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CASE REPORT

Sudden cardiac death with triple pathologies: A case report

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Abstract Sudden cardiac death in young adults may be associated with rare cardiomyopathies such as left ventricular noncompaction (LVNC) and arrhythmogenic right ventricular (ARVC) cardiomyopathies. LVNC is characterised by hypertrabeculations and deep recesses of the left ventricle. ARVC presents with thin myocardium as a result of extensive fibro-fatty infiltrations. In both conditions, death may be due to arrhythmia, thromboembolic events or heart failure. We report a case of a 21-year old athletic young man who collapsed at the futsal court right after the game. He was resuscitated but expired at the hospital after a brief admission. A week earlier, he had a similar episode of syncope and revived through cardio-pulmonary resuscitation at the site. Post mortem examination showed extensive acute myocardial infarction (AMI) involving the papillary muscles and the left ventricular wall. Features of LVNC were also observed. On top of that, the right ventricle showed patchy thin myocardium as the wall was largely comprised of fat. Histology examination confirmed the presence of AMI and massive fibro-fatty infiltrations of the right ventricle. This unfortunate young man had co-existing cardiomyopathies which is rare indeed. As he succumbed to AMI, this mechanism of death is also uncommonly associated with neither LVNC nor ARVC. In conclusion, young and physically active individuals may not be spared of sudden cardiac death. Mild and non-specific symptoms should not be taken lightly as it may be the subtle signs of cardiomyopathies.

1. Introduction

Sudden cardiac death (SCD) is defined as an unexpected death resulting from a cardiovascular cause in an individual with or without pre-existing heart disease.[1] The causes of SCD in the group of young adults of less than 35 years old include myocarditis, valvular heart disease, congenital coronary artery anomaly and cardiomyopathies such as left ventricular non-

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compaction cardiomyopathy (LVNC) and arrhythmogenic right ventricular cardiomyopathy (ARVC).[2,3]

LVNC or 'spongiform cardiomyopathy' is a rare congenital cardiomyopathy which arises from arrested endomyocardial compaction during fifth to eighth week of embryogenesis.[4–6] It is characterised by prominent left ventricular trabeculations of the endocardial layer, deep inter-trabecular recesses and thin compacted layer.[6] The morphological defects may cause clinical manifestations such as syncope, thrombo-embolic events and death may result from heart failure or arrhythmias.[7,8]

Another important type of cardiomyopathy which is associated with SCD in young athletic adult is ARVC. The prevalence of ARVC is reported to be 1/1000 in general population.[3] It is a heritable myocardial disease as a result of mutations in genes encoding cardiac desmosomal proteins.[3,9] At autopsy, ARVC is characterised by markedly thin wall of the right ventricle as it is largely replaced by fibro-fatty tissue.[3] Microscopically, the fibro-fatty infiltrations with chronic inflammatory cells may be seen affecting both the right and left ventricles.[3,10] Common presentations may include palpitations, ventricular tachycardia (VT) and syncope, but many patients are initially asymptomatic.[10]

We present a case of a 21-year-old gentleman who suddenly collapsed at the futsal court right after the game. He had no known medical illness and led an active lifestyle. Autopsy examination showed an extensive acute myocardial infarction (AMI) of the left ventricle. Apart from that, the right and left ventricles revealed interesting pathologies of co-existing LVNC and ARVC.

2. Case report

A 21-year-old Malay gentleman was noted to be having shortness of breath and jerky movements right after playing futsal with his group of friends. A similar episode took place a week ago following the same event; however, he regained consciousness after a cardiopulmonary resuscitation (CPR) procedure performed by his friend. He did not proceed to seek any medical treatment as he believed he was healthy and physically active. During this episode, he became unconscious, unresponsive and immediately brought to the nearest hospital. Spontaneous circulation has returned upon successful resuscitation. He was subsequently referred to our centre for further management. At the Emergency Department, he was haemodynamically unstable despite being on two inotropic agents. He had another episode of cardiac arrest and revived after 12 min of CPR. Bedside echocardiogram that was performed revealed thickened left ventricular wall with increased trabeculations and poor cardiac contractility (Fig. 1). Arterial blood gases showed pH of 6.7 and other parameters were in keeping with severe metabolic and respiratory acidosis. In spite of the maximum medical supports given he eventually succumbed after nearly three hours of admission. A police report was lodged and an order was issued for medico-legal autopsy examination to determine the cause of death.

2.1. Autopsy findings

Autopsy examination showed a muscular, medium-built young adult male measuring 166 cm in height and 76 kg in weight.

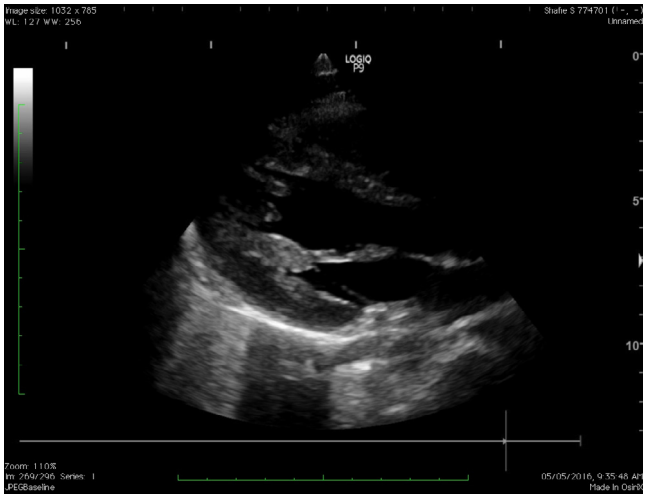

Fig. 1 Bedside echocardiogram showing thickened left ventricular wall and increased trabeculations.

Bluish discolouration was noted on the nail beds, indicating cyanosis. There was no significant injury noted on the body. Internal examination revealed a mild cardiomegaly as the heart weighed 350 gm. The right and left ventricular wall thicknesses measured 2 mm and 15 mm respectively. The left ventricle showed evidence of acute myocardial infarction (AMI) as demonstrated by the haemorrhagic myocardium vividly seen on the papillary muscles (Fig. 2a). The cut surfaces also showed areas of whitish discolouration, hyperaemia and soft parenchyma, consistent with AMI. On top of the ischaemic changes, the left ventricular wall also revealed excessive trabeculations with deep recesses (Fig. 2b). Coarse trabeculae resembling multiple papillary muscles were also observed. These features were consistent with LVNC. On the other hand, examination of the right ventricle also showed a remarkable finding. The free wall of the ventricle was thinner than normal and fatty tissues could be seen displacing the normal myocardium (Fig. 3a). These features were consistent with ARVC.

Other internal organs appeared grossly normal. Histopathology examination of the heart shows large areas of fibrosis, haemorrhage and neutrophilic infiltrations in the

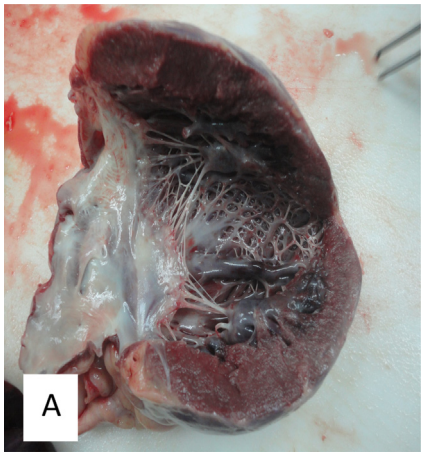

Fig. 2a Haemorrhagic papillary muscles.

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