and scleroderma, and, more rarely, sarcoidosis.

Only 5% of patients with systemic sarcoidosis develop cardiac symptoms; however, pathology studies reveal myocardial granulomas in 20-30% of patients. Pericardial involvement is considered rare even if widespread myocardial infiltration has occurred. Symptomatic acute pericarditis without pericardial effusion is seldom observed in sarcoidosis.

However, unlike sarcoidosis with the involvement of other organs, cardiac sarcoidosis carries an increased risk of sudden death and can be challenging to diagnose. Due to the rarity of the condition, there are no clearly validated guidelines, and our experience relies mostly on published case reports and small series of patients rather than on large population studies.

The early and reliable diagnosis of cardiac involvement remains extremely difficult, and even subtle cardiac symptoms require thorough investigation by the clinician. Cardiac involvement in pulmonary sarcoidosis is associated with a poor prognosis, especially once patients become symptomatic. Conversely, asymptomatic patients have a good prognosis; therefore, early diagnosis and treatment are crucial. Physicians must be aware of silent cardiac involvement in patients with pulmonary sarcoidosis, and thus, these patients should be routinely monitored both clinically and with an ECG, reserving echocardiography, and when necessary CMR, for cases raising a higher degree of suspicion.

Considering cardiac sarcoidosis is associated with an increased risk of sudden death, especially in case of myocardial infiltration, several therapeutic strategies should be considered. Early treatment with corticosteroids...
or other immunosuppressant agents may prevent the progression of cardiac involvement. Certain cases of high-grade AV conduction block or other types of bradyarrhythmia justify cardiac pacemaker implantation.\textsuperscript{10} Moreover, life-threatening ventricular arrhythmias triggered by myocardial infiltrates may require implantable cardioverter-defibrillator therapy for primary prevention of sudden cardiac death in selected high-risk patients.\textsuperscript{10}

**Conclusion**

We here present a case of acute pericarditis, which is an unusual cardiac manifestation of sarcoidosis. The present case emphasizes that early diagnosis of cardiac involvement remains challenging and routine screening of pulmonary sarcoidosis patients is pivotal to monitor the course of the disease.

**Acknowledgements**

We would like to acknowledge the contribution of the staff of the Cardiology department to the work presented.

**Author contributions**

Conception and design of the research: Bostanitis I, Papadopoulou SL. Acquisition of data: Bostanitis I, Papadopoulou SL, Tsalidou M, Triantafyllou K. Analysis and interpretation of the data: Bostanitis I, Papadopoulou SL, Triantafyllou K, Girasis C. Writing of the manuscript: Bostanitis I, Papadopoulou SL, Tsalidou M, Girasis C. Critical revision of the manuscript for intellectual content: Bostanitis I, Papadopoulou SL, Tsalidou M, Triantafyllou K, Girasis C.
Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

Sources of Funding

There were no external funding sources for this study.

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