Acute fulminant myocarditis at autopsy: A clinical masquerade?

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Abstract: Background: Myocarditis is a diagnostic challenge in cardiology. The diagnosis is frequently made post-mortem, with no clinical evidence of myocardial failure. Autopsy studies report a frequency of myocarditis ranging from 0.11-0.55% in the general population. Myocarditis is presents with varied clinical manifestations, ranging from asymptomatic to sudden cardiac death, sometimes mimicking the Myocardial Infarction (MI). Case Summary: A 55 years old male presented with sudden onset of chest pain, breathlessness at rest and flu like symptoms 5-6 days prior to admission. There was tachycardia with low Systolic blood pressure and muffled heart sounds. ECG showed ST segment & T wave changes. CK-MB was moderately raised. Though clinically diagnosed as MI and treated, patient expired the next day. On autopsy, gross examination of heart revealed no significant findings. Histological examination revealed fulminant myocarditis involving right ventricle, interventricular septum, left ventricular wall and A-V Node. Conclusion: It is not uncommon to find Myocarditis patients presenting with manifestations of MI and these cases often misdiagnosed as MI. In such cases, myocarditis should be considered as a differential diagnosis. Endomyocardial biopsies and histological examination of the myocardium is absolutely necessary for reliable diagnosis of the disease. Keywords: Myocarditis, Myocardial infarction, Autopsy.

Introduction

Myocarditis is one of the most challenging diagnostics in cardiology [1]. The entity is rarely recognized, the pathophysiology is poorly understood and there is no commonly accepted diagnostic gold standard [2]. Autopsy studies report a frequency of myocarditis ranging from 0.11-0.55% in the general population [3]. Myocarditis is commonly mild or subclinical disease, it is not uncommon to find patients presenting with chest pain, ST & Q wave abnormalities, segmental left ventricular dysfunction and mild elevation of Creatinine Kinase (CK). These patients have often been misdiagnosed as myocardial infarction (MI). In such cases, myocarditis should be considered as a differential diagnosis owing to its heterogeneous presentation [4]. However, for the reliable diagnosis of the disease, Endomyocardial biopsy and histopathological examination of myocardium is the absolute necessity [5]. We report a case of acute fulminant myocarditis diagnosed at autopsy, clinically diagnosed and treated as myocardial infarction.

Case History

A 55 years old male prisoner was brought to the casualty with sudden onset of chest pain and breathlessness at rest. On examination, there was tachycardia with a pulse rate of 104/min, pulse volume was low and weak and Jugular venous pulse was raised. Systolic blood pressure was low with 80 mm Hg. Heart sounds were muffled. ECG showed ST segment & T wave changes. CK-MB was moderately raised. A Clinical diagnosis of MI was made and treated for the same. Unfortunately patient expired the next day. A cause of death was reported as “Cardiogenic shock, secondary to inferior wall MI and complete heart block with ?Anterior wall MI”. As it was a custodial death, the body was subjected to the medico-legal autopsy.

Patient had been apparently healthy, except for flu like symptoms 5-6 days prior to admission for two days. He was a chronic smoker and non-alcoholic. There was no history of hypertension, diabetes and previous ischemic
heart disease. There was no family history of cardiac disorder.

Autopsy findings: On external examination, there was no significant finding apart from the post mortem changes corresponding to time of death. Internal examination revealed positive findings confined to the heart. Heart weighed 300gms and was cut open along the flow of blood. There was no evidence of pale or necrotic areas suggestive of MI. Left anterior, Left circumflex and Right Coronary arteries showed only yellowish atheromatous plaques without occlusion. On microscopy, the myocardium of the right ventricle, interventricular septum, left ventricular wall around the coronary sinus and A-V Node showed focal areas of myocarditis, with myocardial fibre disarray (Fig.1) and myocytolysis (Fig.2).

Fig-1: Microphotograph showing myocarditis with inflammatory infiltrate comprising of lymphocytes and plasma cells with myocardial fibre disarray (H&E X 200).

Fig-2: Microphotograph showing Myocytolysis and inflammatory infiltrate (H & E X 400).

Discussion

Myocarditis is defined as “a process characterised by an inflammatory infiltrate of the myocardium with necrosis and/or degeneration of the adjacent myocytes not typical of ischemic damage associated with coronary artery disease” [2]. The diagnosis is frequently made post-mortem, where no clinical evidence of myocardial failure has been present [4]. Data as to autopsy frequency of myocarditis are heterogenous. Passarino et al report frequencies ranging from 0.11-5.5% [5]. The causes of acute myocarditis can be Infectious & Non-Infectious. Although viral infection is the most common etiology, myocarditis is often idiopathic. The common viruses causing myocarditis include Coxsackie A and B, Influenza A and B, Cytomegalovirus, Varicella zoster virus, Epstein-Barr virus, Mumps, Respiratory syncytial virus and Hepatitis B. Recently, adenoviruses, various Herpes viruses, Parvovirus B19 and Human immunodeficiency viruses have been identified as potential cardiotropic agents [6]. Other infectious etiologies are bacterial, parasitic and fungal infections which are rare. Among the non-infectious causes are autoimmune.

These areas were extensively infiltrated by inflammatory cells, predominantly lymphocytes and few plasma cells. The left ventricle in most part revealed remarkable degree of hypertrophy. The tissue from the left and right atrial wall revealed normal myocardium. There was no evidence of myocardial infarction. The pericardium also showed infiltration with lymphocytes and plasma cells suggestive of pericarditis. The left anterior descending and right coronary artery showed Grade V atheromatous lesion, without significant narrowing of the lumen. Left circumflex artery revealed Grade III atheromatous lesion. The other organs sent for autopsy were liver, spleen, both kidneys, brain and both lungs, all of which were within normal limits, except lungs which showed features of Chronic venous congestion. The histological diagnosis of Acute Fulminant myocarditis was made. On perusal of clinical history, autopsy findings and histopathological examination, the cause of death was opined as “Complete heart block with heart failure secondary to acute fulminant myocarditis.”
disorders [7] and toxic agents like drugs [2]. However, in the present case, there was no history of exposure to non-infectious agents and infectious cause could not be definitely established. Since patient had flu like symptoms before the admission, viral etiology was considered.

The clinical presentations of myocarditis are variable, ranging from an asymptomatic to profound cardiogenic shock. Majority of the patients have no specific cardiovascular abnormalities, but may have ST segment and T wave abnormalities on ECG. Chest pain may occur in up to 35% of patients and usually reflects associated pericarditis [7]. Occasionally patients present with clinical syndrome identical to Acute MI. Left ventricular dysfunction may be present in less than 50% of patients. At autopsy, coronary arteries are usually patent [7]. Myocarditis mimicking myocardial ischemia or infarction is a rare but known phenomenon [8]. Angelini A et al reported that out of 12 cases clinically diagnosed as MI, 11 cases showed histological evidence of Myocarditis [4]. Patients may present with syncope with A-V block or ventricular arrhythmias. Sudden cardiac death can be initial presentation of myocarditis in some patients presumably, from complete heart block or ventricular tachycardia [7]. In the present case, patient presented with ischemic chest pain, and had ST segment abnormalities & complete A-V block, which clinically masqueraded as myocardial infarction. There is no single commonly accepted gold standard diagnostic test for Acute Myocarditis [2]. An increase in the CK-MB is seen approx in 10% of patients but newer Troponin assays are sensitive for myocardial injury. ECG shows ST-T wave abnormalities. However, similar findings can be seen in MI and are only suggestive but not diagnostic parameters [7]. Histopathological diagnosis is critical to confirm the diagnosis, either by endomyocardial biopsy or at autopsy [4]. ‘Dallas Criteria’ is used to establish histopathological diagnosis and classification of myocarditis [2]. These criteria separate biopsies into Active, Borderline or Non Myocarditis with routine haematoxylin and eosin staining. A lymphocytic infiltrate is characteristic of viral myocarditis [6]. According to clinical pathological classification, myocarditis is divided into 4 subgroups- 1. Fulminant myocarditis, 2. Subacute myocarditis 3. Chronic active myocarditis, 4. Chronic persistent myocarditis [7]. Patients with fulminant myocarditis have distinct onset of the condition, usually within days of the well defined viral illness. They present with severe Left Ventricular dysfunction and often cardiogenic shock. Histologically unequivocally positive with severe inflammatory infiltrate and myocyte necrosis [2]. Clinically, and histopathologically, the present case showed features of fulminant myocarditis.

In the present case, patient presented with clinical manifestations of Acute MI. Chest pain was due to pericarditis and the left ventricular failure could be attributed myocarditis. Patient had flu like illness prior to the development of the disease. Acute infections are associated with multiple host responses triggered by cytokines, which result in wasting of striated muscle, degradation of performance related metabolic enzymes and deterioration of central circulatory function, leading to cardiac failure. Histologically, in this case, A-V node was extensively infiltrated with lymphocytes, which can explain complete heart block on ECG. Since there was no history of drug intake or exposure to toxins and there was nothing suggestive of an autoimmune reaction, the non-infectious causes of myocarditis were ruled out. Considering the history, clinical presentation, gross and histopathological findings, the cause of death was concluded as acute fulminant myocarditis.

**Conclusion**

Myocarditis has highly variable clinical presentation and hence the diagnosis is often made at autopsy. Present case stresses the importance of considering the differential diagnosis of myocarditis while dealing with the case of chest pain. Endomyocardial biopsy is strongly recommended for the detection of myocarditis and its sensitivity may be further increased by immunohistochemical and molecular biological techniques.
References


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