Idiopathic Giant Cell Myocarditis Leading to Sudden Death - A Case Report

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Abstract

Idiopathic giant cell myocarditis(GCM) is an uncommon disease of unknown aetiology, often seen in young adults. The demonstrable lesions are limited to heart but cases of extracardiac involvement are also known. We report an autopsy case of Idiopathic GCM with no extracardiac lesion in a thirty year old daily wage-worker, who died suddenly.

The purpose of this report is to emphasize that GCM may exist in the absence of any symptomatic heart disease.

Introduction

Myocarditis is defined as an inflammatory or infectious disease of myocardium causing damage through production of toxin or by immunologically mediated destruction.[1] A rare type is Idiopathic GCM.[1] It often affects young, healthy adults[2] and is clinically characterized by a rapid downhill course resulting into death.[4]

Case Report

A 30 year old, daily wage-worker fell unconscious while on work and was brought dead to the hospital. He was asymptomatic and never had any ailments of heart disease in the past.

At autopsy, the body was of an ill-nourished and well developed young male. The external examination revealed no abnormality. Heart weighed 500 gms. Focal white patches were seen on anterior surface of left ventricle. Left ventricular wall thickness was 1.2 cm. All valves, chambers and great vessels were unremarkable. No evidence of atherosclerosis or infarction was noted grossly. Lungs weighed 450 gms each with no obvious abnormality. All other organs were unremarkable.

Histopathology of heart revealed interstitial destruction of myocardial fibres with replacement by granulomatous process. Focal collection of lymphocytes, plasma cells, eosinophils and a striking presence of large number of Langhan's and foreign body type of giant cells was noted. The cytoplasm of giant cells lacked asteroid bodies, Schumann bodies or any fungi. The inflammatory process showed interdigital pattern of involvement with inflammatory foci in the form of nodules replacing myocardial fibres and pushing the surrounding myocardial fibres at the periphery. Pericardium and endocardium were normal. Fite stain did not reveal acid fast bacilli.

No granulomatous process was seen in lungs. The diagnosis of Idiopathic GCM was offered.

Discussion

In 1991, Fiedlar described isolated myocarditis and thereafter various forms of myocarditis were named after him as Fiedlars myocarditis.[5]

Tesluk reported several cases of

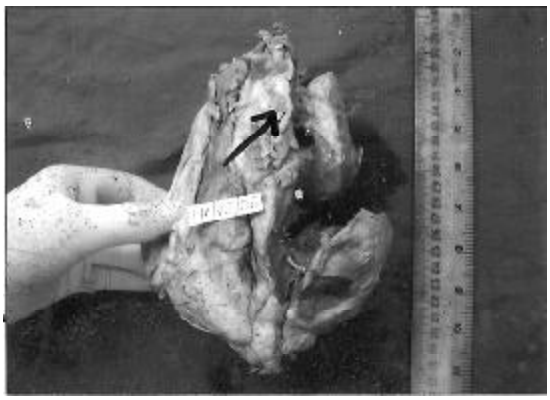

Fig. 1 : Gross photograph of heart with focal white patches on the anterior surface of left ventricle.

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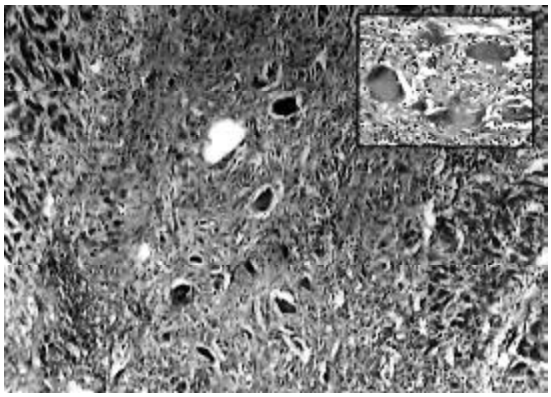

Fig. 2 : Microphotograph of myocardium with replacement of myocardial fibres by granulomatous process. H&E x10.Inset shows high power view to show multinucleated Langhans type of giant cells and lymphocytes. H&E x 40.

myocarditis with extra-cardiac involvement and subtyped it as isolated, granulomatous and giant cell type.[4,5]

Extensive series by Marcose and his co-workers also found extra-cardiac pulmonary lesion, he stated doubt about isolated nature of the lesion.[5] In 1905, Saltikow coined the term Giant cell myocarditis as a disease of unknown aetiology characterised by necrosis of myocardial fibres and infiltration by multinucleated giant cell.[2] GCM is usually diagnosed at autopsy. Median age is forty two to forty six years.[1,6] Youngest reported case in the literature is an infant.[2] Our case presented at 30 year of age.

Many cases of GCM are associated with auto-immune disease like Idiopathic bowel disease.[6] Few cases of GCM showed an association with Thymoma and Myaesthenia gravis.[3]

Major symptom is rapid progressive heart failure or concomitant ventricular arrhythmia.[2,6] Our patient was totally asymptomatic. Aetiology of GCM is unknown. Specific cause of giant cell formation like tuberculosis, syphilis, Chaga's disease, fungal infection and sarcoidosis should be ruled out by thorough evaluation.[3,4]

Histologically it is characterised by widespread circumscribed areas of myocardial necrosis and infiltration by lymphocytes, eosinophils, plasma cells and a large number of multinucleated giant cells. The endocardium and pericardium appear normal.

Our case showed similar interstitial granulomatous infiltrate with normal endocardium and pericardium. Special stains ruled out known aetiological factors associated with GCM.

Unlike lymphocytic myocarditis, GCM had a fairly high incidence of ventricular tachycardia as well as high grade of atrioventricular block. Likelihood of adverse effects- either death or recurrence in cardiac transplantation is higher in GCM than in lymphocytic myocarditis.[4]

Treatment includes immunosuppressive therapy and the indication for cardiac transplantation should be evaluated early as there remains a high risk of recurrence in transplant graft.[1]

Abbreviations

Giant cell myocarditis(GCM).

References

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