Case Report

Indian Journal of Forensic Medicine and Pathology
Volume 14 Number 2, April - June 2021
DOI: http://dx.doi.org/10.21088/ijfmp.0974.3383.14221.16

An Autopsy Study of Rheumatic Heart Disease: A Prevalent Iceberg Disease

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How to cite this article:
Muhammed Aseel Zahir Hussain, Archana B, Thanka J *et al.,*/An Autopsy Study of Rheumatic Heart Disease: A Prevalent Iceberg Disease/ Indian J. Forensic Med Pathol. 2021;14(2):121–124

Abstract

Cardiovascular diseases are a chief cause of sudden death. Rheumatic Heart Disease (RHD) exists as a hidden burden in developing countries. It occurs as a sequelae to Rheumatic Fever caused by Group A β Hemolytic Streptococcus. Despite the existence of anti-biotics and prophylaxis by Penicillin it is still prevalent.

A 33-year-old man was found unconscious in his house and was later found dead. He had a history of abdominal pain for the past six months for which he had been on medication. There was no other relevant past history about him or his family. He had no history of either alcohol consumption or smoking. Complete medico legal autopsy was done and his death was confirmed to be RHD.

RHD is an "iceberg" disease which is still prevalent in developing countries. A cross reaction between foreign antigen and cardiac proteins leads to formation of autoantibodies causing autoimmune reaction. Mitral valve is commonly involved. RHD diagnosis can be established using Jones criteria during clinical examination.

RHD still exists despite use of advanced anti-biotics and Penicillin prophylaxis and needs to be therefore considered as a diagnosis of sudden death of young adults.

Keywords: Caterpillar Cells; Sudden Death; Rheumatic Heart Disease.

Introduction

Cardiovascular diseases are the most significant and prevalent cause of sudden death in a person. Rheumatic Heart Disease (RHD) continues to be a burden in developing countries. It causes significant morbidity in young adults. It occurs as a complication of Rheumatic Fever caused by Group A β Hemolytic Streptococcus in genetically susceptible host. In the past, RHD patients occupied a significant portion of the beds in hospitals of developed countries. In the recent past rheumatic fever has receded in developed countries.

RHD is a preventable condition, if intervention is made at the right time. In the era of advanced antibiotics and penicillin prophylaxis, it is still prevalent in developing countries like ours. We report an incidentally detected case of RHD in a male after an autopsy on him.

Case Report

A 33-year-old man was found to be in an unconscious state in his house and was later declared dead. He had a history of abdominal pain for the past six months for which he had been on medication. There was, however, no other relevant family history on him. He had no history of alcohol consumption or smoking. A complete medico-legal autopsy was done him.

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Macroscopically: There was evidence of petechiae over left ventricle and congestion of liver, spleen and kidneys. Kidney shows thickening of walls of arterioles. Liver showed centrilobular necrosis and features of chronic venous congestion. Lungs were edematous with alveolar septal thickening and focal fibrosis.

Microscopically: Myocardium showed foci of fibrinoid necrosis, lymphocyte infiltration (Fig. 1) and plump macrophages with abundant cytoplasm, round to oval nuclei with central wavy chromatin (caterpillar cells). (Fig. 2) By IHC, the cells infiltrating the myocardium were positive for CD45, Vimentin and CD68 (Fig. 3) and negative for CK and SOX10, confirming them to be macrophages. Histological features were suggestive of Aschoff bodies (Fig. 4) consistent with the diagnosis of RHD.

Sections from liver showed congested sinusoids with focal centrilobular necrosis. Portal area showed lymphocytic infiltration. Hepatocyte zone 3 demonstrated presence of micro and macrovascular steatosis (10%).

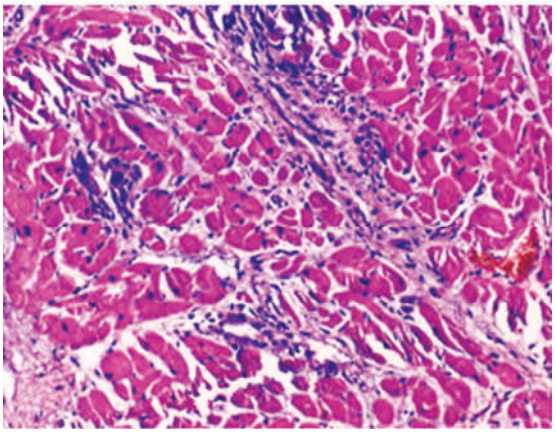

Fig. 1: Histopathological slide (100X) showing features of Myocarditis.

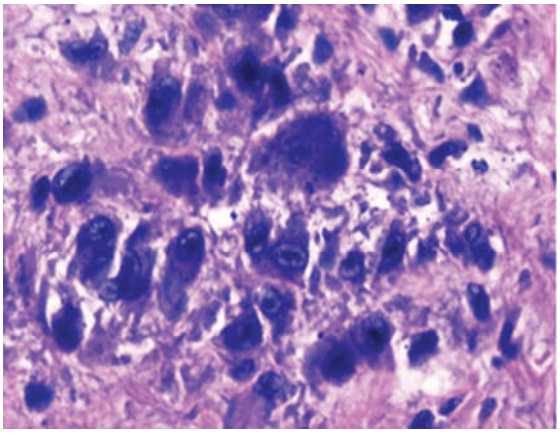

Fig. 2: Histopathological slide(400X) showing Caterpillar cells in the Myocardium.

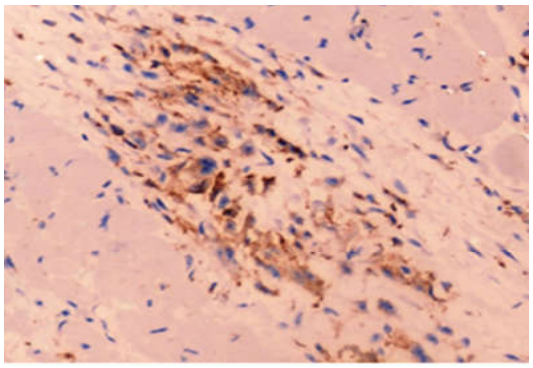

Fig. 3: Immmunohistochemistry; CD68 Positivity noted.

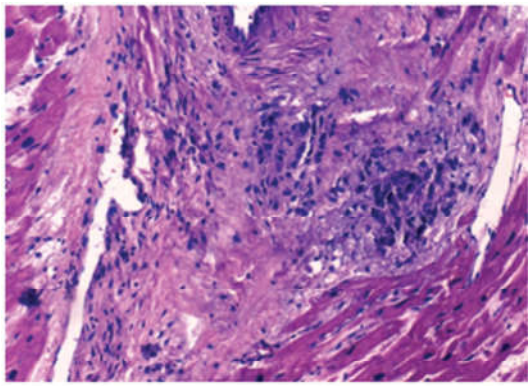

Fig. 4: Histopathological slide (100X) showing Aschoff Bodies.

Discussion

Cardiovascular causes form the most important cause of sudden death. RHD is an important disease causing cardiovascular mortality and morbidity in young adults. It occurs as a complication of Rheumatic Fever by Group A β Hemolytic Streptococcus in a genetically susceptible host due to an autoimmune reaction of the cardiac proteins with the streptococcal antigen.

Epidemiology RHD tends to be more common in females.[1] It also accounts for the greatest cardiovascular related loss of disability adjusted life years in children.[2] The number of disability adjusted life years due to rheumatic heart disease in 2015 was 10,513,200.[3]

RHD has affected 33.4 million people globally and causes 347,000 deaths annually.[4] Premature death contributed more to ill-health in patients than years they lived with disability.

The decrease in incidence in developed countries can be credited to better living conditions resulting in better hygiene, less overcrowding leading to decrease in transmission of infection. Patients suffering from rheumatic carditis are at a high risk of recurrence of RHD and they receive long term antibiotic prophylaxis. Patients with valvular

disease receive prophylaxis for a period of 10 years after the previous attack of acute rheumatic fever or upto the age of 40 years, whichever is longer.

The key issues in the prevention of RHD is the inability to find an effective way for early identification of those with rheumatic heart disease to administer effective prophylaxis rather than going for surgery. The lack of primary preventive measures also adds to the burden of the disease. One of the major roadblocks in the study of RHD in developing countries is the overshadowing of the burden of rheumatic heart disease by other prevalent diseases like Immunodeficiency, Tuberculosis and Pneumonia.

One of the major drawbacks in combatting RHD can be attributed to the reliance on clinical examination for identifying patients of rheumatic heart disease rather than echocardiographic screening citing the lack of funds to do so in a resource-poor set up. But a study conducted on school children by Marijon showed that echocardiographic screening identified nearly 10 times the number of cases detected by clinical examination which represent an exceptional number of cases that could go unnoticed and thus predispose them to the risk of RHD.[5]

This suggests that Rheumatic Heart Disease is another "Iceberg Disease" that warrants attention because of its morbidity. The role of Group A β Hemolytic Streptococcus in RHD has been established due to increase in cases of rheumatic fever following infection and also the increased Anti Streptococcal antibodies like Anti Streptolysin O in patients.

Foreign Antigen M Protein cross reacts with Cardiac Myosin leading to autoimmune reaction against heart valves. Autoantibodies are formed against carbohydrate of heart valve, binding to valve and leading to damage. This causes intracellular cardiac proteins to spill further instigating autoimmune reaction. The most commonly involved valve is Mitral Valve.[6] Vegetations may be present along the line of closure of valve leaflets.

One of the major presenting neural symptoms is Sydenham Chorea. Autoantibodies target dopaminergic receptors leading to altered dopamine levels and thus causing chorea. The diagnosis of RHD is established during clinical examination using Jones Criteria, which includes migratory polyarthritis, carditis, erythema marginatum, Sydenham chorea and subcutaneous nodules. One of the minor criterion for diagnosis is the prolongation of PR interval on ECG.

Echocardiography is the gold standard for the diagnosis of carditis.[7] The most common lesion is mitral regurgitation (MR), while mitral stenosis (MS) is pathognomonic of RHD.[8]

Prophylaxis is administered in order to prevent infection by Group A β Hemolytic Streptococcus. Primordial prevention of RHD mainly involves preventing overcrowding. The strongest evidence for a causal association between a primordial (socio-economic) determinant and RHD risk is for household or bedroom crowding.[9]

Primary prevention of Group A Streptococcus infection includes administration of Benzathine Penicillin and Penicillin V. In case of Penicillin allergy, cephalosporin, clindamycin or Azithromycin can be used.

Secondary prevention of Group A Streptococcus is done by administering Benzathine Penicillin G, Penicillin V and Erythromycin.

Duration of prophylaxis is 10 years since last episode of rheumatic fever or until 21 years of age, whichever is longer with prolongation till 35-40 years. Increased compliance to Penicillin is associated with decreased Acute Rheumatic Fever recurrence and decreased mortality.[10]

The most important prophylactic measure includes the improvement of sanitation, hygiene, nutrition and access to affordable health care.

Interventions like Catheter Intervention is used for stenotic lesions primarily. Patients having severe mitral stenosis benefit the most from catheter-based interventions.[4] Cardiac Surgery is the preferred intervention when patient is symptomatic.

Conclusion

RHD is a prevalent cardiovascular disease in developing countries due to the poor living conditions. It results from an autoimmune etiology. Clinically, Jones criteria serves as a salient method of diagnosis whereas echocardiography has proven to be the gold standard to diagnose carditis. Presence of distinct histopathological features can clinch the diagnosis.

In this modern era of advanced antibiotic treatment, forensic pathologists should still consider RHD as a cause of sudden death in middle aged persons.

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