Giant-cell myocarditis


This is a case report of a 32-year-old Thai woman who was brought in dead at the emergency room of Chulalongkorn hospital. The autopsy revealed a typical giant-cell myocarditis on gross examination which was confirmed on histopathology. Special stains, were done and a review of studies by many authors, was made to confirm the myogenic origin of giant cells and to exclude any infective or other possible causes, such as sarcoidosis etc. The etiology of Giant-cell myocarditis is still unknown. Giant-cell myocarditis is very rare but is one of the causes of sudden and unexpected death. This is the only case found in the Forensic Medicine Department of Chulalongkorn hospital between 1979-1989. It may even be the first case report of this condition in Chula Med J. (computer search in Medline)

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Received for publication. March 13, 1990.

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ประยุทธ์ บุญยอด กล่าวเนื้อหัวใจอัคลาสนิสัยเด่นเหนือ一切. จุฬาลงกรณ์ราชวิทยาลัย 2533 มีอธิบาย:
34 (6) : 467-472

ทุกรอยอาจพยากรณ์ว่าจะเป็นผลไม่หวังผลให้เกิดการติดตามหรือผลประโยชน์ให้เกิดการติดตามหรือผลประโยชน์ให้เกิดการติดตามหรือผลประโยชน์ให้เกิดการติดตามหรือผลประโยชน์ให้เกิดการติดตามหรือผลประโยชน์ให้เกิดการติดตามหรือผลประโยชน์ให้เกิดการติดตามหรือผลประโยชน์ให้เกิดการติดตามหรือผลประโยชน์ให้เกิดการติดตามหรือผลประโยชน์ให้เกิดการติดตามหรือผลประโยชน์ให้เกิดการติดตามหรือผลประโยชน์ให้เกิดการติดตามหรือผลประโยชน์ให้เกิดการติดตามหรือผลประโยชน์ให้เกิดการติดตามหรือผลประโยชน์ให้เกิดการติดตามหรือผลประโยชน์ให้เกิดการติดตามหรือผลประโยชน์ให้เกิดการติดตามหรือผลประโยชน์ให้เกิดการติดตามหรือผลประโยชน์ให้เกิดการติดตามหรือผลประโยชน์ให้เกิดการติดตามหรือผลประโยชน์
Giant-cell myocarditis, considered by some author to be a variant of idiopathic myocarditis\(^1\), is a rare condition which has been reported in the world literature under a variety of names such as idiopathic giant cell myocarditis\(^2\), giant cell granulomatous myocarditis\(^3\), myocarditis of giant cell type\(^4\), granulomatous myocarditis\(^5\), isolated myocarditis\(^6\), isolated granulomatous myocarditis\(^7\), acute isolated myocarditis\(^8\), and giant-cell myocarditis\(^9\)\(^-\)\(^18\) The last name mentioned is the one commonly used by most authors\(^9\)\(^-\)\(^18\).

Like other forms of myocarditis the condition occurs in all ages, from newborn\(^4\) (6 weeks) to the very old\(^5\) (85 yr.) and affects both sexes. And as mentioned above this condition is rather rare, some authors still consider it as extremely rare\(^3\). However the pathological features are unmistakable with serpiginous areas of myocardium necrosis, replaced by the granulomatous-like lesions and fibrosis, respectively.

This report is the first case of this kind found in the department of Forensic Medicine, Chulalongkorn hospital.

**Case report**

A 32-year-old Thai woman was frought in dead at the emergency room of Chulalongkorn hospital on Dec. 10, 1989. No history taking nor physical examinations were done.

**Autopsy examination** (performed about 10 hours after death)

Gross examination revealed a deceased Thai female who weighed 41 kilograms and her height was 153 centimeters. No abnormalities were found.

**Internal examination**

- **Brain**: weighed 1,210 gm. Moderate congestion and edema of the brain parenchyma was seen.
- **Heart**: weighed 350 gm. There was no excess pericardial fluid. The epicardium was smooth and shiny and contained scanty amount of fat. There was a severe degree of dilatation of the cardiac chambers, especially the left ventricle. The myocardium had an extraordinary appearance, within the ventricular wall, the atrial and interventricular septum, there were numerous patchy and confluent areas of greyish discoloration of the myocardium, alternated with extensive fibrosis. No areas of recent infarctions were seen, neither were there mural thrombi. The valvular endocardium was smooth, glistening, and semitransparent. (figure 1 A,B) The valvular circumferences were: aortic 5.5 cm.; mitral 8 cm.; tricuspid 11 cm.; pulmonary 7 cm. The right ventricle wall measured 0.5 cm. in thickness, while the left ventricular wall measured 1.3-1.5 cm. in thickness.

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**Figure 1.** Show gross appearance of the heart. A note the severe dilatation of the cardiac chambers (Global-shape) B. Lt. ventricle shows dilatation. Note the numerous patchy and confluent areas of greyish discoloration in the myocardium. (arrows)
The interventricular septum measured 1.6 cm. Microscopic examination revealed large, diffuse, and focal areas in which the myocardium fibers had been completely replaced by fibrous tissue, some of which, has undergone hyaline degeneration. Other areas revealed extensive necrosis of the myocardial fibers, infiltrated with small round cells (mainly lymphocytes and plasma cells) and giant cells scattered in an apparently indiscriminate fashion. (figure 2 A,B\(^{1,2}\)) The giant cells exhibited marked variation in size, shape and appearance, several of these seemed to represent transitional forms of muscles fibers. The giant cells contained between 5-30 nuclei with or without nucleoli and had pale cytoplasm that was sometimes vacuolated. The nuclei were vesicular or pyknotic in appearance, and were situated both centrally and peripherally throughout the cells. (Langhan’s type, foreign bodies, Osteoid type) Some astersid bodies were seen. (Figure 2C, 1, 2)

Figure 2. A. Myocardium show areas of necrosis, replaced by granulomatous-like lesions (H&E × 200)
B. Giant cells of various types, Langhans, (L) foreign-body (F) (H&E × 200)

Figure 2c(1) Granulomatous lesion near epicardium showing asteriod bodies in the giant-cells (arrows) (H&E × 100). (2) Higher power (H&E × 400)
Special stains of the muscle lesions were done such as Acid fast bacilli, Gram (for bacteria), and spirochetes (Levaditi), all were negative.

Phosphotungstic Acid Hematoxylin and Toluidine failed to demonstrate striation in the giant cells. No areas of caseation necrosis were seen.

Both Lungs together weighed 850 gm. The lung parenchyma revealed areas of congestion alternated with areas of pale color. Microscopic examination revealed areas of widening of the alveolar wall containing round cells infiltration. Another area revealed organized pneumonia. Swab culture from the lung yielded a streptococcus of moderate growth.

Liver: weighed 1,200 gm. There were irregular, dark red areas of severe, extensive congestion and fatty degeneration on microscopy.

The rest of the organs were unremarkable.

Postmortem analysis for blood alcohol yielded 194.2 mg%. No analgesic, benzodiazepine, or barbiturate group were found in the liver, kidney and stomach contents Serum Anti-HIV was negative. (The patient came form nowhere, with no past history, and unknown occupation)

Discussion

According to Saphir, giant-cell myocarditis was first described by Saltykor in 1905. Between then and 1969, there were less than 30 cases reported in the world literature (3) and this included the first case report in Thailand from the Women’s hospital) by Vinijchaikul K(13) in the year 1966. By the year 1970 Dechakaisaya et al(14) reported for the second time in Thailand. They reported 3 cases from the pathology departments of Chulalongkorn hospital together with Police general hospital.

The diagnosis of giant-cell myocarditis is usually made at autopsy because of sudden and unexpected death, occasionally the diagnosis is made as an incidental finding together with myositis associated with Thymoma and myasthenia gravis etc.

The etiology of giant-cell myocarditis remains obscure. The clinical such as course of the disease is not known. On anatomic finding staining for acid fast bacilli, bacteria, and spirochete must be done to rule out such causes. And also this case report had done. Pyun et al(16) reported studies by light & electron microscopy and they reached the conclusions that the giant cells were derived from myocardial fibres and ruled out a viral aetiology. However, this case report failed to demonstrated myofibrils or intercalated disk in the giant cell (Toluidine blue and PTAH) as poision and Pyun had done.

The clinical manifestations of giant-cell myocarditis include persistent cardiac arrhythmia, rapidly progressive cardiac failure, or sudden death. Most of the cases are isolated findings but is may be found in association with a thymoma or myasthenia gravis as mentioned above. According to Dechakaisaya and other authors(16-18) giant-cell myocarditis had been reported following sulphonamide and penicillin therapy and was also found in conjunction with systemic lupus erythematosus as well as other collagen diseases. etc.(19)

In 1980, Tubbs et al(20) reported a case of a 15 year-old boy with giant cell myocarditis in whom he and his co-worker had tried to demonstrate immunocytochemistry for cytoplasmic muramidase. They were successful in showing cytoplasmic muramidase in neutrophils and tissue macrophages but absent in the giant cells. Thus, they concluded that giant-cell myocarditis was separate from sarcoidosis. In the meantime, Singham et al(21) reported the second case of giant-cell myocarditis with complete atrioventricular block. They were able to provide histological prove of atrioventricular and sinoatrial nodes. Giant cell myocarditis is still a rare condition, with only a few more only reports between 1982-1989. Okada et al(18) reported .007 % in their autopopy cases 1985. Recently, Drut RM. and Drut R(22) reported giant-cell myocarditis in a new born with congenital herpes simplex where the found desmin and myoglobin which supported the myogenic origin and negative muramidase and a-L-antichymotrypsin after immunoperoxidase procedure, which excluded a viral origin.

Blood alcohol level of 194.2 mg% would explain severe fatty metamorphosis of the liver as well as pneumonia in this case report. Neither the blood alcohol level nor the pneumonia were the cause of death in this patient. She probably died from cardiac arrhythmia which could occur at anytime from myocarditis.

Acknowledgments

The author is grateful to Dr. Choochart Annoppech, Head department of Forensic Medicine, for help. Thanks are also to Asso. Prof. Prasert Samranvej and to Mr. Chalaw Buanamjued from the Department of Pathology, who performed almost all of the special stains.
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