Sudden Death from Dilated Cardiomyopathy: Case Report

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ABSTRACT

Sudden unexpected death is commonly dealt by a forensic pathologists. sudden cardiac death being a major cause of death in Thai population. Even though dilated cardiomyopathy is the most common type of cardiomyopathy found, it rarely results in sudden death. The author described a case of Thai adult male who was found dead in his bed. The autopsy finding revealed the cause of death to be dilated cardiomyopathy (DCM).

Keywords: Sudden death, dilated cardiomyopathy, forensic autopsy

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INTRODUCTION

Sudden unexpected death is a category of death commonly dealt by forensic pathologists. The purpose of autopsy in these cases is to determine whether the cause of death is due to the criminal act or not. Sudden Cardiac Death (SCD) is known to be the major etiology of sudden unexpected death in a forensic setting.[1] SCD is usually defined as the unexpected natural death from a cardiac cause within a short time period, generally ≤ 1 hour from the onset of symptoms, in a person without any prior condition that would appear fatal.[2]

Cardiomyopathy is found to be the second leading cause of SCD with coronary artery disease being the first.[2] However, the prevalence of cardiomyopathy in forensic cases is relatively sparse.[3] This may be due to the fact that post-mortem examination is requested when sudden unexpected death occurs, while the clinical manifestation of dilated cardiomyopathy is often congestive heart failure. Thus, the patients usually seek medical care and receive diagnosis early before death occurs.

The European society of cardiology defines cardiomyopathy as a myocardial disorder in which the heart muscle is structurally and functionally abnormal, in the absence of coronary artery disease, hypertension, valvular disease and congenital heart disease sufficient to cause the observed myocardial abnormality.[4] Cardiomyopathies can be categorized by their abnormal morphologies and functions into hypertrophic cardiomyopathy (HCM), dilated cardiomyopathy (DCM), restrictive cardiomyopathy (RCM), arrhythmogenic right ventricular cardiomyopathy (ARVC) and

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unclassified cardiomyopathy. Each type can be further classified into familial/genetic form and non-familial/non-genetic form. However, the American Heart Association (AHA) suggests a different classification of cardiomyopathy. The classification scheme stated by AHA is based on the pathogenesis of disease or predominant organ involvement. If the disease is confined only to the heart muscle, it is defined as "Primary cardiomyopathy" which can be further divided into genetic causes such as HCM, acquired causes such as peripartum cardiomyopathy and mixed etiologies such as DCM. Whereas, "Secondary cardiomyopathy" is the pathological myocardial involvement as a part of generalized systemic disorders such as amyloidosis, sarcoidosis, hemochromatosis etc.[5]

The author reports a case of sudden death in a Thai male during sleep which later turned out to be dilated cardiomyopathy.

History

A body of a 36-year-old man was sent to the Department of Forensic Medicine Siriraj Hospital for post-mortem examination. He was an electrician living alone in his apartment. He was found dead in his bedroom one morning by his co-worker. Police investigation revealed that the surroundings were clear of trespass or sign of violence. His brother presented at the morgue next day and stated that the deceased was previously healthy. There was no history of medical condition or any hereditary disease in the family.

Autopsy finding

Autopsy examination was performed one day after the body had arrived. The body was that of a sthenic build male in adult age. The height was 171 cm and the weight was 70 kg. The external examination showed no sign of trauma.

The heart showed cardiomegaly and weighed 560 g as shown in Fig 1. Left atrium and ventricle showed chamber dilation. Scattered foci of myocardial fibrosis were found in papillary muscles of his left ventricular free wall and interventricular septum. No atherosclerotic plaque was found in coronary arteries. Cardiac valves were in normal position and appearance. The left and right lungs weighed 470 g and 630 g, respectively. Other organ systems were unremarkable.

Microscopic examination of the heart showed hypertrophy of cardiac myocyte and enlarged box-shaped nuclei. Perivascular fibrosis and multiple foci of fibrosis were found throughout the myocardium as in Fig 2-3. Lymphocyte infiltration can only be seen in some areas of fibrotic tissue as in Fig 4. There was no myocytolysis or myocardial necrosis found. The microscopic examination of other internal organs was unremarkable. Toxicology screening was negative. The cause of death was given as dilated cardiomyopathy.

DISCUSSION

Dilated cardiomyopathy (DCM) is the most common form of cardiomyopathy. It is characterized by left ventricular chamber enlargement and dilation, which is a systolic dysfunction in the absence of abnormal loading condition such as hypertension or valvular heart disease[6]. Microscopic finding often reveals

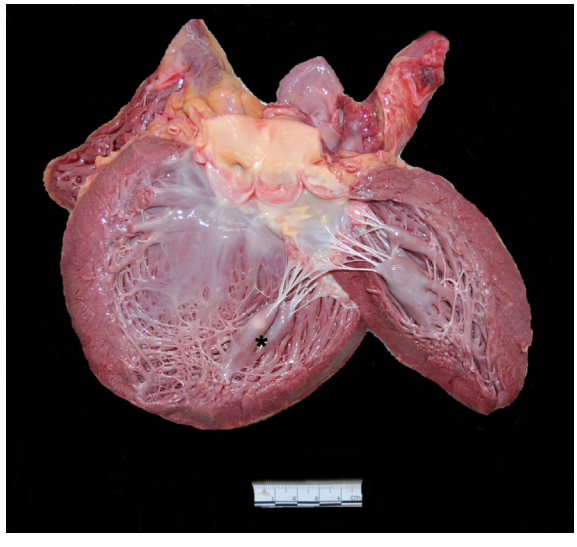

Fig 1. The heart showed cardiomegaly and chambers dilatation. Myocardial fibrosis was noted in papillary muscle (*).

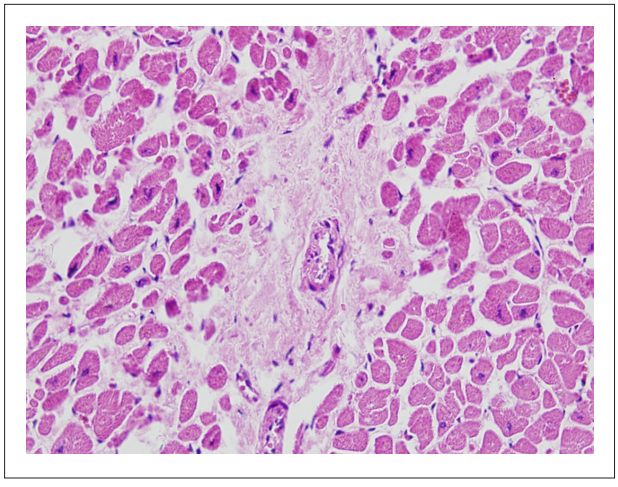

Fig 2. (H&E, x200) Perivascular fibrosis.

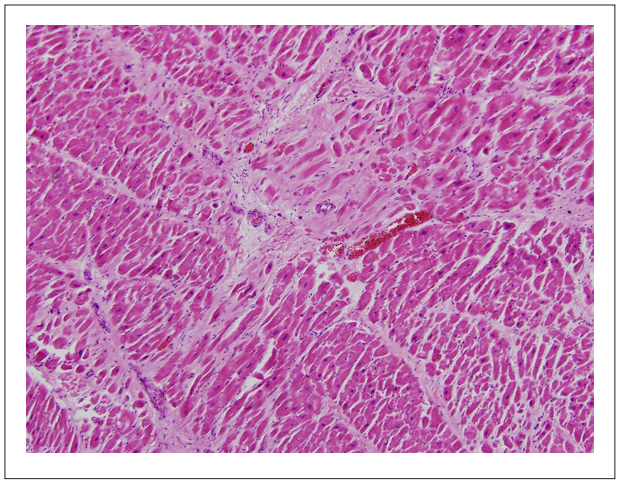

Fig 3. (H&E, x100) Diffuse interstitial fibrosis throughout myocardium.

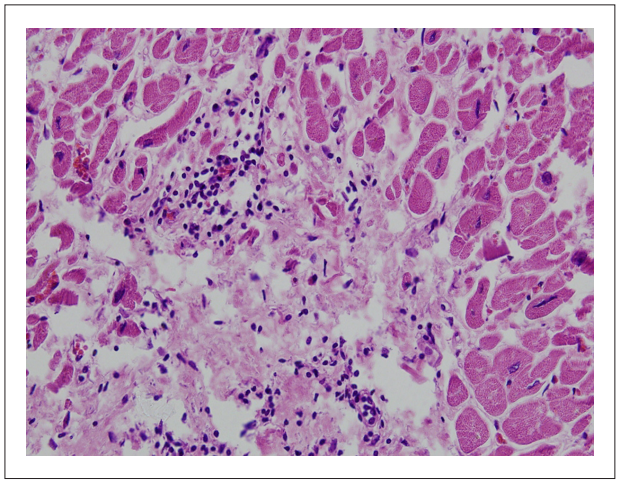

Fig 4. (H&E, x400) Lymphocyte infiltration.

areas of interstitial and perivascular fibrosis, and sometimes small areas of necrosis and cellular infiltrate of mainly lymphocytes. Features of congestive heart failure such as colliquative myocytolysis and endocardial thickening can also be found.[7]

TABLE 1. Acquired causes of DCM and systemic diseases those can cause DCM[5,6]

Post inflammatory
Viral myocarditis
Autoimmune myocarditis
Chagas disease *(Trypanosoma Cruzi infection)*
Toxin-related causes
Alcoholic cardiomyopathy
Anthracyclines
Metals *(cobalt, lead, mercury, and arsenic)*
Peripartum cardiomyopathy
Nutritional deficiencies
Beriberi (thiamine)
Muscular dystrophy
Duchenne/Becker muscular dystrophies
Iron storage disorders
Primary hemochromatosis
Chronic hemolytic anemia treated by transfusion

The cause of dilated cardiomyopathy is very heterogeneous, with genetic (familial) form in 20-35% of cases.[8,9] The mode of inheritance is mainly autosomal dominant and less frequent autosomal recessive and X-linked recessive. The most common gene involved in familial DCM is LMNA, encoding the intermediate filament protein lamins A and C[10]. Some genetic forms of dilated cardiomyopathy are inherited with other phenotypes such as muscular dystrophy, cardiac conduction defect, and sensorineural hearing loss.[9,11]

The acquired causes of DCM and those systemic diseases which can affect the heart, resulting in DCM, are shown in Table 1. Due to non-specific pathology, the etiologies of DCM cannot be differentiated by morphology of the heart alone, except for the DCM phenotype caused by intracellular storage disorders which have distinctive gross and microscopic features such as hemochromatosis.

The family history of the deceased in this case was negative for heart disease or sudden death among relatives. Also, lymphocyte infiltration in the heart of this patient was sug-

gestive of inflammatory DCM. The proposed mechanism of progression from myocarditis to DCM was pathologic remodeling which resulted in excessive tissue fibrosis.[12]

The cause of death in DCM patients is mostly congestive heart failure, sudden death rarely occurs.[13] When sudden death in DCM does occur, it is usually attributed to ventricular tachyarrhythmia,[14] and less than 20% of the patients have bradyarrhythmia. The risk of sudden cardiac death in a patient with DCM has been found to increase with decline in systolic function.[15]

CONCLUSION

The author has described a case of a Thai male from Northern Thailand who suffered from sudden cardiac death during sleep. The cause of death was diagnosed as DCM. The definition, pathological finding and the pathophysiology of genetic and acquired forms of DCM have also been described.

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