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HOCM *vs*. Cardiac Sarcoid - Diagnostic Challenge

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ABSTRACT

The true prevalence of cardiac involvement in Sarcoidosis is unknown; it has been reported as high as 58% in Japan and approximately 20% - 27% in US. The clinical manifestation ranges from incidental finding to heart failure and sudden death. Cardiac sarcoid can present in different ways depending on the location and extent of myocardial involvement. The hypertrophic stage of cardiac sarcoid is rarely seen. Immunosuppressive therapy is the mainstay of treatment and the prognosis largely depends on the extent of myocardial involvement and presence of rhythm abnormalities. We present a case of a 60-year-old gentleman whose presentation was confusing and diagnosis was challenging as it has mimicked hypertrophic cardiomyopathy.

KEYWORDS

Sarcoidosis; Heart failure; Hypertrophic cardiomyopathy; Immunosuppressive therapy

CASE STUDY

A 60-year-old gentleman presented to our acute medical assessment unit with 4 weeks history of dry, irritating cough without any other constitutional symptoms. He has never smoked and had background history of Hypertension, GORD and Cataract surgery. He was on ACE inhibitor (Ramipril 10 mg) for hypertension. There was no significant family history of note; his son was known to have mild pulmonary sarcoid. On clinical examination he had pan systolic murmur at Mitral area and chest was clear on auscultation. He had no peripheral oedema or signs of decompensated heart disease. On investigations ECG findings were consistent with left ventricular hypertrophy (voltage criteria). His Chest X-ray was unremarkable [Figure 1], BNP, Troponins, FBC, electrolytes, serum Calcium and LFTs were within normal limits. His cough was attributed to ACE-Inhibitor and hence was switched to ARB and an outpatient cardiac ECHO (in view of murmur) and pulmonary function test were organised. Trans-thoracic ECHO showed good systolic function with asymmetric apical hypertrophy with septal thickening of 1.7 cm and mild mitral regurgitation. On focused scan, there was systolic anterior motion of mitral sub-valvular apparatus with turbulence and the left ventricular outflow tract (LVOT) peak gradient was noted to be 21 mm HG which increased to 107 mm Hg on Valsalva manoeuvre. His PFTs were normal. He was reviewed with result of ECHO and was clinically well and his cough has resolved after stopping ACE inhibitor. Subsequently an Outpatient Cardiac MRI was organised for him that was reported as "*strongly suspicious for hypertrophic cardiomyopathy with asymmetric helical septal hypertrophy, mitral valve elongation, SAM and LVOT obstruction. No myocardial fibrosis was*

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identified." There was however marked hilar and mediastinal lymphadenopathy at that stage on Cardiac MRI. He was referred for genetic testing as the structural changes were pointing towards hypertrophic cardiomyopathy.

He re-presented to AMAU with dry cough, exertional dyspnoea, unintentional weight loss generalized weakness, headache, paraesthesia and blurring of vision. His corrected serum calcium level was elevated. His CXR showed extensive fibrotic changes (Figure 2), the previous one being unremarkable on 1st presentation. He was admitted, HRCT and MRI brain was organised. HRCT showed extensive bi-hilar and mediastinal lymphadenopathy and parenchymal involvement and likely diagnosis of Sarcoid was considered (Figure 3). MRI Brain imaging was performed suggestive of multiple non-specific foci of predominantly subcortical T2 high signal intensity may represent Neuro-sarcoid (Figure 4 and Figure 5). The diagnosis of Sarcoid was confirmed by endobronchial biopsy of lymph node. Unifying diagnosis of Multisystem Sarcoidosis was established and he was commenced on treatment with high dose steroids with PPI and bone protection, beta-blocker. An outpatient coronary angiogram was normal. The genetic testing is negative for classical Hypertrophic Cardiomyopathy. He improved significantly after being treated with steroids and had complete resolution of his symptoms. A follow up Chest x-ray was clear and transthoracic Echo performed showed significant improvement in LVOT gradient down to 24 mm Hg on valsalva manoeuvre although septal hypertrophy persists. He will be followed up in cardiology and respiratory clinics and is currently doing well.

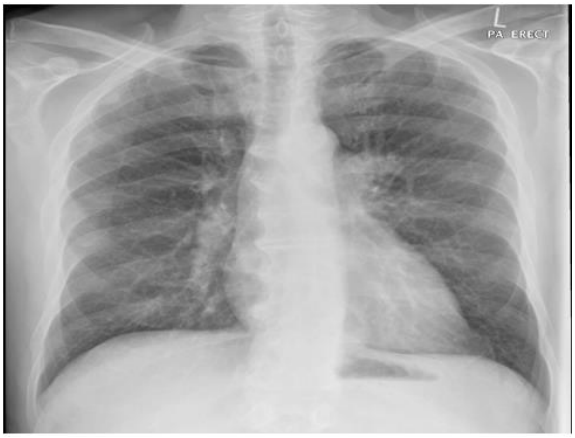

Figure 1: Chest x-ray PA view - unremarkable.

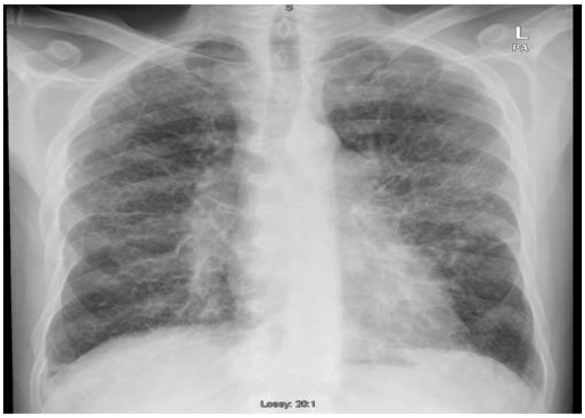

Figure 2: Bi-hilar lymphadenopathy with bilateral parenchymal infiltrate.

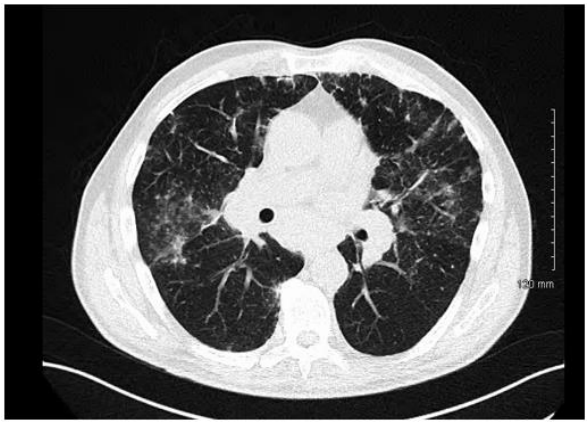

Figure 3: CT Thorax - suggestive of sarcoidosis.

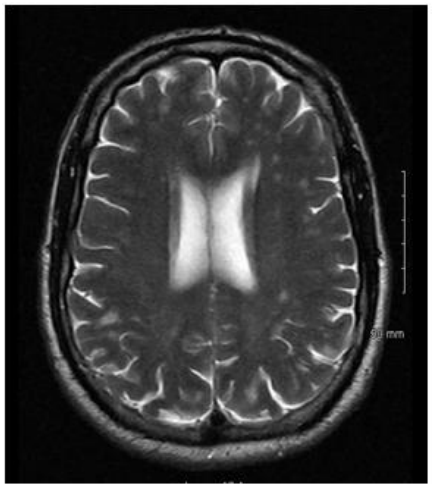

Figure 4: MRI brain.

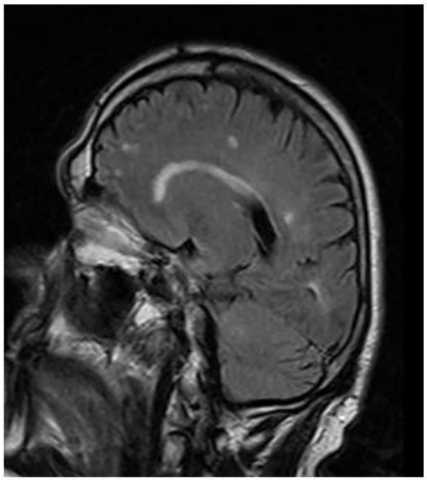

Figure 5: MRI brain.

DISCUSSION

Sarcoidosis is a multisystem granulomatous disorder of unknown etiology, typically involves the lungs. Upto 30% of patients with sarcoidosis would have extrapulmonary manifestations [1]. Cardiac involvement in Sarcoidosis is relatively uncommon but involves all ages and ethnicities. Cardiac Sarcoid ranges from incidental finding to heart failure and death. The most common clinical manifestations are AV blocks, arryhtmia, heart failure and sudden cardiac death [2]. Cardiac involvement is a diagnostic challenge and should be considered in patients with a known diagnosis of extracardiac sarcoidosis, in patients with <60-years of age with unexplained syncope or unexplained conduction abnormalities and patients with sustained ventricular tachycardia who does not have any explanation of sustained VT. In any patient with a clinical suspicion of cardiac involvement 12 lead ECG, Echocardiogram, Holter monitoring and cardiac MRI is needed to make a diagnosis. FDG-PET and Endomyocardial biopsy is rarely needed, however can identify acute inflammation and can identify suitable candidates for immunosuppressive therapy [3].

Echocardiographic findings in patients with CS are variable and may include focal areas of edema resulting in increased wall thickness and mimicking hypertrophic cardiomyopathy (e.g., asymmetric septal hypertrophy) [4]. Among patients with CS, LVEF can be either preserved or reduced. Patients with a dilated cardiomyopathy have dilated LV chambers and depressed LVEF [2]. Patients with a restrictive cardiomyopathy have normal LV chamber sizes and LVEF, and there is evidence of diastolic dysfunction. Echocardiography has low sensitivity for detection of CS of approximately 25 percent to 65 percent as compared with CMR or FDG-PET [5]. The other conditions that can mimic Cardiac Sarcoid are Arrythmogenic Right Ventricular Hypertrophy, Fabry disease, Chagas disease and Tuberculosis.

Our patient underwent genetic testing and Hypertrophic Cardiomyopathy was excluded and FDG-PET confirmed corresponding areas of focal inflammation. Treatment is largely based on the clinical features: i.e. Standard heart failure therapy if EF is low, Beta blockers, ICD in people with risk of sudden cardiac death, Pacemaker implantation in case of AV blocks and Immunosuppression in suitable candidates [6].

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