International Journal of Health Sciences and Research
www.ijhsr.org ISSN: 2249-9571

Case Report

Idiopathic Giant Cell Myocarditis: A Rare Diagnosis on Autopsy

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Received: 04/01/2016 *Revised: 20/01/2016* *Accepted: 26/01/2016*

ABSTRACT

Idiopathic giant cell myocarditis is a rare and frequently fatal disorder usually affecting healthy adults. Etiology of idiopathic giant cell myocarditis is largely unknown and it is considered as one of the cause of unexpected sudden cardiac death in healthy adults. We present a case of 50 years old male who died suddenly. An autopsy was conducted. Heart along with other internal organs was sent for histopathological examination. Histopathological examination revealed presence of mixed inflammatory infiltrate along with multinucleate giant cell; consistent with giant cell myocarditis. The present case highlights the idiopathic giant cell myocarditis as a rare fatal diagnosis made on autopsy.

Keywords: Sudden death, giant cell myocarditis, idiopathic.

INTRODUCTION

Giant cell myocarditis is a rare variant of myocarditis with a very fulminant and fatal course, usually affecting young to middle aged adults. [1,2] Etiology of giant cell myocarditis is unknown. However, it has found to be associated with autoimmune diseases like rheumatic disease, myasthenia gravis, thymoma, SLE, thyroiditis, dermatomyositis, pernicious anaemia and ulcerative colitis. [1] Histopathological features are characterized by mononuclear inflammatory cells along with multinucleate giant cells and histiocytes. Degenerated muscle cells along with fibrosis may be present. Clinically the disease is manifested by cardiac failure, cardiac arrhythmias, ECG changes suggestive of cardiac infarction or sudden unexpected death. [3] We report a rare case of idiopathic giant cell myocarditis in a 50 years old healthy male who died suddenly.

CASE REPORT

A 50 years old male died suddenly. A medicolegal autopsy was conducted to know the cause of sudden death. External examination did not show any obvious abnormality. Heart and other internal organs including pieces of lung, liver, spleen and kidney were sent for histopathological examination. Medical history reviewed for any previous illness. There was no history or medical records suggestive of autoimmune disorder, connective tissue disorder or any infectious etiology etc.

On gross examination heart weighed 370 grams. (Fig.1) Both ventricular cavities were normal. The left and right ventricular wall thickness measures 1.4 cms and 0.6cms respectively. Papillary muscles, chordae tendineae, atria, orifices and valves were normal. (Fig.2) Coronary vessels were patent and traced as far as possible for any thrombus. Cut sections of other organs (pieces of

lung, liver, spleen and kidney) were unremarkable.

Fig. 1. Gross appearance of heart- unremarkable.

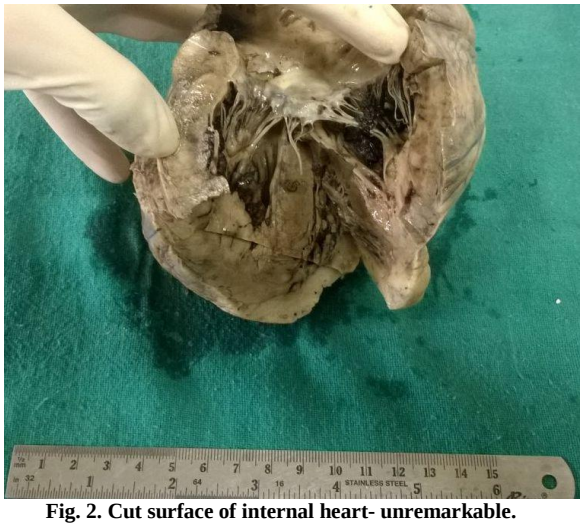

Fig. 2. Cut surface of internal heart- unremarkable.

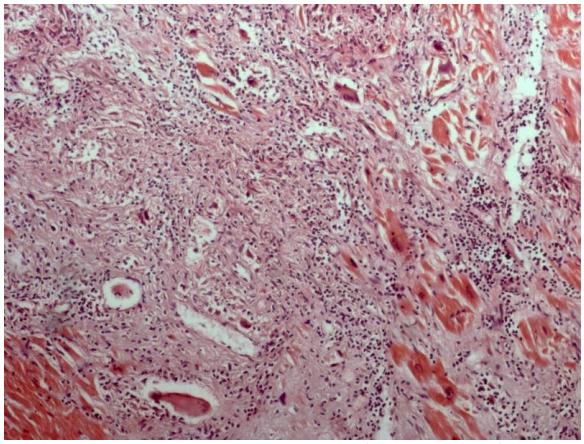

Fig. 3. Microsection revealed mixed inflammatory infiltrate with giant cells in between cardiac muscle. (H&E, 100X)

Representative microsections examined from various portions of heart showed areas of necrosis, mixed inflammatory infiltrate comprising of lymphocytes, plasma cells and multinucleate giant cells.(Fig.3) Special stained sections for acid fast bacilli were negative. No epithelioid cell granuloma or caseous necrosis was noted. No fungi were demonstrated on PAS and GMS stain. Sections examined from other organs like lungs, liver, spleen and kidney were found to be unremarkable. Thus the final diagnosis of idiopathic giant cell myocarditis was suggested.

DISCUSSION

Myocarditis is a pathologic entity in which infectious microorganisms and/or inflammatory processes cause myocardial injury. A rare morphologic type of the disease is idiopathic giant cell myocarditis. [4] The patients are usually young healthy individuals with equal sex incidence and may present with symptoms like dyspnea, peripheral edema, palpitation, arrhythmias or sudden cardiac death. [5]

Sudden cardiac death is defined as an unexpected death which results from cardiac causes in individuals without heart disease or with an early after symptom onset, usually within 1 hour. Idiopathic giant cell myocarditis is a rare clinicopathological entity known to cause death in more than half of the cases of sudden cardiac death. [6] The exact etiology and pathogenesis of giant cell myocarditis is not known.

Approximately 20% patients may have associated autoimmune disorders, mostly inflammatory bowel disease like ulcerative colitis, rheumatic disease, SLE, myasthenia gravis, thymoma etc. It has been postulated to be autoimmune in nature, mediated by T-lymphocytes and tumor necrosis factor-α, the latter stimulating the multinucleation of macrophages. Giant cell myocarditis has to be distinguished from the other forms of myocarditis in view of the difference in prognosis. [5]

Ventricular endomyocardial biopsy might be useful in the diagnosis, with a

high sensitivity rate of 82-85%. However, more microsections need to be studied to rule out sampling error or other causes of myocarditis. [5] Tubercular and cryptococcal myocarditis also has giant cells within the granulomatous lesions. Special stains for the organisms should be performed to rule out tubercular and cryptococcal myocarditis. Rheumatic myocarditis show characteristic interstitial granulomas with giant cells. Other rare variant like syphilitic myocarditis and systemic sarcoidosis usually have distinct clinical profile and diagnostic studies can usually differentiate from idiopathic giant cell myocarditis. [4]

CONCLUSION

Idiopathic giant cell myocarditis is a disease of relatively young to middle aged healthy adults which presents as sudden cardiac death as seen in our case. So, idiopathic giant cell myocarditis must be considered as differential diagnosis in cause of unexplained sudden death in healthy adults.

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How to cite this article: Singh G, Kalyan S, Parmar P et al. Idiopathic giant cell myocarditis: a rare diagnosis on autopsy. Int J Health Sci Res. 2016; 6(2):424-426.

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