Isolated cardiac sarcoidosis: An autopsy case report

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Abstract: Sarcoidosis is a systemic disease of uncertain etiology, characterized by non- caseating epitheloid cell granulomas which can virtually involve any organ. Cardiac involvement of sarcoidosis is seen in around 4-5% patients, while autopsy studies proved incidence up to 20-25%. Sudden cardiac death is common in cardiac sarcoidosis, since it was not diagnosed due to silent clinical presentation. We report a case of cardiac sarcoidosis in a 43 years patient, who presented with chest pain and succumbed to sudden death. Autopsy revealed heart showing gross and histopathological findings of sarcoidosis, thus confirmed the diagnosis.

Key Words: Cardiac sarcoidosis, Sudden death.

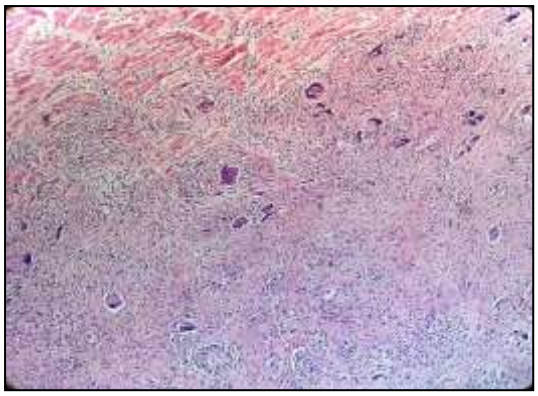
INTRODUCTION:

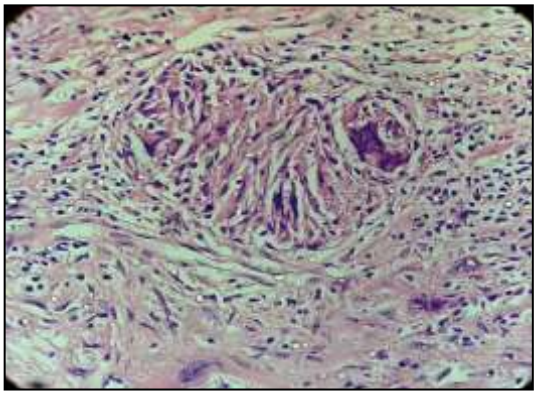
Sarcoidosis is a multisystem granulomatous disorder of unknown etiology which mainly affects lungs, skin, eyes, liver, spleen, lymph nodes and heart. The quoted incidence of clinical heart involvement is around 4-5% while at autopsy at least 20-25% patients show cardiac sarcoidosis and 13-25% deaths occur from the disease. (1) Cardiac sarcoidosis occurs more frequently in young adults. (2) The clinical presentation can vary from benign ectopic to life threatening arrhymias. The diagnosis of cardiac sarcoidosis is often difficult since patients remain clinically silent and cardiac dysfunction is the only sign. The outcome of most of the patients that are not diagnosed in time is sudden death. (3)

CASE REPORT:

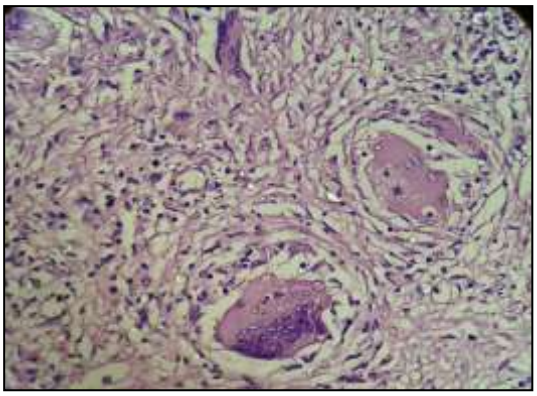
We report a case of cardiac sarcoidosis in a 43 years male patient who presented with chest pain and succumbed to sudden death. Viscera were sent for histopathological examination. Grossly the heart was weighing 300 grams and there were multiple whitish fibrotic areas distributed all around the inter-ventricular wall, myocardium of left ventricle wall (fig 1). Multiple sections were processed from affected area. Microscopy revealed multiple non- caseating granulomas comprised of epitheloid cells, lymphocytes and giant cells (fig2). Asteroid bodies also seen (fig 3).

Multiple whitish fibrotic areas distributed all around the inter- ventricular wall, myocardium of left ventricle wall (fig 1).





Multiple non- caseating granulomas comprised of epitheloid cells, lymphocytes and giant cells (fig2) and fig(2.1).



Thus clinical history, gross findings in heart and histological sections showing multiple non-caseating granulomas in myocardium confirmed the diagnosis of sarcoidosis. We also did special stains like ZN for AFB bacilli, PAS and SM to rule out other causes. The autopsy report was finalized as cardiac sarcoidosis to be the cause of sudden death after ruling out all other causes of granulomatous lesions like tuberculosis affecting the heart. Sine this patient died suddenly hence detailed clinical investigations reports were not available.

DISCUSSION:

Cardiac sarcoidosis is a potentially fatal condition, not frequently diagnosed during clinical examination. It is an incidental autopsy finding. Studies done by Michael et al., Robert et al., and Fleming suggest that sarcoidosis was the cause of sudden death in most patients.[4] Most patients with cardiac sarcoidosis have little or no clinical evidence, of extra-cardiac sarcoid involvement. The main cause of death is arrhymias and progressive cardiac failure. Sudden death may be the 1st manifestation in most of the cases, therefore isolated cardiac sarcoidosis are only diagnosed on post- mortem examination [5, 6] The diagnostic criteria included as per Japanese expert is as: 1) Histological diagnosis showed non-caseating granulomas in myocardium, through biopsy and surgical sections.20 Clinical diagnostic features included are AV block, ventricular arrhymias, CHF and Sudden death.[6] Endo myocardial biopsy is the gold standard for the diagnosis of cardiac sarcoidosis. [7] The diagnosis of cardiac involvement in sarcoidosis may be missed because of patchy involvement.

Our study discussed isolated cardiac sarcoidosis presenting as sudden cardiac death. FDG-PET/CT imaging has higher sensitivity for the detection of sarcoidosis in myocardium. [8] Steroid therapy is the primary treatment of choice for cardiac sarcoidosis. [9]

CONCLUSION:

Cardiac sarcoidosis should be considered in young patients who present with heart failure or sudden death, due to its fatal clinical outcome and silent clinical presentation. Thus Cardiac sarcoidosis

remains a challenge, despite the availability of modern diagnostic modalities. This case is presented here for documentation purpose and to emphasize on subclinical cases which go unnoticed because of lack of awareness.

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