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CASE REPORT

Isolated Cardiac Sarcoidosis- A Rare Case in Bangladesh.

MOSTASHIRUL HAQUE, MOHAMMAD AL- MAMUN, ZAFOR IQBAL JAMALI, CHAYAN KUMAR SINGH, FAKHRUL ISLAM KHALED, NITAY KUMAR GOSH, MD. ASHRAF UDDIN SULTAN, DMM FARUQUE OSMANY

Department of Cardiology, Bangladesh Medical University (BMU), Shahbag, Dhaka, Bangladesh

Address of Correspondence: Dr. Mostashirul Haque, Associate Professor, Department of Cardiology, Bangladesh Medical University (BMU), Shahbag, Dhaka, Bangladesh. E-mail: mostashirul@gmail.com

Abstract:

Cardiac sarcoidosis (CS) is a granulomatous inflammatory disorder of the myocardium with a broad spectrum of clinical manifestations, comprising incidental conduction disorder, heart failure, arrhythmia, and sudden death. We present an unusual case of cardiac sarcoidosis that mimicked hypertrophic cardiomyopathy (HCM). Sarcoidosis has the lowest prognosis when it involves the heart muscle, resulting in more than two-thirds death of all sarcoid patient. Myocardium involvement can occur alone or in conjunction with other organ involvement, most notably pulmonary sarcoidosis which is about 90%. Here our patient has cardiac presentation.

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Case Report:

A 43 years old diabetic, normotensive patient presented with shortness breath. ECG showed Left ventricular hypertrophy with strain, interventricular conduction defect. Chest Xray shows cardiomegaly without bilateral hilar lymphadenopathy. Trans-thoracic echocardiogram (TTE) demonstrated preserved left ventricular (LV) function with asymmetric septal hypertrophy (ASH) measuring 30 mm, without outflow tract obstruction. Abdominal fat pad biopsy revealed no evidence of amyloidosis. Cardiac MRI confirmed ASH and late gadolinium enhancement (LGE) shows spotty enhancement occupying antero- septal, posterior- septum in basal to apical segment along with basal anterior part of LV myocardium. Localized regional wall motion abnormality corresponding to the late gadolinium enhancement (LGE). Non dilated hypertrophied ventricle with both atrial enlargement shows restrictive filling pattern. According to JCS 2016 guideline clinical criteria for isolated cardiac sarcoidosis was satisfied for diagnosis (Focal area perfusion uptake mismatch in FDGPET, IV conduction defect in ECG, Asymmetric septal hypertrophy in Echocardiogram, LGE in Cardiac MRI). Then the patient was treated conservatively with steroids. He becomes symptom free for last 2 weeks.

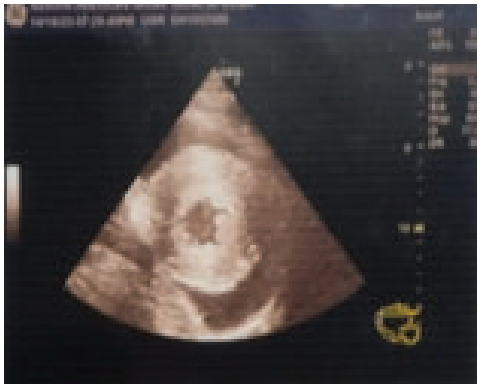

Figure 1: *PSAX view shows hypertrophic ventricular wall*

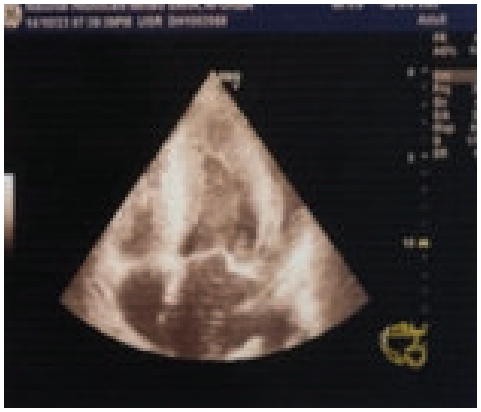

Figure 2: *Apical 4 chamber view shows asymmetric basal septal hypertrophy*

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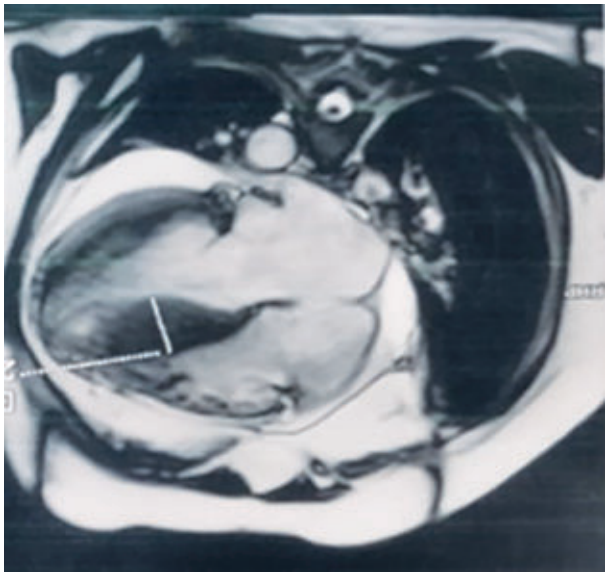

Figure 3: *Cardiac MRI shows asymmetric septal hypertrophy*

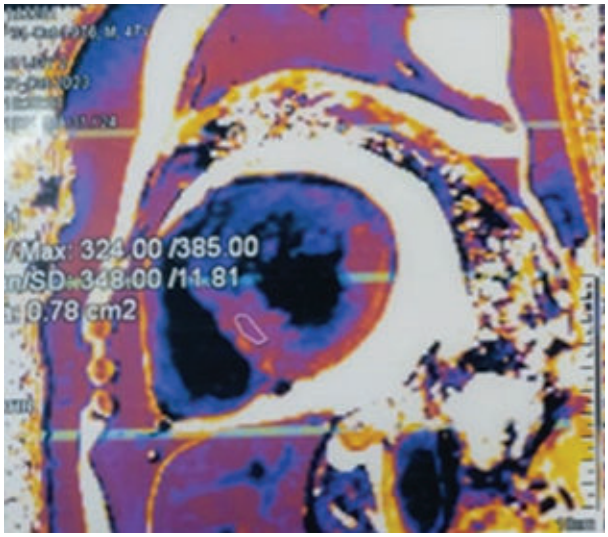

Figure 4: *Late gadolinium enhancement*

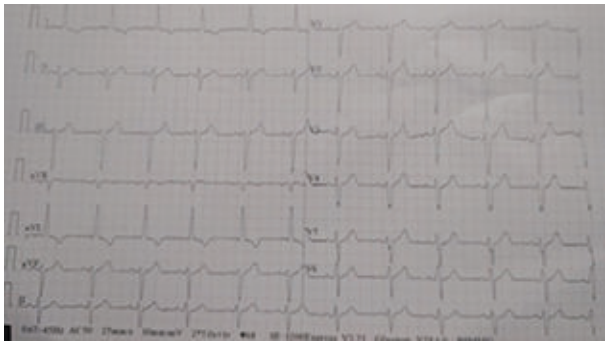

Figure 5: *ECG shows LVH with IV conduction defect (LBBB)*

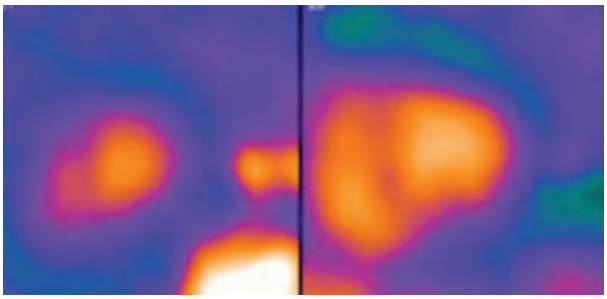

Figure 6: *FDGPET shows normal perfusion with mild increased uptake at basal septal region*

Discussion:

Cardiac sarcoidosis is the most concerning form of sarcoidosis, with a frequency of 20 to 30% based on autopsy studies and cardiac MRI screening of known sarcoid patients.[1] Only 5% of these people, however, had clinical manifestations of the disease.[2] Although isolated CS is considered to account for approximately 27% to 54% of all CS cases, current studies using whole-body PET scans suggest a lower and varied prevalence (3). As a result, careful evaluation is required to rule out the presence of sarcoid lesions in other organs before diagnosing isolated disease. Data regarding cardiac sarcoidosis in Bangladesh is very rare.

The wide spectrum of clinical manifestations of CS arises from the variable locations of sarcoid lesions in the myocardium. There is however a predilection for the base of the interventricular septum, the conduction system, and the left ventricular (LV) free wall.[4] In consistence, presented case had involvement of the base of the interventricular septum, the conduction system, and the left ventricular (LV) free wall. In our cases ECG shows LBBB, Schuller et al reported that a bundle branch block pattern is associated with cardiac involvement in a cohort of individuals with biopsy proven pulmonary sarcoidosis.[5] Cardiac MRI is important because it is a noninvasive method of diagnosing morphological and functional problems associated with CS. Aside from its excellent sensitivity and specificity, it is useful in our country where EMB is not available. The fluor deoxy glucose positron emission tomography (FDGPET) scan, which uses radioactive glucose to detect areas of active inflammation in CS, has a sensitivity and specificity of 89% and 78%, respectively. However, FDGPET is considerably more difficult to get in many resources constrained countries.[6] FDGPET report of our patient shows normal perfusion with mild increased uptake at anterior basal, inferior basal region suggesting early active inflammation.

Systemic corticosteroids remain the cornerstone of CS treatment and are considered first line drugs due to their efficacy and ability to achieve a meaningful response in a

very short period of time. In the event of corticosteroid failure and the development of adverse effects, nonsteroidal immunosuppressive drugs such as methotrexate, azathioprine, mycophenolate mofetil, leflunomide, cyclosporine, or cyclophosphamide are alternatives, particularly when higher doses are required (>10 mg/day prednisolone). In a cohort study of 36 CS patients, Harper et al found that using infliximab resulted in lower steroid doses and fewer dysrhythmia.[7] ICD implantation is an option for patients with LVEF more than 35% who have had syncope, evident myocardial scarring on cardiac MRI or PET scan, and/or induced ventricular arrhythmias during EPS.[8]

Our patient responds well with steroid.

Conclusion:

Diagnosis of CS requires a high level of suspicion with multimodality imaging such as MRI, CT, and PET, is essential particularly in patients with systemic sarcoidosis. Active screening of these patients will encourage early diagnosis and treatment, which may spare individuals.

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