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Tropical Endomyocardial Fibrosis: Case Report

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ABSTRACT

Tropical endomyocardial fibrosis is a peculiar disease involving the heart. Recent reports on it have appeared in alphabetical order in Brazil, Casablanca, France, India, Mozambique and the United Kingdom. Therefore, this case report from Nigeria is deemed to be worthy of documentation.

Keywords: Heart, Muscle, Fibrosis, Etiology, Early death

INTRODUCTION

Endomyocardial fibrosis is also known as Tropical Endomyocardial Fribrosis, is a unique cardiac cellular lesion, which is reportedly replaced by fibrous tissue [1,2]. Single cases were reported from Brazil (1), Casablanca (2) and the UK (8). Therefore, the purpose of this report is to present a Nigerian case in which an indigene of the Igbo ethnic group was involved [3]. This was facilitated by the establishment of a Regional Pathology Laboratory at Enugu, the erstwhile capital city of Eastern Region of Nigeria with the corresponding author as the pioneer pathologist. In this context, it had been considered by the Birmingham (UK) group that the establishment of a histopathology data pool facilitates epidemiological analysis [4].

CASE REPORT

The patient was a man aged 21 years that consulted one of us (CN) at the Enugu Specialist Hospital, Enugu. The clinical summary was as follows: Ill for 8 years with febrile episodes, anorexia and breathlessness, worse on exertion. Swelling of abdomen and legs was noticed 2 months after the onset of the illness and had been present since then. Hospitalized four times, he was operated on for abdominal pains. No major illness in childhood, playing normally. On examination, he was emaciated. Ascites. ++ Leg edema +. JVP over 10 cm. Irregular fibrillating pulse. Normotensive. Weak apex beat in 5th intercostal space along mid-axillary line. Much increased area of precordial dullness. Well marked triple rhythm. S3 heard all over. Accentuated pulmonary second sound. Systolic murmur. Liver concealed by ascites but edge was roughly 4 fingers below the ribs. Solid slightly mobile mass was felt at the lower abdomen. Diagnosis: “Endo myocardial fibrosis.”

The correspondent author’s postmortem report

External appearances: The body was that of an emaciated young man who was very pale. The ankles pitted on pressure. Both legs were slightly sabre shaped, especially on the left, whose convexity was surmounted by a large healed ulcer. The umbilicus was absent, there being a transverse abdominal surgical scar, which suggested having undergone operation probably for the repair of umbilical hernia. The abdomen was greatly distended and a mass was palpable on the left side. The right breast was slightly enlarged. There was no lymphadenopathy or finger clubbing.

Internal appearances:

Abdomen: On opening the peritoneum, there was much blood-stained fluid. Layers and layers of fibrinous matter with purulent looking parts were noted. All the organs were matted together; the mass palpated externally being seen to be a spurious tumor formed by coils of the intestine. A small quantity of altered blood was present in the stomach, but the source could not be identified. Melena was apparent in the lower gut. Both the spleen, which was slightly enlarged, and the liver were unsheathed by white fibrous tissue, the

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parenchyma being congested. The pancreas and kidneys were not remarkable.

Chest: Both pleural cavities contained much straw colored exudates, the lungs lying free, except for firm adhesions over the left apex. On section, both lungs were edematous. The pericardium was greatly dilated, occupying much of the chest cavity. It contained about a liter of slightly blood stained fluid. The heart itself was picturesque. A few whitish plaques were noted on the external surfaces of the atria and ventricles, especially the left ventricle. Enlargement was striking on the right side, especially the atrium. Both ventricles exhibited thickened walls. The atrial walls were dilated, being paper thin on the right side. The endocardial surface was glistening white throughout the left atrium, especially around the valves. The right chamber was less impressive in this respect. As for the ventricles, the left was also more generalized and thicker, especially in the apical area, whereas in the right chamber this was limited to the posterior areas. Of the valves, both the pulmonary and the aortic ones were unexceptional, but both atria-ventricular ones were thickened. Their chordae tendinae were shortened, especially on the right side. No thrombi were found.

Head: Not examined.

Microscopy: The myocardium was the seat of considerable fibrosis which special staining confirmed to be more marked nearer the endocardium. Lymphocytic infiltrates were noted in the epicardium. Both layers of the pericardium were gummed up in parts by inflammatory infiltrates which featured lymphocytes and plasma cells with occasional polymorphs. The peritoneum was similarly affected, fibrinous exudates abounding as well as both organizing and organized exudates. Perisplenitis was evident together with a covering of fibrinous exudates. These appearances were obvious on the liver, whose parenchyma was the seat of chronic venous congestion and incipient cirrhosis. The lung showed patchy edema and interstial pneumonia; a single localized area of eosinophilic granuloma was identified. Gynaecomastia was present. Apart from autolysis and congestion, the pancreas, thyroid and kidney revealed nothing noteworthy.

DISCUSSION

Although a 63 year old female sufferer was reported from Brazil (1), the commonest age ranges were described as "highest among persons 10 to 19 years of age" [5], "mainly children and adolescents" [6] or "predominantly children and young adults" [2]. Hence, our patient, aged 21 years, fits the above pictures.

Incidentally, our female patient was not in tune as regards sex predominance. Thus, although Mozambique authors found that the incidence is that of males being higher than in females [5], the Brazilians wrote in terms of "Gender seems to lack any predominance, and varies according to the series and the country of the study" [1].

Demise was usually due to heart failure [1,2,7,8]. It was prominent in our patient.

Marked ascites was noted in France [6]. So it was locally.

Intraventicular thrombus formation was generally a feature. This was linked with the valvulopathy seen in France [7]. Here, there was the specific "No thrombi were found." If anything, there were "fibrinous exudates abounding as well as both organizing and organized exudates."

CONCLUSION

Tropical endomyocardial fibrosis "remains a mysterious and challenging cardiovascular disease" [8]. Some answers have come from countries as far apart as Brazil (1), Casablanca (2), France (4), India (5), Mozambique (6,7) and the UK (8). Therefore, it is deemed to be important to add a Nigerian case; it is from the Ibo ethnic group [3]. In sum, forensic autopsy of a 21 year old man confirmed the existence of this odd disease in this African country.

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