

CASE REPORT

ENDOMYOCARDIAL FIBROSIS ASSOCIATED WITH SCHISTOSOMA HAEMATOBIIUM INFECTION

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SUMMARY

Endomyocardial fibrosis (EMF) is a form of restrictive cardiomyopathy common in the tropics and subtropics. The aetiology of EMF is unknown but helminth infestations such as schistosomiasis have been implicated. Two boys aged 8 and 10 years with EMF associated with *Schistosoma haematobium*, are described. The schistosomes in both cases may have been acquired from contact with contaminated water collected and stored in containers and subsequently used for bathing. Both patients were managed conservatively. Overall prognosis of EMF is poor and this report emphasizes the importance of public health interventions in the control of schistosomiasis.

Keywords: Endomyocardial fibrosis; *Schistosoma haematobium*; Cardiomyopathy; Ghana; Public health intervention

INTRODUCTION

EMF is the most common restrictive cardiomyopathy in the tropics and subtropics and a cause of death in these areas.¹ The disorder is caused by deposition of fibrous tissue on the endocardial surfaces, resulting in impaired filling of one or both ventricles.² The aetiology of EMF remains unclear although it is frequently associated with parasitic infestations.³

Cases of EMF associated with *Schistosoma mansoni* disease are published in the literature.^{4,5} In Ghana, *Schistosoma haematobium* is the predominant schistosome species with a prevalence of up to 60% in some communities.⁶ Infestations occur through contact with water contaminated with cercariae, the free-living infective stage of the parasite, which penetrate intact human skin and cause urinary schistosomiasis.⁷

To the best of our knowledge this is the first report of EMF associated with *S. haematobium* in the West African sub-region.

CASE REPORTS

Case 1

An 8-year-old boy from Big Ada in the Greater Accra Region of Ghana presented with a distended abdomen of a year's duration and worsening respiratory distress. He had mild pedal swelling, orthopnoea and associated weight loss. His urine was amber and of adequate volume. He had been treated with praziquantel for schistosomiasis, two years prior, as part of a community screening exercise. He denied ever wading or swimming in the nearby Volta lake, but the lake was the family's source of water for domestic activities including bathing.

On examination, he looked chronically ill with massive abdominal distension and bilateral pitting oedema up to the thigh. He was dyspnoeic with reduced breath sounds on the left side of the chest. Blood pressure was 100/76 mmHg and heart rate, 100/min. Heart sounds were muffled with no audible murmur. His abdomen was grossly distended and massive ascites was demonstrated by a positive fluid thrill. No abdominal masses were ballotable.

Investigations

Haemoglobin was 9.4g/dl, total white cell count $6.4 \times 10^9/L$ with eosinophils $0.3 \times 10^9/L$. Sickling test was negative. ESR was elevated at 54mmfall/hr and liver function tests showed a low albumin of 24g/L. HIV and Mantoux tests were negative and renal function was normal. Urinalysis was also normal and microscopy was negative for schistosoma ova.

Stool microscopy was negative for helminths. The schistosome specific antibody test for *Schistosoma haematobium* was positive for IgG and negative for IgM. Both IgG and IgM were negative for *Schistosoma mansoni*. Chest x-ray revealed cardiomegaly and a left-sided pleural effusion. ECG showed sinus rhythm, low voltages and tall P waves. Echocardiogram showed a very large right atrium, thickened and calcified right ventricular apex and small right ventricle. Left heart chambers were normal in size and function. A moderate pericardial effusion was present with no evidence of cardiac tamponade. These findings were consistent with endomyocardial fibrosis.

The patient was managed conservatively on diuretics and also received one dose of Praziquantel. He subsequently improved, with resolution of respiratory distress and decreased abdominal distension, and was discharged after 21 days, for continuing follow-up as an outpatient. A letter was sent to the chief of his home town and public health officials concerning the patient's diagnosis and possible link to schistosomiasis.

Case 2

A 10-year-old boy from Kpando in the Volta Region of Ghana presented with progressive abdominal distension of 7 months duration and fever and cough of 3 months duration. He also had dyspnoea and weight loss. There was no facial oedema, urine volume was unchanged and there was no past or present history of gross haematuria.

The water used for domestic activities was collected from a nearby river whose source was the Volta Lake. The patient had never swum or waded in the river. On examination, he looked chronically ill with gross abdominal distension. He had no lymphadenopathy or clubbing. There was minimal pedal oedema. He was dyspnoeic with a respiratory rate of 40/min and heart rate of 92/min.

The apex beat was located in the 5th left intercostal space at the mid-clavicular line and was normal in character. His blood pressure was 100/55mmHg. Heart sounds were distant with no audible murmur. The abdomen was grossly distended with marked ascites but no masses were ballotable.

Investigations

Haemoglobin was 10.8g/dl, total white cell count $6.4 \times 10^9/L$ (eosinophils $0.5 \times 10^9/L$) and ESR 65mmfall/hr. Liver and renal function tests were normal. Urinalysis showed microscopic haematuria with no proteinuria, leukocytes or casts and microscopy was negative for

Schistosoma ova. Stool microscopy was negative for helminths. HIV and Mantoux tests were negative.

Ascitic fluid biochemistry was normal and no acid fast bacilli were seen. IgG antibodies to *S. haematobium* was positive and IgM negative. IgG and IgM were both negative for *S. mansoni*. Chest x-ray showed cardiomegaly and ECG showed sinus rhythm, tall P waves and non-specific T wave changes. Echocardiogram showed severe dilation of the right atrium, mild dilatation of the right ventricle and thickening of the anterior aspect of the right ventricular wall. The left ventricle was normal and a small pericardial effusion was present. The findings were consistent with endomyocardial fibrosis. He was managed on diuretics and received one dose of praziquantel. The microscopic haematuria resolved within two weeks and he was discharged also with a letter to the chief of his town and public health officials.

DISCUSSION

Endomyocardial fibrosis is a major cause of death in areas where it is endemic although the pathogenesis is not completely known.¹ In EMF, ventricular thrombosis results in thickening and fibrosis of the endocardial surface of the heart, leading to reduced compliance and restrictive physiology.^{2,3} The disease is associated with a poor prognosis and treatment is largely conservative.^{1,2} As was seen in our patients, symptomatic therapy with diuretics may be useful.¹ For patients with severe symptoms surgery may be required although this option is largely unavailable in most resource-poor countries where EMF predominates.³

EMF is frequently associated with concomitant parasitic infestations and their attendant eosinophilia.^{4,8} It has been proposed that the initial myocardial damage may be associated with abnormalities of eosinophils.^{8,9} There are published reports of EMF associated with *S. mansoni* infection.^{4,5} Our patients did not have any evidence of *S. mansoni* infection but IgG to *S. haematobium* was positive. EMF associated with co-infection of *S. haematobium* and *intercalatum* has been reported previously from Equatorial Guinea.¹⁰

In Ghana, *S. haematobium* infection remains a public health problem, with the highest prevalence seen in communities closest to the Volta Lake, where inhabitants are completely dependent on lake water for their domestic use.^{6,7,11} Traditionally, infestations occur when people wade, swim or walk in water containing the infectious larvae or cercariae.⁷ Neither of our patients admitted to going into the infested water body but had used the water for bathing at home.

Clinicians must be aware that disease transmission to humans may occur from various forms of contact with contaminated water and not just in those who swim or wade in the water body.

The possible association with EMF is another reason to intensify efforts at control of schistosomiasis. Effective public health interventions include health education, provision of safe drinking water and toilets, treatment of established infestations including routine screening in high risk areas and mass treatment with praziquantel.^{7,12} Support must also be given for more research in the development of antischistosome vaccine as well as newer drug targets.^{13,14}

CONCLUSION

EMF may occur in children infested with *Schistosoma haematobium*. Efforts to control schistosomiasis in endemic countries should be intensified.

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