

## Giant Cell Myocarditis: A Case Report.

Afifa Azhar<sup>1\*</sup>, Sunitha S Vernekar<sup>2</sup>, and Zaheer Abbas Ali Khan Pathan<sup>1</sup>.

<sup>1</sup>Department of Pathology, SDM College of Medical Sciences and Hospital, Dharwad, Karnataka, India.

<sup>2</sup>Department of Pathology, Karnataka Institute of Medical Sciences, Hubli, Karnataka, India.

### Case Report

Received: 13/03/2014

Revised: 21/03/2014

Accepted: 27/03/2014

#### \*For Correspondence

Department of Pathology, SDM  
College of Medical Sciences and  
Hospital, Dharwad, Karnataka,  
India.

Mobile: +91 9480575596

**Keywords:** Giant cell myocarditis,  
arrhythmias, congestive heart  
failure.

#### ABSTRACT

Giant cell myocarditis (GCM) is a rare and morphologically distinctive form of myocarditis. It is a disease of young healthy adults. It is very rare in pregnant women. It may present as a fatal disease with initial symptom itself as in our case. About half of the patients suffer a sudden death due to arrhythmias, congestive heart failure and the disease is usually diagnosed at autopsy.

#### INTRODUCTION

Giant cell myocarditis(GCM) is a rare and morphologically distinctive form of myocarditis, characterized clinically by a rapid downhill course resulting into death and histologically widespread cellular infiltrate composed of multinucleate giant cells interspersed with lymphocytes, plasma cells, eosinophils and macrophages and at least focal but frequently extensive areas of necrosis. About half of the patients suffer a sudden death due to arrhythmias, congestive heart failure and the disease is usually diagnosed at autopsy <sup>[1]</sup>. GCM is seen occasionally in association with thymoma, sarcoidosis, tuberculosis, systemic lupus erythematosus, thyrotoxicosis and inflammatory bowel disease <sup>[1,2]</sup>.

Extensive search in English literature yielded two reported cases of GCM in pregnant women with ours being third case.

#### Case Report

A 29-year pregnant lady presented to the obstetrician with history of 2 months amenorrhea. She was diagnosed to be pregnant. While on the way back to her home, she complained of breathlessness, collapsed and died. There was no history of cough, pleuritic pain, fever, allergy, intoxication, vascular/respiratory disease in the past.

#### Pathologic features

After autopsy specimen of heart, uterus with bilateral adnexa, brain and thyroid were sent for histopathological examination. Specimen of heart weighed 280gms. External surface was unremarkable. Multiple grey white foci were noted throughout the left ventricular myocardium (Fig.1). Coronary vessels were patent and the great vessels appeared normal. There was no cardiomegaly, cardiac hypertrophy, congenital or acquired anomalies of the heart.

Specimen of uterus was bulky weighing 300gms and measured 14x10x6cms with bilateral adnexae. Cut section showed gestational sac and thickened endometrium. Bilateral adnexae were unremarkable.

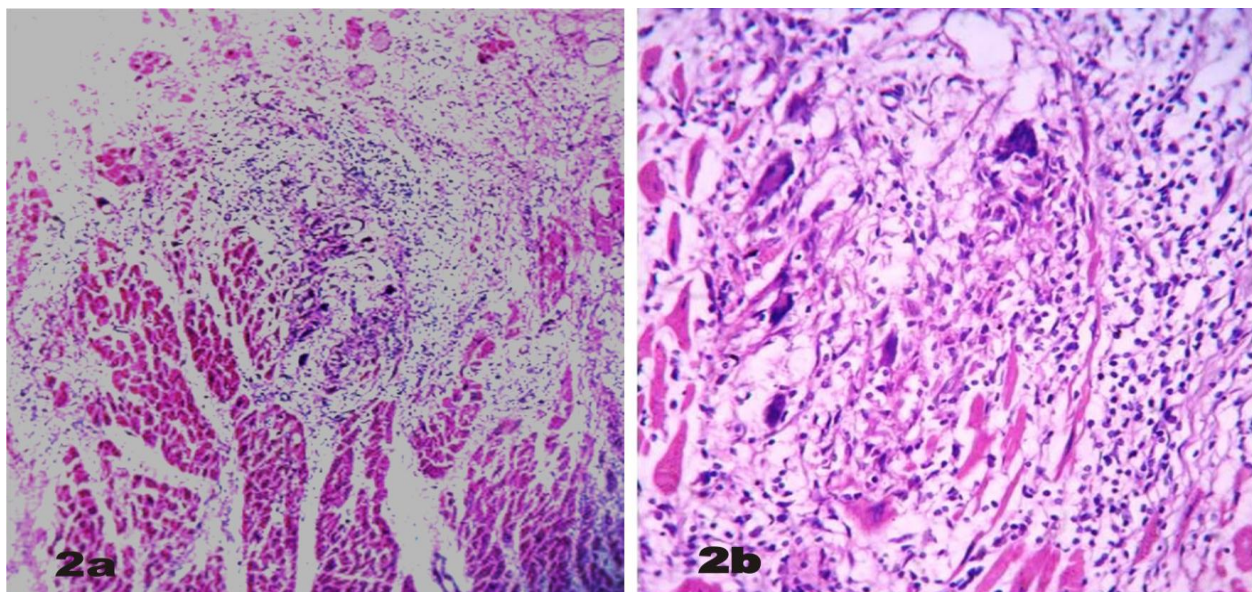
Brain and thyroid appeared unremarkable.

Multiple sections from the myocardium of the left ventricle showed scattered, diffuse and patchy areas of granulomas consisting of numerous multinucleated giant cells, plasma cells, lymphocytes, neutrophils, histiocytes, eosinophils and fragments of degenerated or necrotic myocardial fibers (Fig.2). H&E, Ziel-Nelson, PAS, GMS and Gram's stains failed to demonstrate bacteria, parasite, fungi or protozoa. No significant lesions were present in any other organs. A diagnosis of giant cell myocarditis was made on histopathology examination. The endometrium was of secretory type with decidual change.

**Figure 1: Left ventricular myocardium showing multiple areas of hemorrhage**



**Figure 2: 2a Showing myocardial necrosis and granulomas in the myocardium (H&E 10X) and 2b. Showing giant cells in the granuloma (H&E 40X)**



## DISCUSSION

Giant cell myocarditis is a rare and morphologically distinctive form of myocarditis. In 1905, Saltykow reported the first case of GCM which seemed idiopathic in nature. [3,4] Idiopathic GCM and giant cell myocarditis are used as synonyms by different workers [3].

GCM is a disease of young healthy adults. Male and female are equally affected [5]. The age of onset varies from six weeks to 84 years, with mean age of 42.6 years [3,4]. Generally GCM presents as heart failure in 70% cases with dyspnea, hypotension, orthopnea and peripheral edema. Occasionally the initial presentation may be intractable arrhythmias, complete heart block, symptoms mimicking myocardial infarction or sudden death [5, 6]. A diagnosis of GCM should be considered in patients with declining left ventricular function and ventricular

arrhythmias despite usual supportive treatment with inotropic and antiarrhythmic agents. In our case the initial symptom had turned out to be fatal disease.

Twenty percent of cases are seen in association with autoimmune diseases, such as ulcerative colitis, crohn's disease, Hashimoto's thyroiditis, Rheumatoid arthritis, pernicious anemia and Takayasu's arteritis. A disease similar to GCM has been described in rats after inoculation with cardiac myosin, which suggests an autoimmune origin [5,6].

The differential diagnosis of GCM includes sarcoidosis, rheumatic fever, and rheumatoid arthritis, giant cell containing tumors, syphilis, brucellosis and tularemia. Eosinophils, myocyte damage and foci of lymphocytic myocarditis are more frequent in idiopathic GCM, while granulomas and fibrosis are more frequent in cardiac sarcoidosis [3].

Litovsky SH et al described histologic phases as acute, healing and healed phases. The acute phase is characterized by extensive infiltration of lymphocytes, eosinophils, macrophages and macrophage derived KP-1 positive giant cells (GCs) associated with myocyte necrosis and no granulomas. The healing phase with granulation tissue, moderate macrophage GCs and scattered KP-1 negative myogenic GCs. A healed phase shows a dense scar with no GCs [3].

GCM and idiopathic myocarditis of pregnancy differ in some aspects. The latter condition is seen in post partum period and predominantly in Negroes. The myocardial changes consist of degeneration of myocardium with interstitial edema and cellular infiltrate predominantly of lymphocytes and macrophages. Giant cells were rarely observed [7].

The course of the GCM is invariably fatal unless short term transplantation is done or strong immune-suppressive therapy is started along with some ventricular assist device. Cooper LT et al reviewed 63 cases of GCM and reported an overall mortality rate of 89% with a median survival of 5.5months regardless of the fact whether a patient underwent transplantation or not [5].

## CONCLUSION

In GCM myocardial fibers undergo unusual type of degeneration necrosis, with cellular infiltrate of lymphocytes, eosinophils and giant cells. Further investigations are required to determine the cause particularly the possible role of virus hypersensitivity or autoimmune reaction. GCM is a rare disease by itself and still very rare in pregnant women. In literature we could find two cases, ours is the third case. The GCM may present as a fatal disease with initial symptom itself as in our case.

## REFERENCES

1. Murty OP. Giant Cell Myocarditis with Cardiac Tamponade -A Rare Combination. The American J Forensic Med Pathol. 2008; 29:245-248.
2. Singh A M et al. An Autopsy Case of Sudden Death in a Patient with Giant Cell Myocarditis. The Indian J Chest Dis Allied Sci. 2003; 45: 195-198.
3. Konapur PG, Dhaded AV, Udaykumar M. Idiopathic giant cell myocarditis – a case report and review of literature. Indian J Pathol Microbiol. 2003; 46(2):241-243.
4. Momin YA, Raghuvanshi SR, Lanjewar DN. Idiopathic Giant Cell Myocarditis Leading to Sudden Death – A Case Report. Bombay Hospital J. 2008;50(2):288-289.
5. Haelst PL, Rossem M, Valentijn R M and Bejer G J P. Giant cell myocarditis: a fatal cause of dyspnea in pregnancy. European J Obstetr Gynecol Reprod Biol.2001;100: 105-107
6. Koul D et al. Fulminant Giant Cell Myocarditis and Cardiogenic Shock: An Unusual Presentation of Malignant Thymoma. Cardiol Res Pract.2010;2010:1-4
7. Pyun KS et al. Giant Cell Myocarditis – Light and Electron Microscopic Study. Arch Pathol. 1970;90:181-188.