

Giant Cell Myocarditis: A Rare Diagnosis Made On Autopsy

K Jhaji, S Nibhoria, S Sandhu

Citation

K Jhaji, S Nibhoria, S Sandhu. *Giant Cell Myocarditis: A Rare Diagnosis Made On Autopsy*. The Internet Journal of Pathology. 2010 Volume 13 Number 1.

Abstract

An autopsy case report of 60 year old female patient who died suddenly after getting in minor altercation with neighbors. An autopsy was conducted and the heart was sent for histopathological examination. Histopathological examination revealed presence of multinucleated giant cells, lymphocytes, neutrophils, plasma cells and macrophages around areas of necrotic zone with absence of granulomas. No previous history of tuberculosis was known. The present case highlights the giant cell myocarditis as a rare fatal diagnosis made on autopsy.

INTRODUCTION

Giant cell myocarditis is rare morphologically distinct form of myocarditis with a very fulminant and fatal course.^[1,2] This disease lacks granuloma formation and is now considered to be a separate entity from granulomatous myocarditis. Etiology of giant cell Myocarditis is unknown till date however in some patients there is an association with autoimmune diseases like rheumatic disease, myasthenia gravis, thymoma, systemic lupus erythematosus, thyroiditis, dermatomyositis, pernicious anemia and ulcerative colitis. The purpose of this case report is to highlight a very rare diagnosis which has fatal outcome and is diagnosed incidentally on autopsy..

CASE REPORT

A 60 year old female patient collapsed suddenly after getting in a minor altercation with neighbors. A medico legal autopsy was conducted and the heart was sent for histopathological examination. There was no external injury and patient was moderately built and nourished with no previous history of any kind of major ailment. The cause of death was ascertained only on autopsy and histopathological examination of heart. On gross examination of the heart it weighed 420 gms. Both ventricles were normal and the left ventricular wall thickness was 1.6 cms and right ventricular wall thickness is 0.8 cms . multiple irregular grayish white areas were noted on both ventricular walls . Papillary muscles , chordae tendinae, atria, orifices, ostia and valves were normal. Coronaries were patent [Figure 1].

Figure 1

Figure 1 Heart showing irregular grayish white areas on ventricular wall



Microscopic examination of several sections taken from ventricular wall showed necrosis. Diffuse inflammatory infiltrate comprising of lymphocytes, neutrophils, plasma cells, histiocytes and multinucleated giant cells alongwith degenerated myocytes and some showing reactive changes [Figure 2, 3] .No epithelioid cell granulomas or caseous necrosis noted. Acid fast bacilli were not seen. Sections from atria, valves and coronaries were unremarkable.

Figure 2

Figure 2 Section shows diffuse inflammatory infiltrate , multinucleated giant cells and reactive changes in myocytes and no granulomas (H&E, x100)

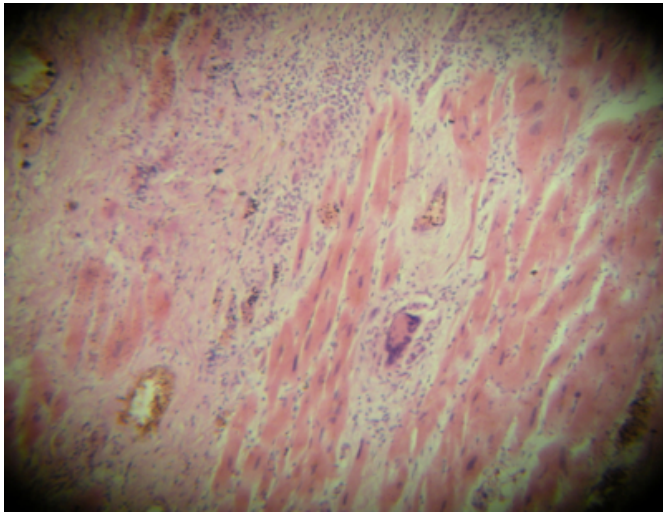
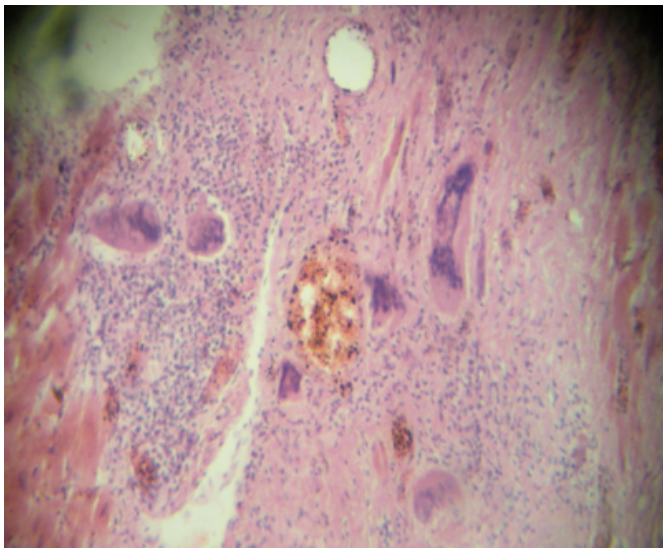


Figure 3

Figure 3 Section shows diffuse inflammatory infiltrate and multinucleated giant cells (H&E, x100)



DISCUSSION

Giant cell Myocarditis is rare, morphologically distinct form of Myocarditis with a Fulminant and fatal course.^[1,2] It was first described by Saltykow in 1905 in a 37 year old man who died suddenly after surgical drainage of an abscess.^[5] In early part of the twentieth century the term giant cell Myocarditis was used to describe both granulomatous and diffuse inflammatory myocardial infiltrates that contained

multinucleated giant cells.^[6,7] Teslak first distinguished the term and called it giant cell Myocarditis.^[8] It is a pathological diagnosis . A diffuse or multifocal inflammatory infiltrate consists of lymphocytes, histiocytes, plasma cells, eosinophils and numerous multinucleated giant cells and absence of true granulomas.

It is a rare condition and the incidence is very low. In a Japanese autopsy registry the incidence was 0.007% (25 of 377841 cases from 1958 to 1977).^[3] The incidence of giant cell Myocarditis was similarly low 3 of 12815 necropsies from 1950 to 1963 at Oxford Infirmary.^[4] Cooper et al reported the largest series consists of 63 cases from multiple centers world wide.^[9] The median age of presentation was 42 years (range 15 to 69 years) but patients younger than 15 years and older than 70 years have been reported.^[10, 11, 12,13] Equal incidences have been reported in males and females.

It is a fatal disorder with short course generally less than 3 months and diagnosis is made incidentally on autopsy. All most all cases are reported on autopsy which highlights the importance of autopsy in this rare myocardial disorder.

References

1. Wilson MS, Barth RF, Baker PB et al: Giant Cell Myocarditis .AmJ Med 79:647,1985
2. Cooper LT, Berry GJ, Rizeq M, Schroedar JS: Giant Cell Myocarditis. J Heart Lung Transplant 14:394,1995
3. Okada R, Wakafuji S: Myocarditis in autopsy. Heart Vessels Suppl 1985; 1:23-29
4. White head R: Isolated Myocarditis. Br Heart J 1965; 27:220-230
5. Saltykow S: Ueber diffuse Myocarditis. Virchows Arch Pathol Anal 1905; 182:1-39
6. Saphir O: Myocarditis; General review, with analysis of 240 cases. Arch Pathol 1941;32:1000-1051;1942;33:88-137
7. Kean BH, Hoekenga MT: Giant cell myocarditis . Am J pathol 1952;28:1095-1105
8. Teslak H: Giant cell versus granulomatous myocarditis. Am J Clin Pathol 1956;26:1326-1333
9. Cooper LT, Berry GJ, Shabetai R: Idiopathic giant cell myocarditis-natural history and treatment. N Engl J Med 336:1860,1997
10. Goldbeg GM: Myocarditis of giant cell type in an infant. AmJ Clin Pathol 1995; 25:510-513
11. Whitehead R; Isolated myocarditis. Br Heart J 1965; 27:220-230
12. Fenoglio JJ Jr , Ursell P C , Kellogg CF et al . Diagnosis and classification of myocarditis by Endomyocardial biopsy .N Engl J Med 1983 ,308 :12- 18 .
13. Litovsky SH , Burke AP , Virmani R . Giant cell myocarditis : an entity distinct from sarcoidosis characterized by multiphasic Myocyte destruction by cytotoxic T cells and histiocytic giant cells .Mod Pathol 1996 , 9 : 1126- 1134 .

Author Information

Kanwardeep Kaur Jhaji

Assistant Professor, Department of Pathology, GGS Medical College

Sarita Nibhoria

Associate Professor, Department of Pathology, GGS Medical College

Samarjit Kaur Sandhu

Professor, Department of Pathology, GGS Medical College