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MAIESE A. (*) , BOLINO G.*

A Case of Unexpected Cardiac Death Due To an Abnormality Of The Antolateral Papillary Muscle

(*)Rome University "Sapienza" – Dipartimento di Scienze Anatomiche, Istologiche, Medico Legali e dell'Apparato Locomotore, Viale Regina Elena, 336 – 00161 Rome. Corresponding author: aniellomaiese@msn.com

Key Words: sudden death, anomalies of mitral subvalvular apparatus, ventricular hypertrophy.

ABSTRACT

The unexpected death (UD), despite the identification of more appropriate and targeted preventive and therapeutic strategies, continues to be a "challenge" for modern medicine and even more for the forensic pathologist. In determining the precise cause of sudden cardiac death the only possible way is that represented by the survey autopsy. One cause of UD can be represented by abnormalities of the mitral subvalvular. The case we observed, is a peculiar case of sudden death secondary to papillary muscle supernumerary, in the absence of other causes of death of justification.

RIASSUNTO

Un caso di morte improvvisa conseguente ad un' anomalia del muscolo papillare anterolaterale.

La morte improvvisa (MI), nonostante l'individuazione di sempre più adeguate e mirate strategie preventive e terapeutiche, continua a rappresentare una "sfida" per la medicina moderna e ancor più per il patologo forense. Nello stabilire la causa precisa di morte improvvisa cardiaca l'unico mezzo possibile è quello rappresentato dall'indagine autoptica. Una causa di MI può essere rappresentata da anomalie dell'apparato sottovalvolare mitralico. Il caso da noi osservato, tratta di un peculiare caso di morte improvvisa secondaria a muscolo papillare soprannumerario, in assenza di altre cause giustificative di decesso.

1. INTRODUCTION

Among all the congenital heart diseases, valvular structures abnormalities account for 1% and the mitral changes (0.49%) are the most several, difficult and common [1]. Their physiopathological consequences are stenosis, insufficiency, steno-failure or mitral prolapse [2]. In spite of the constant spotting of adequate and targeted preventive/therapeutic strategies, the unexpected death (UD) is still a "challenge" for the modern medicine and for the forensic pathologist.

Here below, we present a case of UD of a young man, apparently healthy, whose autopsy revealed just the presence of an extra-tip of anterolateral papillary muscle near the mitral valve.

2. CASE REPORT

A 18-years-old boy, waking up in the morning, fainted and instantly died. The medical history of his family only highlighted that the father of the boy died because of uncertain cardiac reasons. The recent and past anamnesis were negative. The external examination revealed multiple small excoriations on the face, with front dental fracture referable to the fall after the fainting. The internal examination didn't show any pathologic change, with exception of the heart. It weighted 360g, 50% depended by each ventricle; the epicardium showed no changes. Seriate section of the left descendant coronary artery showed an intramural course of about 3mm. The anterolateral papillary muscle had a relevant distortion, presenting an accessory tip over the anteromedial edge of mitral valve (*Fig.1)*, whose edges showed no changes. The cut surface of the myocardium had no focal changes; on the contrary, the histological investigation highlighted light myocellular hypertrophy, with focal myocardial disarray prevalently observed at the free wall of left ventricle and subaortic septum , perivascular fibrosis, and hyperplasia of the *tunica media*. The toxicological investigation showed negative results.

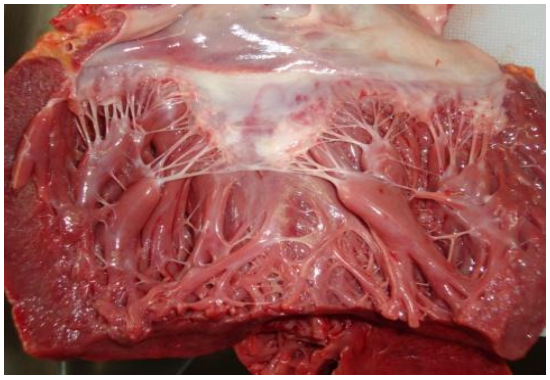
A  B 

FIGURE 1: A, Section of left ventricle showing the accessory tip of the anterolateral papillary muscle. B, insertion of the anterolateral tip of the papillary muscle over the mitral valve.

3. DISCUSSION

A topographical classification [3] subdivides valve anomalies in valvular (valvular edges and valvular ring) and sub-valvular (tensor system: papillary muscles and tendineous cords). Mitral stenosis is mainly caused by congenital fusion of commisures or presence of additional mitral tissue. On the other hand, mitral deficiency is typical in case of *cleft* isolated from the frontal edge of the valve or dysplasia of the edges [4]. In these cases, there is evidence of shortened tendineous cords together with dysplastic valvular edges. When an isolated cleft of the mitral valve, the defect of the flap is directed towards the left ventricular outflow tract. If the valve is dysplastic ,the chordal apparatus is shortened with varying degrees of dysplasia of the flaps.

The most frequent congenital abnormality of sub-valvular structures is the so-called “parachute mitral valve” , that presents one papillary muscle inside the left ventricle or two that are very close to each other. All the tendineous cords, turned to both edges, come from the end of a singular papillary muscle, hence the intercordal spaces are very narrow, causing a sub-valvular obstruction. In adults, this condition can be present as isolated phenomenon. In newborn or infants, often “parachute mitral valve” is associated to aortic stenosis or coarctation, and to a supravalvular mitral membrane, confirming in this way the Shone Syndrome [5]. There is also another anomaly worth to be mentioned, the so-called “mitral one with arch” [6], in which the two papillary muscles are joined on the free limit of the frontal edge, creating a muscular arch. Finally, we can find dysplastic abnormalities of sub-valvular system as lengthening of a papillary muscle, agenesis of papillary muscles, accessory papillary muscles,

lengthening or agenesis of tendineous cords; all these changes may cause valvular deficiency or prolapse [7] .
Sudden unexpected death may mainly have a cardiac or non cardiac etiology . Sudden cardiac death (SCD)may be linked to coronary and non coronary causes (early cardiomyopathies; congenital cardiomyopathies; valvulopathies; etc.). In post-mortem series ischemic cardiomyopathy (IC) is the most frequent cause of SCD, followed by the cardiomyopathies (10-15%) and by valvular diseases [8].
To establish the exact cause of a SCD, the only way is a post-mortem investigation.
However, at autopsy, the heart often looks macro and microscopically normal. For MIC caused by arrhythmia, it can also happen that the myocardic substrate presents subacute or chronic pathologies, and that the precipitant event – "trigger" (stress, electrolytic changes, etc.) remains unknown. The causes of an unexpected death can be classified as: clear causes (i.e. pulmonary embolism, breaking of aorta or heart with cardiac plugging, rupture of papillary muscle, etc.), probable causes (i.e.: arteriosclerotic plates with stenosis >75%, post-infarction scars, cardiomyopathy, etc.), possible causes (i.e.: abnormalities of coronary arteries, mitral prolapse without valvular deficiency, etc.).

The presence of an accessory papillary muscle can cause the development of valvular deficiency or a prolapse of the mitral valve, as well as myocardium hypertrophy [9-10], ideal substrate for the development of uneven phenomena (ventricular fibrillation), one of the causes of unexpected death. In the current case, in light of our findings, we believe that a malignant cardiac arrhythmia is a possible explanation for the patient's death, in the absence of any other pathologic findings or major structural abnormalities, although histo-pathological investigation could have been more accurate to highlight anomalies on a genetic cause of fatal arrhythmias.

Furthermore, we underline the importance to inform the family of a possible, and genetically derived, congenital cardiac pathology [11], in order to evaluate the need to perform preventive diagnostic-instrumental analysis. In the case we examined, the genetic analysis hasn't been done because the Public prosecutor has no interest in investigating the specificity of a natural death; also the mother decided to put off since she has no other children and the previous death of her husband.

In conclusion, we believe that clinicians and forensic pathologists should be aware of this entity both for a more detailed pathologic and anatomic studies of anomalous insertion classification and for differential diagnoses.

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