Sudden death of two infants with dilated cardiomyopathy: A case report

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Abstract: Dilated cardiomyopathy (DCM) occasionally contributes to sudden death, and in rare cases, undiagnosed DCM may be encountered during forensic autopsy. It is difficult for forensic experts to confirm the cause of DCM because only limited examinations can be performed during forensic autopsy. In this report, we presented two cases of sudden infant death due to DCM transitioned from lymphocytic myocarditis. In case 1, we observed biventricular dilation, moderate diffuse lymphocytic infiltration, irregular arrangement of myocardial fibers, and fibrosis. In case 2, we observed biventricular dilation, mild lymphocytic infiltration, and mild diffuse fibrosis. In these two cases, the patient was diagnosed with DCM that had transitioned from myocarditis only during autopsy but had not been diagnosed earlier owing to the absence of abnormalities during medical examination. Both forensic experts and clinical doctors should consider the possibility of inapparent cardiomyopathy in subjects who have a clinical course similar to the abovementioned cases.

Key Words: dilated cardiomyopathy, lymphocytic myocarditis, forensic autopsy, infant, sudden death.

INTRODUCTION

Dilated cardiomyopathy (DCM) usually causes congestive heart failure, blackout due to arrhythmia, and generalized embolization [1]. DCM occasionally progresses to an advanced stage without apparent symptoms and causes sudden death. Undiagnosed DCM is rarely encountered in the forensic field, regardless of its contribution to death. Except for genetic disorders, DCM is the terminal phase of myocardial damage caused by various factors such as infections, toxicity, and abnormal metabolism; acute viral myocarditis is reported to be the most common cause of DCM via an immunological response [1]. However, it is very difficult to confirm the cause of DCM because forensic autopsy involves several limitations with regards to examination processes; for example, the cardiac output cannot be measured because the heart has stopped functioning at the time of autopsy. Additionally, although the time for progression of DCM varies from several months to several years, patients with DCM, especially children or young adolescents, tend to have a poor prognosis.

This report presents two cases of sudden infant death, in which the infants were diagnosed with DCM for the first time during autopsy.

CASE REPORT

Case History

Case 1
A 10-month-old female child had not shown any abnormality from the time of birth to a regular health check-up performed at 6 months of age. She showed reluctance in drinking milk and began crying incessantly on the evening before the day of her death. On the day of death, she had been sleeping on a cushion in the living room; after a while, her mother noticed that she was not breathing and requested for emergency medical services. She was brought to the hospital but died despite undergoing resuscitation. A medicolegal autopsy was performed 18 hours after the patient’s death.

Case 2
A 3-month-old male child, who was born in the 34th gestational week and had a low birth weight of 1980...
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...showed no particular abnormality after birth as well as during the 3-month health check-up. Because he had been crying since the morning and refused to drink milk, his parents took him to the hospital. He was hospitalized owing to facial pallor, cold limbs, dehydration, and an oxygen saturation rate of 91%. Eventually, his respiration rate increased but his vital status suddenly deteriorated. Subsequently, he died despite undergoing resuscitation. A medicolegal autopsy was performed 33 hours after the patient’s death.

**Autopsy Findings**

During autopsy, all internal organs were macroscopically examined, and tissue samples were collected. The samples were fixed with 10% formalin solution and cut into 3-μm-thick sections after being dehydrated and embedded in paraffin; subsequently, the tissue sections were subjected to hematoxylin–eosin and Elastica-Masson-Goldner staining.

**Case 1**

The body weight and height of this infant were 69 cm and 6.4 kg, respectively, and her nutritional status was considerably poor. Examination revealed a moderate number of clots in cardiac blood; we collected and pooled 12 mL of left pleural fluid, 45 mL of right pleural fluid, and 40 mL of peritoneal fluid. Her heart weighed 270 g (Fig. 1), (the normal heart weight for a 10-month-old female infant is approximately 36 g [2]). The thickness of her right ventricular wall was 0.2 cm and of her left ventricular wall was 0.7 cm. Her heart showed severe biventricular dilation and left ventricular hypertrophy (Fig. 2). She had spongy left ventricular papillary muscles. No other diseases, malformations, or considerable damage were noted in her other organs. The pathological findings were as follows: (1) both ventricular walls had...
moderate diffuse interstitial lymphocytic infiltration, partially involving nodular collection of lymphocytes (Figs 3, 4); (2) irregular arrangement of myocardial fibers, fibrosis, and collapse of myocardial fibers were relatively frequent (Fig. 3); and (3) the endocardium was moderately hypertrophic (Fig. 5). We diagnosed the patient with DCM due to lymphocytic myocarditis based on these macro- and microscopic findings.

Case 2

The body weight of this infant was 5.8 kg and his height was 59 cm; his nutritional status was also considerably poor. Examination revealed a moderate number of clots in cardiac blood; we collected and pooled 2 mL of left pleural fluid, 6 mL of right pleural fluid, and 90 mL of peritoneal fluid. His heart weighed 100 g (Fig. 6) (the normal heart weight of a 3-month-old male infant is approximately 32 g [2]). The thickness of his right ventricular wall was 0.3 cm and of his left ventricular wall was 0.7 cm. The heart showed biventricular dilation and hypertrophy (Fig. 7). He had spongy left ventricular papillary muscles and patent foramen ovale. There were no signs of other diseases, malformations, or considerable damage in other organs. The pathological findings were as follows: (1) the left ventricular walls had mild lymphocytic infiltration; (2) mild diffuse fibrosis of myocardial fibers and intramuscular interstitium and irregular arrangement of myocardial fibers (Fig. 8); (3) there was noticeable anisokaryosis (Fig. 9); and (4) the endocardium was moderately hypertrophic (Fig. 10). We diagnosed him with DCM based on these macro- and microscopic findings.

DISCUSSION

DCM is characterized by an increase in heart weight and dilation of the endocardial cavity, and fibrosis
and fat infiltration in the myocardial wall has been observed in many patients. Because the morphological position of the papillary muscle to which the chordae tendineae of the mitral valve adheres changes owing to dilation of the endocardial cavity, mitral valve regurgitation is frequently observed. From a microscopic point of view, hypertrophy of cardiomyocytes and associated vacuolar degeneration coexist with fibrotic lesions (usually less than 20% of the cross-sectional area). On rare occasions, extensive substitutional fibrosis may be observed [3, 4]. However, many nonspecific findings could indicate DCM and at present, exclusion diagnosis for validating DCM has not been established [5-9]; hence, it is often difficult for a forensic medical physician to diagnose DCM.

In case 1, moderate lymphocytic myocarditis of relatively acute phase occurred, and interstitial microfibrillar collapse in the myocardium occurred as a whole. It was speculated that lymphocytic myocarditis had transitioned to DCM owing to the dilation of both ventricles.

In case 2, fibrosis was mild but diffuse between myocardial fibers, suggesting that the myocardial fibers had been injured previously. Furthermore, mild lymphocytic infiltration was observed. We suspected a history of myocarditis due to viral infection, and this condition then developed into DCM.

The recently reported annual incidence of DCM occurring during childhood was 0.65% (2.4 per 100,000 children), and DCM in children results in poor prognosis (1-year survival rate, 75%; 5-year survival rate, 60%) [10]. The mean age of 63 children with idiopathic DCM at the time of diagnosis was 4.96 ± 5.3 years [11]. In children with DCM, arrhythmia occurs frequently, although it is believed that arrhythmia does not affect prognosis [12-13]; however, mortality is infrequent in children with restrictive cardiomyopathy [14].

CONCLUSIONS

The two cases we encountered could be diagnosed during autopsy as DCM transitioned from myocarditis, and the ages of onset of these patients were lower than the mean age of DCM onset; moreover, no abnormalities, such as arrhythmia, could be detected during regular medical examinations. In the field of forensic medicine, cases involving sudden deaths of infants often become autopsy subjects. Although abnormalities could not be revealed during the regular medical examinations in these cases and the stage at which mortality can be prevented remains unclear, there is a possibility that mortality can be prevented if diagnosis and treatment are performed at a very early stage, immediately after the appearance of symptoms. Both forensic and clinical doctors should always consider the possibility of cardiomyopathy when cases follow a clinical course similar to those of the two cases described here, although such cases occur rarely.

Conflict of interest. The authors declare that there is no conflict of interest.

References


