

Idiopathic giant cell myocarditis in a newborn: case report

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Idiopathic giant cell myocarditis (IGCM) is a rare cause of arrhythmia, heart failure and death in children. The etiology of IGCM is unknown. Endomyocardial biopsy and histological examination are essential for diagnosis. In our case, a one-month-old baby boy with no prior medical history died during breastfeeding. The gross autopsy and drug screen failed to disclose a cause of death. Hemotoxylen Eosin (H&E) stained cardiac tissue slides showed widespread and multifocal necrosis accompanied by mixed inflammatory infiltrate. Admixed with the infiltrate were fibroblastic proliferation and multinucleated giant cells in the absence of sarcoid like granuloma. Although myocarditis is usually thought to be a disease of older children, infants can also be affected. This entity can be the cause of some cases of sudden unexpected death in infancy.

Key words: myocarditis, newborn.

Myocarditis is clinically and pathologically defined as 'inflammation of myocardium.' Although this definition sounds straightforward, the diagnosis and treatment of myocarditis have been controversial¹. The Dallas pathological criteria published in 1986, presented the initial effort to develop standardized diagnostic guidelines for histopathological classification of myocarditis. This attempt to create reproducible histopathologic criteria, made it possible to discriminate myocarditis from ischemic necrosis and other conditions. By this definition, active myocarditis is characterized by an inflammatory cellular infiltrate accompanied by myocyte necrosis, whereas borderline myocarditis shows inflammatory infiltrate but no evidence of myocyte injury².

To diagnose myocarditis, endomyocardial biopsy and histological examination should be performed^{3,4}. Thus, a pathologist is an important member of the multidisciplinary approach to the diagnosis and management of such cases⁵. The role of the pathologist is to establish a histopathologic diagnosis of myocarditis using the Dallas criteria; to exclude other morphological and clinical

mimics of inflammatory myocardial disease, to classify the specific type of myocarditis (eg, lymphocytic, giant cell, hypersensitivity, toxic, infectious, sarcoidosis), and to preserve tissue for research purposes (e.g.: microbiologic, immunohistochemical, molecular)⁶.

A large retrospective pediatric study revealed that myocarditis is the most common cause of sudden unexpected cardiac death in childhood, followed by hypoplastic left heart syndromes and dilated cardiomyopathy⁷. The major etiological agents of myocarditis are viral, bacterial, fungal, protozoal and parasitic microorganisms; toxins, hypersensitivity and immunologic syndromes (Churg-Strauss, inflammatory bowel disease, giant cell myocarditis, diabetes mellitus, sarcoidosis, systemic lupus erythematosus, thyrotoxicosis, Takayasu's arteritis, Wegener's granulomatosis)¹. IGCM is a rare cause of arrhythmia, heart failure and death in children, of which the etiology is unknown⁸⁻¹⁰. The Giant Cell Myocarditis Study Group identified heart failure symptoms as the primary presentation in 75% of patients with giant cell myocarditis⁴. In about 20% of cases, myocarditis is associated with a number of immune diseases^{9,10}.



Figure 1

Fig. 1. On gross examination, the heart weighed 50 g. The left ventricle wall was 1 cm and the right ventricle wall was 0.3 cm in thickness. Intracardiac anatomy was normal as were the origin and anatomy of the coronary arteries

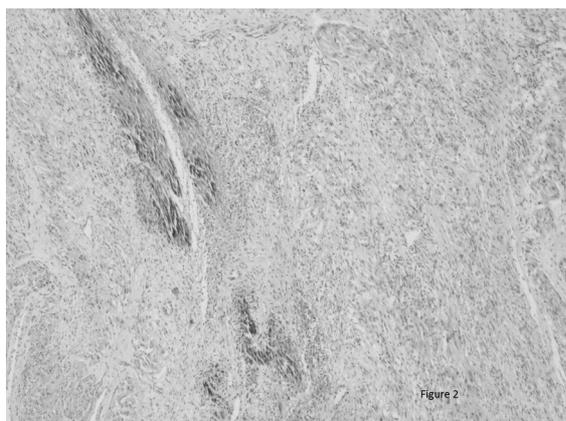


Figure 2

Fig. 2. Widespread and multifocal necrosis of myocytes was observed (H&E, X100)

Case Report

In our case, a one-month-old baby boy died during breast-feeding without any diagnosed disease in advance. Parents stated that they did not notice any significant problem with the baby until the incident. They also report that they have been warned about cardiac arrhythmia by the family physician when they took the baby for routine control. Since they did not pay attention to the warning, no further diagnosis was made. He was the first child of the couple and when family history was enquired they mentioned no other example of similar condition. Additionally the mother had a problem-free pregnancy.

The autopsy was performed in the Forensic Sciences Institution of Turkey, Ankara Head Office. Tissue sampling from main organs was done by a pathologist. Light microscopic

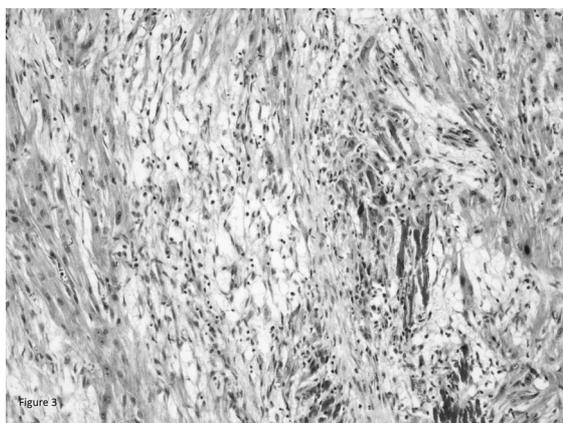


Figure 3

Fig. 3. Admixed with the inflammatory infiltrate were fibroblastic proliferation and multinucleated giant cells in the absence of sarcoid like granuloma (H&E, X200)

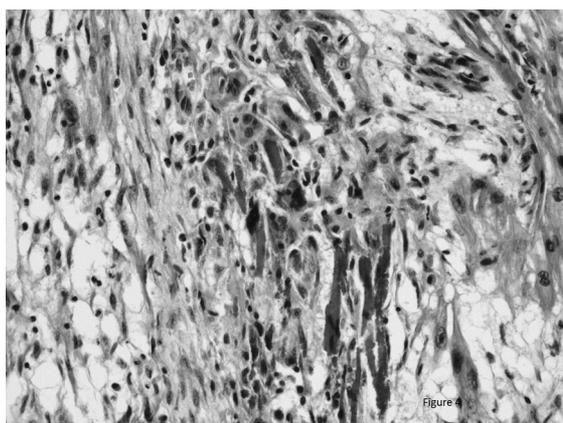


Figure 4

Fig. 4. Granulation tissue composed of mixed inflammatory infiltrate: lymphocytes, histiocytes scarce eosinophilic polymorphocytes and plasma cells. (H&E, X400)

evaluation of the H&E stained slides was performed after tissue processing. On gross examination, the heart weighed 50 g. The left ventricle wall was 1 cm and the right ventricle wall was 0.3 cm in thickness. Intracardiac anatomy was normal as were the origin and anatomy of the coronary arteries (Fig. 1). Neither any gross pathology of the other organs, nor any postmortem toxic substance were detected. When H&E stained cardiac tissue slides were examined by the pathologist, widespread and multifocal necrosis of myocytes was observed. Accompanying mixed inflammatory infiltrate, which is composed of lymphocytes, histiocytes, scarce eosinophilic polymorphocytes and plasma cells, was detected. Admixed with the inflammatory infiltrate were fibroblastic proliferation and multinucleated giant cells in the absence of sarcoid like granuloma (Figs. 2-4).

Microscopic sections of the other internal organs revealed no pathology but congestion¹¹.

Discussion

At the first step of the differential diagnosis, one should keep in mind considering neonatal myocardial infarction and overlap with myocarditis¹².

The differential diagnosis of IGCM includes drug reactions and systemic diseases such as sarcoidosis, Wegener granulomatosis, foreign body reaction, hypersensitivity myocarditis, cardiac lymphoma, fungal myocarditis, tuberculosis and rheumatic carditis. The different clinical presentations and/or histologic findings of these disorders enable pathologists to make a differential diagnosis¹³. Endomyocardial histopathological evaluation is still the gold standard for making the diagnosis of cardiac disease^{1,10}. It establishes to diagnose the disorders of heart such as cardiomyopathies, myocarditis, infiltrative lesions, arrhythmias and drug toxicities¹⁰. But myocarditis can affect only focal areas of myocardium, which is the most crucial handicap of histopathological diagnosis. According to a Mayo Clinic study of endomyocardial samples obtained post mortem from the hearts of patients who died of myocarditis, they reported a false negative rate of 37% for the right ventricle¹⁴. So it is recommended that to reduce sampling error, at least 4 to 5 pieces from each specimen must be obtained⁶.

If there is any suspicion for infection, special stains for microorganisms should be performed¹³.

The etiology of giant cell myocarditis is not known, but mostly predicted to be autoimmune^{3,4,9-11,13}. Experimental studies suggest that genetic factors may play a major role in tendency to disease¹³. IGCM is usually a lethal disorder that generally affects young otherwise healthy people¹³. Unlike lymphocytic myocarditis, which generally has good prognosis, IGCM usually results in death or heart transplantation. The overall rate of IGCM is equal for males and females. Compared to males, IGCM in females may be more strongly associated with autoimmune disease in other organs⁹.

Our case is presented in order to highlight that IGCM is a rare and highly lethal disease which can only be diagnosed by microscopical

examination. The diagnosis of giant cell myocarditis should be considered for all patients with subacute heart failure of unknown cause. Infants, as we present in our case, can also be affected by myocarditis, which is usually thought to be older children's disease. Some cases of sudden unexpected death in children can be caused by this entity.

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