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Sudden Death due to Dilated Cardiomyopathy and Cardiac Cirrhosis: An Autopsy Diagnosis

Braja Kishore Dash*, Jyotish Chandra Choudhury*, Pallavi Bhuyan, Manoj Kumar Jena***, Sitaram Mahapatro****, Tushar Bhutada*******

Abstract

Amongst unnatural deaths, the sudden death is the most common challenges faced by the forensic experts and pathologists in their day to day practice. It becomes a tough assignment to uncover instances of cardiac diseases that cause sudden natural death in the absence of coronary artery disease. A 62 years old male was declared as brought dead to the hospital within an hour of sudden onset uneasiness and chest discomfort. An autopsy was performed as per the guidelines laid down by AECP (Association for European Cardiovascular Pathology -2007) and findings were found to be consistent with Dilated Cardiomyopathy with Cardiac Cirrhosis. We emphasize on adoption of uniform methods of investigation in cases of sudden cardiac deaths for improvements in standards of practice and valid comparisons between different regions and communities.

Keywords: Sudden Death; Dilated Cardiomyopathy; Cardiac Cirrhosis.

Introduction

Sudden deaths create a lot of unrest in the society that along with the law implementing bodies looks forward to forensic experts and the histopathologists for thorough investigation and scientific unfolding of the facts. The World Health Organization (WHO) defines sudden death as a death occurring within 24 hours of the onset of symptoms, when the death is neither instantaneous nor due to sudden cardiac death or sudden infant death syndrome (ICD-10 code R96.1) [1]. Sudden Cardiac Death (SCD) is defined as 'Natural death due to cardiac causes, heralded by abrupt loss of consciousness within one hour of the onset of acute symptoms; pre existing heart disease may have been known to be present, but the time and mode of death are unexpected' [2]. Non-ischemic etiology of sudden cardiac death is less often reported as an autopsy diagnosis.

**Authors Affiliation: ***Associate Professor, ***Professor and Head *****MBBS, Department of F M T., **Assistant Professor ****Professor and Head, Department of Pathology, SCB Medical College and Hospital, Cuttack, Odisha, India.Pin- 753 007.

Reprints Requests: Tushar Bhutada, Department of FMT, SCB Medical College, Cuttack, Odisha, India. PIN- 753 007.

E-mail: doctortushar@gmail.com

Case Report

We present a case report of a 62 yr old male living alone as a tenant complained of sudden onset uneasiness and chest discomfort was brought by his landlord to the hospital where he was declared as being brought dead. Since the person was living alone, it was difficult on the part of the police to provide adequate information. Therefore an autopsy examination was looked upon to establish (1) whether death was due to poisoning with the possibility of toxic and illicit drug abuse (2) whether the death was attributable to cardiac disease or to other natural causes (3) the nature of the cardiac disease, and whether the precipitating factor was arrhythmic or mechanical (4) whether the cardiac condition causing SD warrants screening and counselling of the next of kin.

Autopsy Findings

External examination revealed a thin built person with swelling of both the limbs with a distended abdomen. Keeping in mind the possibility of an infectious disease, full Universal Work Precaution was reconfirmed. Body weight and length was 48 Kg and 1.58 meters respectively. There was the absence of any external injury with no evidence of intravenous access, intubation, ECG pads, Defibrillator burns,

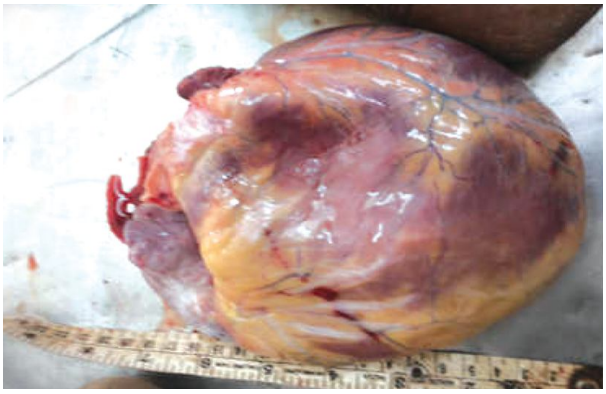

Fig. 1: Enlarged Heart

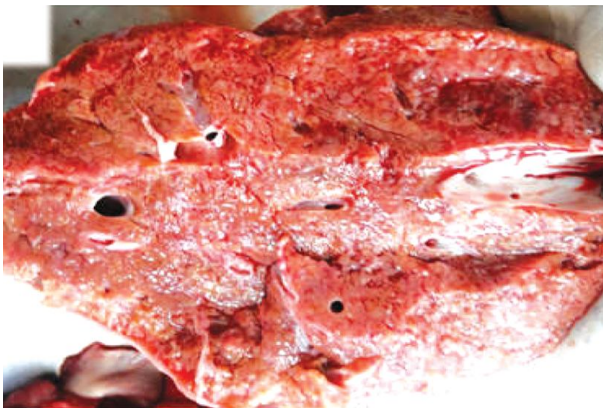

Fig. 2: Gross image of cut liver surface showing nodular subdivision of liver parenchyma.

drain sites or implantation of pacemaker device.

The dissection of the cranial cavity followed by that of the neck did not reveal any significant finding. The wall of the thoracic cavity was found normal; the mediastinum was found enlarged and about 250 ml of clear straw coloured free fluid in the right side pleural cavity. The pericardium was devoid of any adhesions, effusion, exudates and hemopericardium. Heart appeared globular and grossly enlarged (Figure 1), the transverse distance as measured from the obtuse to the acute margin along the posterior atrioventricular Sulcus was 11 cm. The longitudinal size by measuring the distance between the crux cordis and the apex on the posterior surface was found to be 13 cm. The great arteries revealed normal anatomy and were transected 3cm above the aortic and pulmonary valves. The pulmonary veins were checked and transected. Superior venacava was normal and was transected preserving the S.A node. The inferior venacava and the right atrium were dilated and devoid of any thrombi. The left atrium was normal. Pockets on the left and the right atrial endocardium with their openings directed to the valve suggested functional insufficiency of the respective valves. The leaflets of respective AV valves along with chordae tendineae were found intact, thickened, calcified and shrunken. The aorta, pulmonary artery and their respective valves were inspected from above and no significant finding could be noted. The coronary Ostia were found normal after examination in size, shape, position, number and potency. The size and the course of major epicardial arteries were found normal with right dominance. They were also found patent when multiple transverse cut sections were given at 3mm interval along the course and there was no evidence of any significant calcification, intraluminal stenting or bypass grafting. A complete transverse cut (short axis) at the mid ventricular level followed by parallel slices of ventricles at 1 cm interval towards the apex revealed dilatation of the RV and LV cavity, the mid cavity free wall thickness of the left side 1.1 cm, right side 0.4 cm, interventricular septum 0.9 cm excluding the trabeculae and the left ventricular cavity at the level of the papillary muscles, in a transverse cut was 4.5 cm. There was absent of any old scar or ischaemic patch in the walls or papillary muscles. The basal half of the heart was dissected along the flow of blood and the atrial and ventricular septum was found intact. The weight of the heart after emptying the blood was 0.74 kg. Ventricular sides of AV valves were inspected and the thickening of the leaflets and shortening of the chordate tendineae was appreciated more so on the right side. The heart was then preserved in 10% formalin for histopathology examination. Both the lungs were intact and congested. The abdominal cavity contained about 500 ml of clear straw coloured fluid. Mesentery contained a minimal amount of fat. Examination of the hollow viscera did not reveal contact to any toxic or irritant substance Liver was found shrunken weighing 1.45 kg, and cut section revealed large confluent central zones with a granular surface because of the presence of connective tissue (Figure 2). Spleen was found moderately enlarged. Kidneys, supra renal glands, abdominal aorta and bladder were found normal.

Toxicological Analysis

Blood from femoral vein (10 ml), blood from the heart (30 ml), routine viscera, urine (30 ml) and vitreous humor was preserved as per standard practice and was sent for toxicological analysis at the State Forensic Science Laboratory. Search for common poisons was found to be negative.

Biochemical Analysis

Biochemical examination of the pleural and ascitic fluid suggested each to be a transudate.

Histopathology Examination

Histopathology examination showed bridging fibrous septa between terminal hepatic veins with intact portal tracts within the centre of the parenchymal islands. (Figure 3)

A Masson's Trichrome strain showed the blue collagen fibres. (Figure 4)

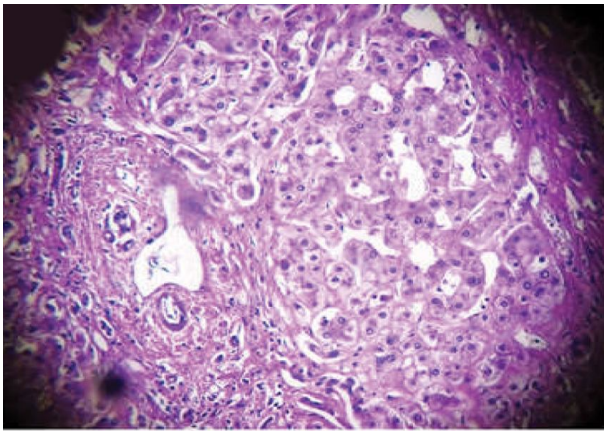

Fig. 3: Photomicrograph showing bridging fibrous septa between terminal hepatic veins, with intact portal tracts within the centre of parenchymal islands. [HE X 400]

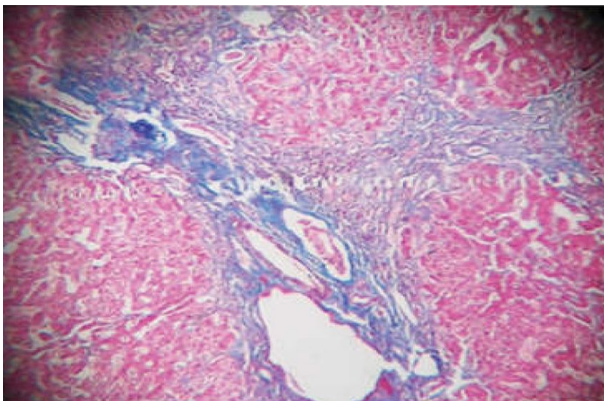

Fig. 4: Fibrosis with intact portal tract in the centre. [Masson's Trichrome X 400]

Discussion

Idiopathic dilated cardiomyopathy (DCM) is a chronic heart muscle disease characterized by left ventricular dilatation and impairment of systolic function. Epidemiologic studies provide an incidence of approximately 20 / 100000 / year and a prevalence of 38 / 100000 [2]. Up to 40% of cases may be familial; inheritance patterns are predominantly autosomal dominant, but X-linked families are reported (2–5%) [2]. DCM constitutes 25.0% of all cases of congestive cardiac failure [3]. The associated right sided failure causes the transmission of right atrial pressure to the liver, where at a cellular level, venous congestion impedes efficient drainage of sinusoidal blood flow into terminal hepatic venules. Sinusoidal stasis results in accumulation of deoxygenated blood, parenchymal atrophy, necrosis, collagen deposition and ultimately hepatic fibrosis [4]. Cardiac cirrhosis (CC) are characterized by venocentric cirrhosis also called reversed-nodularity cirrhosis [5]. This pattern is caused by dominant hepatic vein outflow obstruction with relatively intact portal veins that serve as outflow tracts. In this situation hepatic vein to vein fibrosis results in nodules composed of portal tracts in the centre and veins within the peripheral fibrous septa. Comparative sex data for CC are unavailable because of obvious reasons, however the available data suggest that its prevalence increases with age and it is more common in men [4].

Herman and Blumgart [6] in a series of 2000 consecutive autopsies established a causal significance of chronic passive congestion in the production of hepatic fibrosis by emphasizing the increasing incidence and severity of fibrosis with increasing duration of congestive cardiac failure. They also added that the predominant type of liver fibrosis in relation to cardiac decompensation was central fibrosis. Identical sort of fibrosis were observed in our case. Alvarez and Mukherjee [7] explain that a spectrum of hepatic derangements is possible in setting of right heart failure and the primary pathophysiology being either passive congestion from increased filling pressure or low cardiac output and consequence of impaired perfusion and microscopic, the hallmark features of hepatic venous hypertension are prominence of central veins, central vein haemorrhage, and sinusoidal engorgement. Instead, the ischaemic liver injury is characterized by the centrilobular necrosis of zone 3 hepatocytes [7]. Findings in our case show a preponderance of fibrosis consistent with that of the two studies mentioned above. Apart from cardiomyopathy, another cause of cardiac cirrhosis is constrictive pericarditis, severe pulmonary arterial hypertension (PAH), mitral stenosis, tricuspid regurgitation, cor pulmonale, and as a postoperative consequence of the Fontan procedure for pulmonary atresia, hypoplastic left heart syndrome and tricuspid atresia [6, 8]. Pillarisetti J, et al [9] also reported a rare instance of cardiac cirrhosis as a complication of primum atrial septal defect. Ortiz-Olvera NX et al [10] in a series of 1,176 autopsies demonstrated that liver cirrhosis were per se associated with a high frequency of cardiac abnormalities, regardless of cirrhosis aetiology and the most frequent alterations were cardiomegaly and left ventricular hypertrophy but they excluded CC from the study. However, in our case we have sufficient evidence to believe that hepatic fibrosis was

a consequence of cardiac decompensation. Sakashita et al [11] reported an instance of cardiac cirrhosis developing in a transplanted liver as a sequel of Amyloid cardiomyopathy. The microscopic architecture of reversed lobulation and prominent centrilobular hemorrhagic necrosis in their case was analogous to findings in our case.

SCD accounts for at least 30% of all deaths in DCM and may occur in patients with advanced as well as mild disease and the cause of death may be due to malignant ventricular arrhythmia, bradyarrhythmias, pulmonary or systemic embolization, or electro-mechanical dissociation [2]. However, malignant ventricular arrhythmia remains the commonest single cause of SCD in DCM [2]. Our autopsy did not reveal presence of any embolization, hence, we designate arrhythmia and / or electromechanical dissociation as possible causes of sudden onset circulatory failure triggering death in present case.

Conclusion

DCM with Cardiac cirrhosis rarely reported following a medicolegal autopsy case and could very easily go unnoticed in the absence of stringent protocol and high level of suspicion. We emphasize on adoption of uniform methods of investigation as laid down by AECP [12] (Association for European Cardiovascular pathology) while performing autopsies in cases of sudden cardiac death for improvements in standards of practice and valid comparisons between different regions and communities.

Conflict of Interest

None declared.

Source of Funding

This research received no grant from any funding agency in the public, commercial and not-for-profit sectors.

Research Ethics

The study satisfies legal and ethical criteria.

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