

Isolated Myocarditis - An Autopsy Case Report

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Abstract: *Isolated myocarditis is a type of myocarditis with unknown origin affecting mostly healthy individuals without any preceding cardiac symptoms. This type is further divided into two broad categories diffuse lymphocytic interstitial and granulomatous myocarditis of which later has been reported very rarely. Due To absence of any recorded abnormalities on electrocardiogram and other modalities these cases are most of the time go undiagnosed. Also, as they follow a very steep course of events granulomatous myocarditis has been associated with considerable morbidity and mortality. early institution of therapy can slow the progression of disease which requires use of ancillary techniques to diagnose the case and high index of suspicion.*

Keywords: isolated myocarditis, granuloma, sudden cardiac death.

1. Introduction

Granulomatous myocarditis is one of the rare forms of myocarditis with the involvement of all the layers of myocardium of unknown etiology. The patients usually die of ventricular arrhythmias and heart failure. Diagnosis is most commonly established by endomyocardial biopsy, histological, immunological and immunohistochemical criteria [1]. Of all the causes most, common cause of granulomatous myocarditis is tuberculosis followed by sarcoidosis and rheumatoid fever while rest of the category falls into idiopathic list.

2. Case Report

The viscera of a 23 years female came to our hospital. We received whole heart, pieces of both lungs, pieces of both kidneys, liver, uterus and cervix in Department of Pathology to establish the cause of death. She died in sub-district hospital with complaints of low grade fever for 1 month. Evidence of uveitis not found. No history of joint pain and rash was given. ECG and cardiac MRI were not done. Her postmortem was conducted by medical officer of sub-district hospital.

On gross examination, heart was of normal weight 280gms with pericardial fat. On trimming right and left atrial walls, thickness was 0.2 cm each, right ventricular wall thickness 0.4cm and left ventricular wall thickness 1.1cm. Interventricular septum measured 1.2 cm. All the great vessels were normal. Whitish patches were seen over the right and left ventricular wall extending upto the valves. [Fig1]. On gross examination lungs, kidney and liver appeared normal.

Microscopically sections showed heavy lymphocytic infiltration with both Langhans type and foreign body type of giant cells and myocytolysis found in both ventricles and left atrium [Fig 2]. All the above features suggestive of Non

caseating granuloma. Other organs lung, kidney and liver showed normal histology.

Special stains PAS for fungi and Wade fite stain for tuberculosis was negative. Final diagnosis of granulomatous myocarditis classified under isolated myocarditis [ICD code - I 40.1] was given as a cause of sudden death.

3. Discussion

Although myocarditis first was described in 1837, the distinction between histologic subtypes did not occur until the late 19th century [2]. In 1929, a different histologic type of myocarditis with giant cells was described in which the giant cells were located within noncaseating granuloma and associated with fibrosis rather than with lymphocytic infiltrates [3]. This disorder is now called cardiac sarcoidosis (CS) or idiopathic granulomatous myocarditis. Granulomatous myocarditis follows a very rapid, progressive and frequently fatal disease that mainly affects young to middle aged previously healthy individuals. A retrospective study of 112 consecutive patients with myocarditis at the Massachusetts General Hospital demonstrated prevalence of granulomatous myocarditis 10% [4]. Only few cases of isolated granulomatous myocarditis had been reported so far in India.

In our case the sections from different areas of heart showed noncaseating granuloma composed of epithelioid cells, multinucleated giant cells both Langhans type and foreign body type with lymphocytic infiltration in between myocytes causing myocytolysis. Special stains AFB, PAS and Warthin starry did not reveal mycobacteria, fungus or spirochete. Asteroid bodies in giant cells, Schuamann bodies with concentric fibrosis in the granulomas are absent, thus rules out cardiac sarcoidosis in this case. Sections from other organs showed normal histology. So in the absence of any causative agents, these granulomatous lesions were classified as isolated myocarditis.

While reviewing literature we came across two case series^[5]^[6] in which patients with similar presentation who dropped dead suddenly with all of the heart chambers enlarged and dilated were reported. Neither the pericardium nor the endocardium showed any abnormalities. No abnormality was noted on echocardiogram as well. In Present case as well, patient was admitted with nonspecific complaints other than low grade fever for one month and upper respiratory tract infection without any myocardial symptoms.

The etiology of granulomatous myocarditis remains to be fully elucidated evidence suggests that it arises owing to immune dysregulation mediated by T lymphocytes^[7]. Few researchers suggest formation of giant cells as a response of myogenic fibers to inflammation caused by viral etiology. In few studies authors^[8] stressed that isolated myocarditis should receive serious consideration in any case of sudden and unexpected death in which, at autopsy, naked eye examination reveals no anatomic lesion which could be held responsible for the event and we attest their view.

To conclude granulomatous myocarditis runs an indolent & fatal course so a high index of suspicion along with the use of ancillary techniques like endomyocardial biopsy should be used to diagnose the case. Rapid diagnosis is critical with early institution of therapy as it can dramatically impact the disease course. The aim of this case report was to create an awareness amongst clinicians regarding this rare entity.

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Figure 1: Gross examination of opened up heart showing white patches present over ventricle;

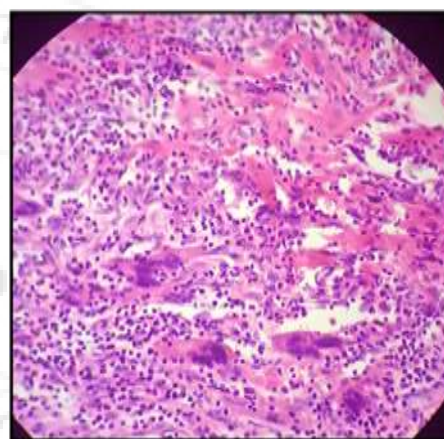


Figure 2: [H&E X 400] Microscopy of noncaseating granuloma showing lymphocytic infiltration in myocardium with giant cells and myocytolysis