Case Report

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Idiopathic Giant Cell Myocarditis: A Report of 2 Cases

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ABSTRACT

Two cases of idiopathic giant cell myocarditis are reported for their rarity .The first case was of a 47 years old male brought to hospital with a history of uneasiness and sudden death. The second case was of a 55 years old male. Gross examination of heart showed whitish fibrotic areas. Microscopy of the heart in both cases revealed large confluent areas of myocyte necrosis along with plenty of giant cells and lymphocytes and occasional eosinophils. Ziehl Neelsen (ZN) and periodic acid Schiff (PAS) stains were negative. There were no asteroid bodies or Schumann bodies in the giant cells.

Keywords: *Myocarditis, Giant Cell Myocarditis, Sudden Cardiac Death.*

Introduction

The term myocarditis may be applied when histological examination shows evidence of an inflammatory process in the myocardium.On microscopy, myocarditis is classified into acute nonspecific myocarditis and granulomatous myocarditis, which includes idiopathic giant cell myocarditis, tuberculous, fungal, rheumatic, syphilitic myocarditis, and sarcoidosis. We report 2 cases of idiopathic giant cell myocarditis which are presented for their rarity.

Case Report

The first case was of a 47 years old male brought to hospital who had died suddenly. There was a prior history of uneasiness. On gross examination of heart, the weight was 440gm. Left ventricular thickness was 2.2 cm and right ventricular thickness was 0.6cm. Coronary arteries were patent. Serial transverse cut sections showed extensive whitish firm areas in the left ventricle on anterior and posterior walls and interventricular septum(Fig 1). Microscopy of the heart revealed large confluent areas of myocyte necrosis along with plenty of giant cells and lymphocytes and occasional eosinophils(Fig 2). Spleen showed a whitish nodular lesion 1x1 cm in size which on microscopy showed many giant cells and lymphocytes. The liver showed an occasional area of necrosis with giant cells and lymphocytes. ZN, PAS and silver methenamine (SM) stains were negative. The giant cells did not show asteroid bodies or Schaumann bodies.

The second case was of a 55 years old male who had died suddenly. There was a prior history of hematemesis. The weight of the heart was 200 grams. Gross examination of the heart revealed large whitish fibrotic areas in the anterior and posterior wall of left ventricle. Left ventricular thickness was 1.5cm, right ventricular thickness was 0.5cm. Microscopy of the heart revealed large confluent areas of myocyte necrosis along with plenty of giant cells and lymphocytes and occasional eosinophils. ZN, PAS and SM stains were negative. The giant cells did not show asteroid bodies or Schaumann bodies.

Viscera was examined grossly and microscopically in the department of Pathology, B J Government Medical College, Pune, Maharashtra. Medicolegal post-mortem was performed in the department of forensic medicine and viscera was sent for histopathologic examination. Routine tissue processing and H& E (hematoxylin and eosin) stain was used for microscopic examination. ZN (Ziehl Neelsen) and PAS (Periodic Acid Schiff) stains were used to identify acid fast bacilli and fungi if any.

Discussion

Idiopathic giant cell myocarditis (IGCM) is a rare condition. The incidence of IGCM has been reported as 0.007% in the literature.[1] We encountered 2 cases of IGCM out of 2040 autopsies in 2015 (0.098%). The age at presentation is variable, with an average age of 37 years and 48 years. [2] In our cases, there was no history of preceding illness in one case and in the second case there was a history of hematemesis which was followed by sudden death.

IGCM is frequently fatal and patients usually die of heart failure or ventricular arrhythmias unless cardiac transplantation is performed. Diagnosis is based on histological findings at autopsy or endomyocardial biopsy. [3] The presence of multinucleated giant cells along with a large number of lymphocytes, eosinophils and plasma cells make this a specific group among the myocarditides. In the present cases, the diagnosis of IGCM was inferred after ruling out tuberculosis and fungal infections using

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Fig. 1: Whitish fibrotic areas in left ventricular myocardium.

ZN, PAS and SM stain. There were no well delineated granulomas. Giant cells did not contain Schaumann bodies or asteroid bodies . Hence the diagnosis of sarcoidosis was not considered.

The giant cells in IGCM are believed to be of myogenic origin; however some of the giant cells are also believed to be of histiocytic origin. The lymphocytes in IGCM were found to be predominantly CD8+ and those of sarcoidosis were found to be CD4+. [4]IGCM has been associated with autoimmune disorders in 19 percent of cases.[3,5]Amongst our cases, extracardiac involvement was present in one case, in which granulomas were present in the spleen and liver. Granulomas in liver in cases of IGCM have also been reported in the literature.[6]

Conclusion

IGCM is a rare condition which is frequently fatal, and shows multinucleated giant cells on histopathological examination**.**

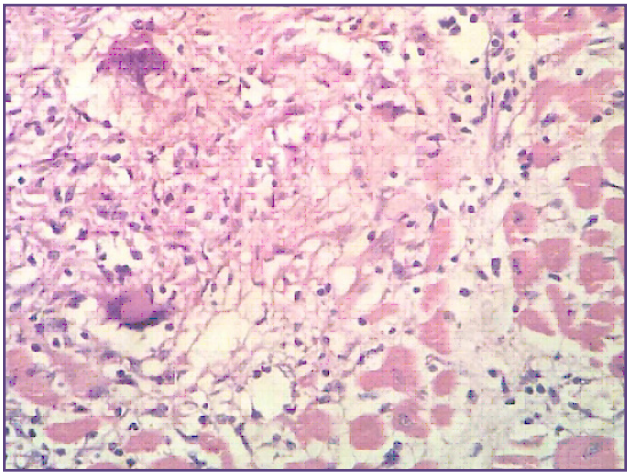

Fig. 2: Microscopy of myocardium with giant cells and plenty of lymphocytes. (H&E stain, 400x)

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