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Sudden Death in Systemic Amyloidosis– A Case Report

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Abtract:
Hereby, we present a case of an elderly lady who had a previous history of chronic inflammatory disorder and was diagnosed to be suffering from systemic amyloidosis, was brought dead to the hospital with a history of fall from the bed and sudden death nesseciating an autopsy.
Autopsy showed that although all the organs suffered from chronic changes, heart was particularly affected showing gross changes due to restrictive pericarditis. This case demonstrates that cardiac amyloidosis can present acutely and may be fatal. The other gross features along with the post-mortem biochemical parameters, histological changes will be discussed.

Key words: Sudden death, Amyloidosis, Restrictivepericarditis, Histopathology

Introduction:

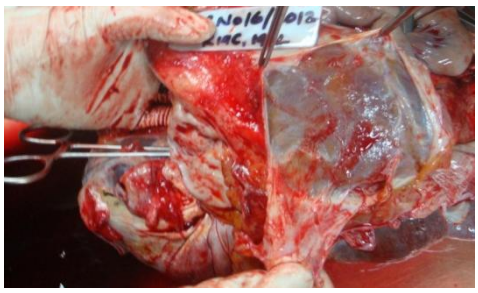
Amyloidosis is not a single disease but a term for diseases that share a common feature: the extracellular deposition of pathologic insoluble fibrillar proteins in organs and tissues. Symptoms vary widely depending upon where in the body amyloid deposits accumulate. Amyloidosis may be inherited or acquired.[1.] In the mid-19th century, Virchow adopted the botanical term “amyloid,” meaning starch or cellulose , to describe abnormal extracellular material seen in the liver at autopsy.[2]

Amyloidosis is a disorder characterized by extracellular deposition of proteins in an abnormal fibrillar configuration. Amyloidosis can be localized or systemic and may affect any organ. The systemic amyloidosis is an uncommon group of disorders characterized by the extracellular deposition of amyloid in one or more organs. Cardiac deposition, leading to an infiltrative/restrictive pericarditis is a common feature of amyloidosis. It may be the presenting feature of the disease or may be discovered while investigating a patient presenting with non-cardiac amyloidosis

CaseReport

History -A 61 year old Female, suffering from systemic amyloidosis from past 6months came with a history of sudden fall from the cot and was brought dead to the

hospitalnesseciating an autopsy. At autopsy on external examination, she was emaciated;there were somesuperficial lacerations over the face.Contusions over the face and upper limbs.Internally all the organs were hard in consistency with thePericardium firmly adherent to the heart a feature seen in case of restrictive pericarditis.As seen below in (Fig– 1)

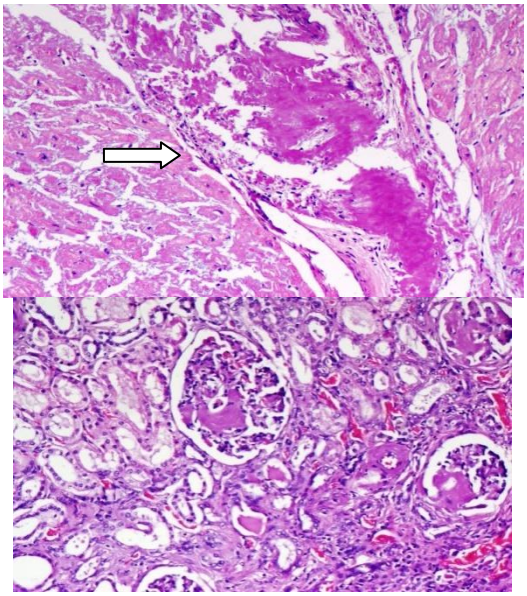


(FIG – 1)

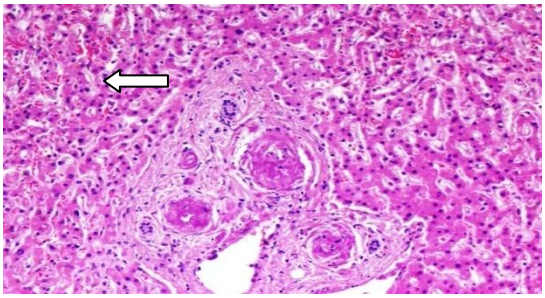
Heart was enlarged, weighing 460grams; hyperaemic area was present over the anterior surface of right ventricle with 50% occlusion of right coronary artery. Left ventricular thickness was 3cms and right ventricular 2cms.Lungs, liver, spleen, and the kidneys were hard in consistency with nodular appearance on cut section.

Blood and viscera were preserved for toxicological analysis and pieces of tissues (heart, lung, spleen and kidney) were sent for histopathological examinationMicroscopic examination:(FIG 2-4) revealed the following

Microscopic Examination – Special stain Congo red was used which showed – positivity in the extracellular eosinophilic material of the (Heart, Liver and Kidney) confirming the amyloid nature of the specimen.



CONGO RED STAINING OF HEART (FIG-2)CONGO RED STAINING OF KIDNEY (FIG-3)



CONGO RED STAINING OF LIVER TISSUES (FIG-4)

Toxicological analysis showed absence of any poison in the body fluids sent.

Postmortem Biochemical Analysis ofthe bloodshowed

CK(MB): 283.8ng/ml [0.6 – 6.3ng/ml]

Troponin-T: 1.88ng/ml [0.00-0.03 ng/ml]

NT PRO BNP: 183.8 pg/ml [40 – 125pg/ml]

Cause of death: was given as- Deceased died due to complications of generalized systemic amyloidosis

Discussion

The amyloidoses are of a series of diseases characterized by the deposition of extracellular

proteinaceous material known as amyloid. Amyloid is a homogenous extracellular material which stains pink with hematoxylin and eosin and which, when stained with Congo red, demonstrates apple-green birefringence if viewed under polarized light. The origin and composition of this material varies considerably,although the appearance of amyloid deposits, whether viewed by light or electron microscopy, is identical among forms. By electron microscopy, the fibrils are seen to be nonbranching, with a consistent diameter of 7.5 to 10 nm.The Congo red appearance of amyloid has been recognized for decades,but with the advent of specialized stainingtechniques, it has become possible to differentiate among the various types of amyloid, thereby allowing specific therapies to be targeted at a specific type of amyloid deposition.[2]

The cardiovascular system is a common target of amyloidosis. Cardiac amyloidosis is classically thought of as an infiltrative cardiomyopathy with subsequent cardiac failure which may be refractory to conventional treatment. [5] It may mimic the clinical picture of constrictive pericarditis and electrocardiographic changes of healed infarction.[5]

Myonecrosis and small vessel ischemia due to amyloid deposit cause an increase in cardiac troponins, whereas diastolic dysfunction and increased genetic expression of natriuretic peptide genes in the amyloid infiltrated ventricles cause increase in plasma BNP levels.[4]

Sudden cardiac death is common in patients with amyloidosis involving the heart. In most other cardiacdiseases associated with sudden death such as coronaryartery disease, dilated and hypertrophic cardiomyopathy,ventricular fibrillation is the commonest etiology ofsudden death and the implantable cardioverter-defibrillator(ICD), used either as prophylactic therapy orimplanted in a survivor of aborted sudden death hasconvincingly been shown to prolong life. Althoughisolated cases of recurrent ventricular arrhythmia withappropriate ICD function have been documented incardiac amyloidosis, most deaths appear to be due topulseless electrical activity, an event not generallyamenable to therapy.[2]

Conclusion

Heart is the organ that normally determines prognosis, death usually occurring as a result of refractory heart failure or sudden arrhythmic death. [3,5]Diagnosis of cardiac amyloidosis requires a high index of clinical suspicion.[3,4]It should be considered whenever a patient presents with heart failure of unexplained cause and a restrictive pattern of cardiomyopathy on echocardiography “sparkling appearance”.[3]

The cardiovascular system is often affected by amyloidosis— the extracellular deposition and accumulation of insoluble fibrillar proteins. The heart is targeted most frequently in the primary and age related forms of amyloidosis, less frequently intransthyretin familial amyloidosis, and only rarely in secondary forms of amyloidosis. Cardiac infiltration results in cardiac symptoms dominated by congestive heart failure, arrhythmias, and cardiomyopathy. The diagnosis of amyloidosis requires a multidisciplinary approach, including clinical examination, biochemical tests, imaging, and genetic analysis, and should be confirmed by Congo red staining in polarised light of a tissue sample. In addition, immunohistochemistry should be used todefine the protein fibril type because it is of diagnostic, prognostic, and therapeutic importance.[6]

The diagnosis of amyloidosis requires tissue sample confirmation. Congo red staining in polarised light is the method of choice at the present. However, the

pathologist should not only make the generic diagnosis of the presence ofamyloid, but should also determine the protein fibril type by
means of immunohistochemistry, because it is of diagnostic,prognostic, and therapeutic importance.By definition, the only diagnosis of amyloidosis is morphological assessment of cytological/histological or post-mortem material.[6]
Inthe past few years advances in molecular biology, immunology, and chemotherapeutic techniques have combined to vastly improve our understanding of the amyloidosis.[4,5]

This review presents the current clinical and diagnostic approach to amyloidosis, with the emphasis on cardiovascular involvement.[6]

Statement of Conflict of Interest

The authors declare that there are no conflicts of interest

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