NON ISCHEMIC CARDIOMYOPATHY – A CASE REPORT

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ABSTRACT
Cardiomyopathies are a group of diseases characterized by the structural and functional abnormalities of the heart muscles in the absence of other illnesses that might be responsible for the observed myocardial anomaly. Hypertrophic and dilated cardiomyopathies are the most prevalent types. Rare types include restrictive cardiomyopathy and restrictive cardiomyopathy with arrhythmia. Dilated cardiomyopathy has a documented prevalence of 36 cases per 100,000 population in Europe and North America, and its yearly incidence varies between 5 and 7.9 cases per 100,000 population. Non-ischemic dilated cardiomyopathy, which causes heart failure, dramatically increases the global burden of cardiovascular diseases. For both men and women, acute dilated cardiomyopathy is mainly brought on by myocarditis. This case was present in Aga Khan University Hospital; and was selected to be reported. The diagnosis in the case scenario is non-ischemic cardiomyopathy associated with heart failure and pulmonary oedema.

Key Words: non-ischemic cardiomyopathy, Dilated Cardiomyopathy

INTRODUCTION

Cardiomyopathies are a group of diseases characterized by the structural and functional abnormalities of the heart muscle in the absence of other illnesses causing the myocardial abnormality. Hypertrophic and dilated cardiomyopathies (DCM) are the most prevalent types. Rare kinds include restrictive, restrictive with arrhythmia. The pathogenesis of DCM includes ventricular enlargement and systolic dysfunction when neither severe coronary artery disease nor established aberrant loading conditions are present. It is regarded as one of the leading causes of heart failure globally. The DCM has a documented prevalence of 36 reported cases per 100,000 population in Europe and North America, and its yearly incidence varies between 5 and 7.9 cases per 100,000 population. The DCM appears to be more common in Africa and Latin America than in Eastern Asia, such as 14 reported cases per 100,000 in Japan, where it is more prevalent in comparison to Europe(1).

Previously reported studies have shown that non-ischemic cardiomyopathy is very common in young adults below the age of 50 years. Non-ischemic DCM, which causes heart failure, dramatically increases the global burden of cardiovascular diseases. For both men and women, acute DCM is mainly brought on by myocarditis. However, recent clinical and experimental data indicate that the pathophysiology and prognosis of DCM vary by gender (2). Here we present a relatively rare case of a young adult non-ischemic cardiomyopathy patient presenting with heart failure and pulmonary oedema.

CASE SCENARIO

A 29 years old male patient was presented in the cardiology clinic with complaints of shortness of breath (SOB) and generalized oedema. The patient had SOB with fatigue and a productive cough for the last two months. There were no known comorbidities; however, he had a positive family history of cardiac diseases. His father was diagnosed with Heart failure at the age of 20 years and died at 34 due to the complications. The patient's echocardiogram revealed a left ventricular ejection fraction of 10% with biventricular dysfunction. The patient was admitted electively for Left Heart catheterization, which later turned out to be normal. However, because of increased filling pressures and decreased cardiac output, an Intra-aortic balloon pump (IABP) was inserted

and inotropic support was initiated. The patient was diagnosed with Non-ischemic Cardiomyopathy and transferred to CCU. Gradually, the patient developed anuric renal failure, prompting an increase in inotropic support despite IABP. Since the patient was a candidate for Heart Transplant, his family was recommended to look for a donor. Meanwhile, an atrial septostomy was advised for Left Ventricular unloading, for which the patient was transferred to the United States of America on family preference.

Findings from Physical Examination and Clinical Presentation

On arrival, the patient's Glasgow Coma Scale was 15/15, oriented to time, place, and person. During physical assessment, the skin appeared cool and clammy. The cardiac evaluation revealed murmurs at the point of maximal impulse (PMI) consistent with an enlarged left ventricle. On respiration, bilateral equal air entry was accompanied by basal crept. Bilateral pleural effusion was evident by dull percussion at the bases. In the musculoskeletal System, tendon reflex was +2 with a range of motion limited at the right leg (due to IABP). Lymph nodes were non-palpable. The abdomen was soft and non-tender. No significant finding was noted in any other system. The patient was anuric during an assessment.

Laboratory and diagnostic test findings with rationale

There are multiple diagnostic measures to rule out cardiomyopathy or associated risk factors depending on the symptoms. Initial evaluation of non-ischemic cardiomyopathy typically includes blood tests, radiological assessments such as Magnetic Resonance Imaging or Computerized Tomography scan, echocardiogram, electrocardiogram (EKG), Holter monitoring, and Cardiac catheterization. A summary of the investigations is presented in Tables 1 and 2.

Table. 1. Summary of the cardiac of the patient

Diagnostic Measures	Results
Electrocardiogram	Heart rate > 90-110bpm Atrial Fibrillation Premature ventricular Contraction Q waves
Echocardiogram	Left Ventricle Ejection Fraction 10% Biventricular Dysfunction Left Ventricular Thrombus
Cardiac Magnetic Resonance (CMR)	Dilated left ventricle Trabeculation in stellate pattern Thin myocardium apical anterior wall.

Table 2. Summary of Hematological Investigations of the Patient

HB	**10.6**	**K**	***4.9**
HCT	***32.8**	**Na**	**138**
RBC	**6.52**	**HCO3**	**24.8**
WBC	**21.5**	**Cl**	**101**

Platelets	***140**	**Ca**	**9.1**
PT	**12.7**	**BUN**	***27**
APTT	**23.5**	**Cr**	***2.8**
INR	**1.0**	**Trop-I**	***204**

Differential diagnosis and current diagnosis with rationale

Cardiomyopathy is a broad term, and it leads to many cardiac complications. As it can either be ischemic or non-ischemic, sometimes it gets difficult to distinguish between both disorders. Furthermore, cardiomyopathy is divided into four types; dilated, hypertrophic, arrhythmogenic, and restrictive cardiomyopathy (3). The final diagnosis in the case scenario is non-ischemic cardiomyopathy associated with heart failure and pulmonary oedema. Non-ischemic dilated cardiomyopathy, which accounts for >50% of all heart transplant surgeries, is currently the leading cause of progressive heart failure(4).

Management of the Non-ischemic Cardiomyopathy

The causes of non-ischemic dilated cardiomyopathy determine the course of treatment. Treatment aims to lessen signs and symptoms, enhance blood circulation, and avoid additional damage to the heart. Apart from medical management, surgical implants are also used as part of a dilated cardiomyopathy treatment plan to improve cardiac contractility, ultimately increasing the cardiac output and minimizing the manifestations(5).

Pharmacological Therapy

A number of drugs are available, and a combination is preferably used to treat dilated cardiomyopathy and avoid any further complications. Table 3 shows the typical medications used in the treatment plan for non-ischemic cardiomyopathy (5).

Table 3. List of drugs available for treatment of cardiomyopathy

Drug Class	Mechanism of Action
Angiotensin-converting enzyme (ACE) inhibitors (lisinopril, enalapril, etc)	Reduces afterload through peripheral vasodilation.
Beta Blockers (atenolol, metoprolol, carvedilol, etc)	Reduces cardiac workload and oxygen demand that occurs as a response to sympathetic activation.
Angiotensin receptor blockers (ARBs) (losartan, candesartan, etc)	Inhibits Vasoconstriction and Myocardial fibrosis
Hydralazine/nitrate combination	Acts as a vasodilator
Diuretics	Eliminates excess fluid from the body (Volume overload)

Selective Antagonist Eplerenone	Attaches selectively to the mineralocorticoid receptor, limiting the binding of aldosterone and preventing sodium reabsorption as well as other harmful aldosterone related processes.
Ipratropium Bromide	Anticholinergic Bronchodilator

Non-Pharmacological Treatment

Non-pharmacologic treatments for non-ischemic cardiomyopathy include both device therapy and surgical intervention. Implants that are frequently utilized to treat non-ischemic cardiomyopathy involve the following options:

- *Implantable Cardioversion Defibrillator:* Used as a secondary prevention in patients with ventricular arrhythmias regardless of the involvement of coronary syndrome.
- *Cardiac Resynchronization Therapy:* Monitors cardiac activity such as mitral regurgitation, cardiac index, and Left Ventricular volume. It is recommended to improve quality of life.

Apart from Device therapy, some surgical procedures are performed to temporarily improve cardiac activity and reduce further complications, such as Mitral Valve Annuloplasty, Left Ventricular Assist Device, and Atrial Septoplasty. Since there are not enough studies to support the long-term benefits of these procedures, Cardiac transplantation is ultimately the ''gold standard'' for the treatment of end-stage cardiac failure(5).

DISCUSSION

After reviewing the case, genetic factors emerged as the root cause of non-ischemic cardiomyopathy for this patient. The pathogenesis of heart failure in DCM includes immune-mediated processes, peripartum cardiomyopathy (PPCM), endocrine and metabolic disorders, hereditary reasons, direct cardiac injury from viral or toxic agents, and genetic factors. A genetic factor can be found in 30–50% of cardiomyopathies cases and up to 40% of DCM patients. However, this fraction is likely overestimated, given the variability in disease penetrance and clinical presentation (1).

Given its higher spatial resolution, cardiac magnetic resonance (CMR) is the gold standard for determining ventricular ejection fraction and volume. CMR may accurately diagnose a number of diseases, including cardiomyopathies. In contrast to an echocardiogram, CMR has the potential to preclude other possible causes of the cardiomyopathies like amyloidosis, coronary artery diseases and iron overload. Reduced cardiac function, ventricular enlargement, and thinning of the myocardial wall can be easily seen on CMR. Due to its capability to enable the evaluation of numerous different tissue features in a single test, cardiac MRI is essential to evaluating patients with cardiomyopathy(6).

Critical points for reflection

After going through this patient's history, we selected this patient for our case study because we felt that non-ischemic cardiomyopathy in young adults is reported more frequently. After going through the patient history and literature, we found that genetics and heredity can be the leading cause of non-ischemic cardiomyopathy, and its early detection can increase the chances of survival and prevent complications. For this combination of situations there is limited literature available. Thus further studies to assess risk factors, early detection methods, and screening protocols needs to be studied.

Ethical Consideration: The case report was submitted with the permission of the patient

Conflict of Interest: There is no conflict of interest.

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