

**Case Report**

Massive Cardiac Lipomatosis of the Right Heart as Cause of Sudden Cardiac Death

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Abstract

Cardiac lipomatosis is a rare and generally benign condition characterized by adipose infiltration of the heart muscle. The aetiology is unknown, and few cases are described in the literature. It is usually asymptomatic, but in some cases can cause fatal arrhythmias. We report on an autopsy case of an elderly patient with massive cardiac lipomatosis of the right heart, which causes dysfunction of the atrioventricular node and sudden cardiac death.

Keywords: Massive Cardiac Lipomatosis; Atrioventricular Node Dysfunction; Right Heart; Sudden Cardiac Death

Introduction

Cardiac lipomatosis is a degenerative process of unknown aetiology in which fatty tissue accumulates within the myocardium and replace it, mostly in the right ventricle. It is a rare condition, therefore there are not precise epidemiological data on its prevalence, however, it is believed to affect mainly females and elderly individuals and it is commonly associated with obesity. The condition is generally considered as benign, but it can cause symptoms such as fatigue and chest pain and can also increase the risk of cardiac arrhythmias and heart failure.

In most cases, since the patients are typically asymptomatic, this finding is incidental, found either by imaging studies or during autopsies [1]. Over 200 cases have been described [1] in the literature. We describe the autopsy case of a patient with sudden cardiac death caused by massive cardiac lipomatosis of the right heart with dysfunction of the atrioventricular node.

Case Report

A 75-years-old-woman, obese, was hospitalized at the

Policlinic Hospital of Bari-Southern Italy to be submitted to surgery for a locally advanced carcinoma of the tongue. The hospital course was regular, but six days after the operation the patient suddenly manifested atrial fibrillation on ECG, hypertensive crisis, blood desaturation, and an increased troponin test. The patient died of cardiovascular arrest and, in the absence of a certain cause of death, an autopsy was performed. At autopsy, the weight of the heart was 460 g due to symmetrical hypertrophy of the left ventricle and the following diameters were recorded: longitudinal 10 cm, transverse 11 cm, antero-posterior 6,5 cm; free wall thickness of the left ventricle 1.5 cm; interventricular septum thickness 1 cm; right ventricle free wall thickness 0.7 cm; on cut section, an extensive adipose tissue infiltration involved the anterior wall (Figure 1 A) and the infundibulum of the right ventricle, the right atrium (Figure 1 B - C) and the atrioventricular junction (Figure 1 D). In particular, the lipomatous infiltration of the atrium exhibited a pseudo-tumoral shape protruding into the lumen between the septum and the coronary sinus orifice. Coronary arteries were patent with few neither obstructive nor complicated atherosclerotic plaques. Histologically, a widespread adipose infiltration of the right ventricle (Figure 2 A) and right atrium (Figure 2 B) was observed, with mild interstitial and subendocardial fibrosis, hypertrophy of residual cardiomyocytes

and scattered interstitial lymphocytes; a fibrotic degeneration of the atrioventricular node was also noted (Figure 2 C - D). No morphological changes associated with myocardial infarction were detected. The pathological report concluded for sudden cardiac death in patients with massive cardiac lipomatosis of the right heart associated with fibrotic degeneration of the atrioventricular node.

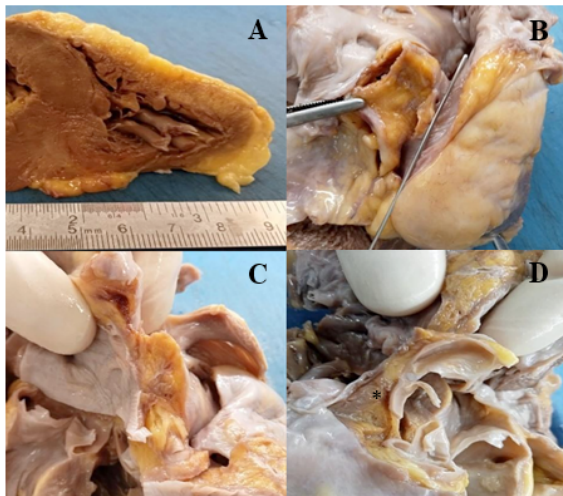


Figure 1: A) Adipose tissue infiltration of the anterior wall of the right ventricle (Transverse section). B - C) Right atrium (Free wall and atrial septum; probe in coronary sinus). D) Atrio-ventricular junction (asterisk).

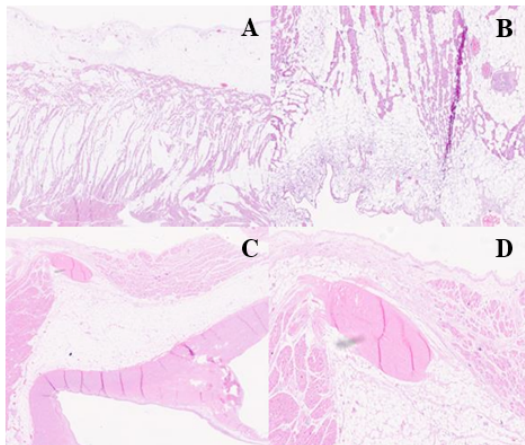


Figure 2: Haematoxylin-Eosin. Massive adipose infiltration. A) Right ventricle (Magnification 2,5x). B) Right atrium (2,5x). C - D) Fibrotic degeneration of the atrio-ventricular node (1,25x and 2,5x respectively).

Discussion

We investigated on the case of a 75-year-old woman,

obese, with atrial fibrillation on ECG, hypertensive crisis, blood desaturation and increased troponin, died by sudden cardiac death after the excision of a tumour of the tongue. The macroscopic and microscopic pathological study demonstrated massive cardiac lipomatosis of the right heart with a concomitant fibrotic degeneration of the atrio-ventricular node. In literature, a clear association between cardiac lipomatosis and sudden death is not reported, probably due to insufficient information about it [2], in fact it is usually considered as a benign, asymptomatic condition, incidentally diagnosed at autopsy. In addition, it is important to emphasize that differential diagnosis with other pathological conditions is necessary, such as adipositas cordis, adipose infiltration of the right ventricle and arrhythmogenic cardiomyopathy of the right ventricle [3]. In the reported case, the absence of significant fibrosis and/or degenerative myocytic changes were in favour of fatty infiltration of both right atrium and ventricle. Various arrhythmias and conduction disturbances have been reported in association with this condition [1]. In the most frequent type of cardiac lipomatosis, the infiltration of the interatrial septum, commonly cause premature atrial contractions, supraventricular tachycardia and atrial fibrillation [4], conversely, the involvement of the atrio-ventricular node can lead to a conduction block [5]. In our case, the massive adipose infiltration of the right atrium and fibrotic degeneration of the atrio-ventricular node could have caused sudden cardiac death by fatal arrhythmia or conduction block.

Conclusions

In the described case, the autopsy demonstrates a massive cardiac lipomatosis of the right heart associated with fibrotic degeneration of the atrioventricular node. Other possible causes of the sudden death were excluded. In particular, we conclude that the sudden cardiac death was caused by a fatal arrhythmia, caused in turn by adipose infiltration of right atrium and fibrotic degeneration of the atrio-ventricular node.

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Informed Consent Statement: Informed consent for publication was obtained from the husband of the patient.

Conflicts of Interest: The authors declare no conflict of interest.

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