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Idiopathic Giant Cell Myocarditis: A Case Report

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Abstract
Giant-cell myocarditis is an extremely rare disease of relatively young, predominantly healthy adults. The patients usually die of heart failure and ventricular arrhythmia unless a cardiac transplantation is performed. We present a case of sudden death in a healthy 45 years old male due to giant cell myocarditis having history of sudden collapse and ultimately death. Histopathology findings of the heart showed non-caseating granulomas with numerous giant cells. Ziehl-Neelsen staining of the sections for acid fast bacilli was negative; stains for fungi were also negative. So, this case is strongly considered to be a case of sudden death due to myocarditis as a result of idiopathic giant cell myocarditis, a rare case in literature.
Keywords: *Giant-cell myocarditis, non-caseating granulomas, asteroid body and Schumann body.*

Introduction

Idiopathic giant cell myocarditis is a cardiovascular disease of the muscle of the heart.[1] It is a rare disease characterized by inflammation of the myocardium, a condition referred to as myocarditis. Inflammation is caused by widespread infiltration of giant cells associated with other inflammatory cells and heart muscle cell destruction. Giant cells are abnormal masses produced by the fusion of inflammatory cells called macrophages. Individuals with giant cell myocarditis may develop abnormal heartbeats, chest pain and, eventually, heart failure. Many individuals eventually require a heart transplant. The disorder most often occurs in young adults. Median age from the time the disease is diagnosed to the time of death is approximately 6 months.[2] Performing autopsies in sudden and unexpected deaths gives a specific, objective and scientific cause of death.

Case Report

According to the inquest report, hospital records and post mortem report received in the department of pathology, deceased was a 45-year-old male, working as a laborer who fainted at around 12 noon while talking at his workplace as per eyewitness' history. He had reported to work after

his breakfast. He was rushed to the hospital by his co-workers where he was admitted in the ICU and died within a few hours of admission. There was no history of previous episodes or co-morbidities. There was no family history of sudden deaths. His body was shifted to the mortuary. After routine inquest, autopsy was conducted the next day. The deceased was a well built adult male, with no external injuries on examination. Stomach contained semi digested food particles.

Gross and microscopic examination

Heart weighed 395 g. There were numerous elevated grayish white patches over the pericardial surface and ventricular walls (Fig. 1). Ventricular septum was grayish white as a whole and papillary muscles were whitish in color. Coronaries were patent. Histology of the heart showed areas of fibrosis in the myocardium with chronic inflammatory cells infiltration (predominantly plasma cells, a few lymphocytes and eosinophils). Numerous large, multi-nucleated giant cells resembling osteoclastic giant cells were seen (Fig. 2, 3and 4). Cause of death was opined as cardiogenic shock due to idiopathic giant cell myocarditis.

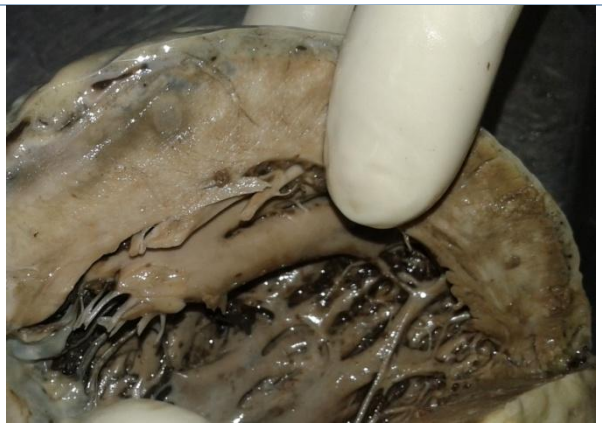

Figure 1 Heart showing grayish white elevated patches on the myocardium.

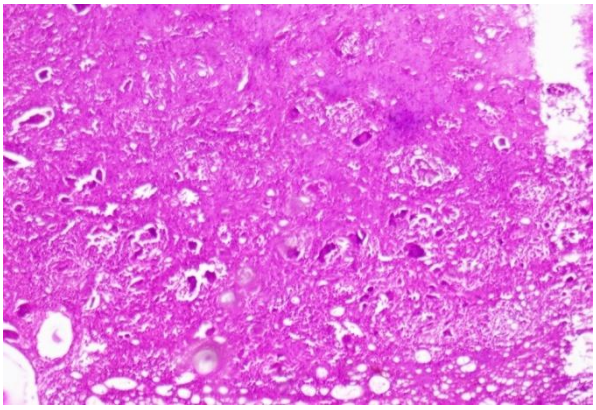

Figure 2 Section shows numerous giant cells in myocardium (H&E, × 100).

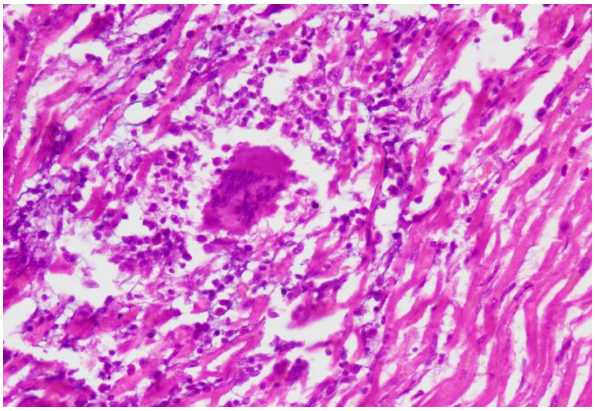

Figure 3 Section of myocardium showing many multinucleated giant cell and granuloma formation. (H&E, × 400).

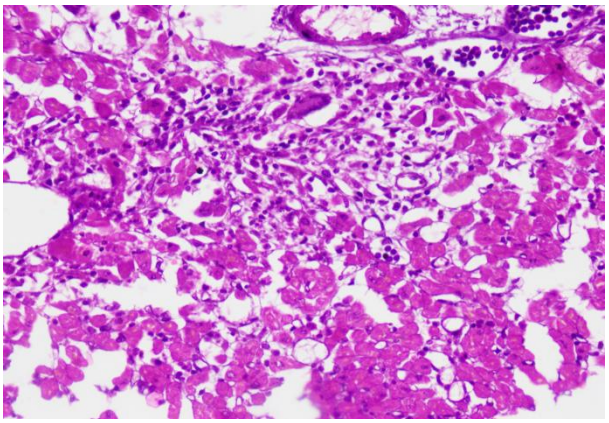

Figure 4 Section of myocardium showing non caseating granuloma lying along with the cardiac muscle (H&E, × 400).

Discussion

CVD is the most important cause of death all over the world. About 40-50% of all cardiovascular deaths have been attributed to SCDs and about 80% of these are caused by cardiac arrhythmias.

SCD is a major public health problem all over the world.[3]

Inflammation of the myocardium is "myocarditis". In previous studies, myocarditis has known to contribute to 6-12% [4,5] of sudden cardiac deaths in young athletes. Myocarditis following an infection is common, of which viral etiology is the most common. A number of viruses including Coxsackie, hepatitis, adenovirus and HIV have known to cause myocarditis. Bacterial, fungal and protozoal infections and hypersensitivity reactions to drugs immunologic syndromes are other causes for myocarditis.[6] The Aschoff lesions of rheumatic myocarditis evolve into characteristic, focal interstitial granulomas with giant cells. Tuberculosis and cryptococcosis also have giant cells within the granulomatous lesions. Special stains for the organisms should be performed whenever there is a suspicion of an infection. Rarely, giant cells may be seen in syphilitic myocarditis. Foreign body reaction, Wegener granulomatosis, and systemic sarcoidosis must be considered in the differential diagnosis as well. These disorders usually have distinct clinical presentations and the appropriate diagnostic studies can usually prevent their confusion with idiopathic giant cell myocarditis.[7]

Serpiginous areas of necrosis and grayish patches in the myocardium allowing an easy naked eye diagnosis on gross examination are common. [8, 9,10] Histologically, ICGM is confirmed by muscle necrosis with giant cells at the margins. Within the areas of necrosis, a florid histiocytic and eosinophilic cell infiltrate can be seen.[11] Adjacent myocardium can be normal or inflammatory cellular infiltration within the myocardium can also be present. Along with the infiltration, multinucleated giant cells and granulomas interspersed with lymphocytes can also be seen. The presence of eosinophils has been noted in most cases. There was typical absence of sarcoid-like granulomas. [12]

Of the 377,841 cases of autopsy taken from the Annuals of Autopsy Records for Japan from 1958 to 1977, 25 cases (0.007%) were recorded as IGCM. The youngest reported patient was of six weeks age. [13] The oldest reported patient was 88 years old.[14] majority of cases of giant-cell myocarditis occur in otherwise healthy persons (81 percent).The mean age in a few case series has been observed as 42.6 to 57 years. majority of cases of giant-cell myocarditis occur in otherwise healthy persons (81 percent).

Although it is referred to as idiopathic several lines of evidence suggest that giant-cell myocarditis is an autoimmune disorder dependent on CD4-positive T lymphocytes. Experimental giant-cell myocarditis can be produced in Lewis rats by autoimmunization with myosin.[15,16] Both human and experimental giant-cell myocarditis are characterized by an infiltrate of T lymphocytes and histiocytes. Giant-cell myocarditis in the native heart is distinct from lymphocytic myocarditis, in that giant-cell myocarditis has a more fulminant clinical course.

Symptomatic individuals usually present with cardiac failure, acute coronary syndromes, dysrhythmias, heart block, dyspnea, fatigue, febrile illnesses, cardiogenic shock or sudden death. IGCM is one of the causes for refractory cardiac failure. Diagnosis in the living is by endomyocardial biopsy, apical-wedge sampling or histology of the explanted heart. [12]

The minimum information available on the condition is from the various case reports and case series. Better understanding of the etiopathophysiology will enable appropriate management of the condition.

Conclusion

Idiopathic giant cell myocarditis is a rare disease but an important cause of sudden death in young individuals. Because of the rarity of procedures like endomyocardial biopsy and apical wedge sampling in our country, most of the cases go undiagnosed, while the patient could still be saved. In cases of sudden death in young individuals, without any known cause, a careful search for gross and histological evidence of idiopathic giant cell myocarditis is essential.

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