COR-PULMONALE- A RARE ASSOCIATION WITH SCHISTOSOMA HAEMATOBIUM INFESTATION

Asad Mahmood, Muhammad Tariq*, Muhammad Saeed**

AFIC Rawalpindi, *Military hospital Rawalpindi, **PNS Hafeez Islamabad

INTRODUCTION

After malaria and amoebiasis, Schistosomiasis (bilharziasis) is the third leading endemic parasitic disease in the world. The main disease-causing species are S haematobium, S mansoni, and S japonicum. S. mansoni is found in Africa, the Arabian Peninsula, and South America; S. japonicum is found in Japan, China, and the Philippines; and S. haematobium is found in Africa and the Middle East. In nonendemic countries the prevalence of imported schistosomiasis is steadily rising owing to an increase in traveling to endemic areas[1].

The chronic schistosomiasis, the third stage, is caused by the heavy deposition of eggs in the intestine, bladder and the liver. It results from the host's immune response to schistosome eggs and the granulomatous reaction evoked by the antigens they secrete. Eggs of S. mansoni and S. japonicum embolize to the liver, where the granulomatous inflammatory response induces periportal fibrosis. Periportal collagen deposition leads to portal hypertension, collateral esophageal varices, splenomegaly, and hypersplenism. Eggs may then be shunted from the liver to the lung, with the possible sequela of pulmonary hypertension. S. haematobium infection occasionally causes mild colonic or hepatic disease. Urinary tract disease is a specific trait of infection with S. haematobium[2].

Infection with S haematobium is usually associated with such sequelae as obstructive uropathy, renal failure and bladder cancer.[1] We present a case of Schistosoma haematobium infestation with pulmonary hypertension and cor-pulmonale in which the pattern of illness was atypical.

Correspondence: Lt Col Muhammad Tariq, Classified Medical Specialist, Military Hospital Rawalpindi
E-mail: docarique@gmail.com
Received: 19 Sep 2008; Accepted: 15 Oct 2008

CASE REPORT

A 46 year old Liberian presented at Pak Level II Field Hospital, Tubmanburg, Liberia on 13 June 2006, with 2 month history of progressive swelling of his feet. He also had mild dyspnoea and fatigue on moderate to severe exertion. He denied chest pain, paroxysmal nocturnal dyspnoea, syncope, cough, fever, chills, flank pain, weight loss, or other constitutional symptoms. There was no history of gross haematuria and terminal dysuria. There was no past history of any significant illness. He recalled no prior episodes of haematuria.

He was born in Guinea and migrated to Liberia at the age of 16 years and settled at Boomy county. He had to flee to Seirra Leon 16 years earlier due to civil war in Liberia but came back to Boomy county after 7 years and since then he had been in Boomy county, Liberia. He had been engaged in many jobs including agriculture, fishing and rubber collection. He had a long history of repeated fresh water contact in Boomy county. He had been taking alcohol occasionally but there was no history of smoking.

On examination he appeared comfortable and had an opacity of left cornea. His vital signs were normal and there were no skin lesion.. His JVP was raised and he had oedema of both feet. Rest of his general physical examination was unremarkable. Cardiac examination revealed split of the second heart sound with accentuation of its' pulmonary component. His lungs were clear on auscultation, there was no neurologic deficit and no organomegaly was detected on abdominal examination.

His Hb was 11.9g/dl, ESR 30mm/hour, TLC, DLC and platelet count were normal with no increase in eosinophils. His S. urea was 9 mmol/L and Creatinine 118 mic mol/L. His S. bilirubin was 10 micmol/L, ALT 32 U/L and Alkaline Phosphatase 176 U/L. His urinalysis showed trace protein, numerous pus cell and

thin walled ovoid eggs with terminal spines suggesting ova of Schistosoma haematobium. Results of an HIV antibody test were negative.

His ECG showed right axis deviation, right ventricular hypertrophy and other nonspecific ST segment and T wave abnormalities. His CXR suggested cardiomegaly with pulmonary hypertension. Pulmonary hypertension was later confirmed by cardiac ultrasonogaphy. No calcification was seen on abdominal radiography. His Ultrasound of abdomen revealed mild to moderate dilatation of pelvicalyceal system of both kidneys with dilatation of both ureters. The kidneys were normal in size and structure. Urinary bladder showed thickening of its' wall but had no mass or calculus. Prostate was of normal size and texture. Rest of the abdominal viscerae showed no abnormality.

He was advised tab Cysticide (Praziquantel 600mg) 2 stat then 2 after 8 hours i.e., a total dose of 40mg/kg body weight in two divided doses, salt-free diet and a diuretic. He was also advised tab ciprofloxacin 500mg BID empirically for his UTI after obtaining his urine for culture sensitivity. He reported back to us after few days with slight improvement in his symptoms. Then he was was lost to follow-up.

DISCUSSION

Occasionally, Schistosoma eggs are deposited outside the genitourinary system. These ectopic eggs may go unnoticed or may cause severe pathology, depending on their position. It is the ova and the host response to them that are the major cause of morbidity and urologic sequelae of S haematobium infection. The severity of infection and the frequency of complications is related directly to egg burden[3]. In the case presented the egg burden was great as there were numerous ova per high power field of urinary deposit examined.

Haematuria is the first sign of established disease, appearing 10 to 12 weeks after infection. Dysuria and haematuria are common in both early and late disease. Although late manifestations like thickening of bladder, dilatation of the ureters and renal pelvicalyceal system were present but gross haematuria was absent at the time of presentation in the case presented and neither did the patient recall any such symptoms in the past.

The lung is mandatory step in the parasite cycle but pulmonary manifestations are limited. There are two forms of pulmonary involvement. The acute form usually occurs about 6 weeks after the infection as a part of Katayama syndrome. Pulmonary manifestations during the early stage of schistosomal infection may occur with either S. haematobium or S. mansoni infection. Nonspecific influenza-like symptoms including cough and transient chest radiographic abnormalities can occur. The chronic form is seen in endemic areas and may cause pulmonary hypertension and cor pulmonale, pulmonary granulomatous schistosomiasis, and pulmonary arteriovenous fistulas[4].

The cardiopulmonary bilharziasis is more frequent on pathological examination than on clinical features. Evidence of pulmonary involvement has been reported in 5-54% of patients. The diagnosis of cardiopulmonary bilharziasis is made by pulmonary biopsy which shows an occlusive angiitis with chronic inflammatory cells, and arteriolar medial hypertrophy caused by a granulomatous inflammatory response to ova trapped and killed in the circulatory bed. The prognosis is dependent upon development of pulmonary hypertension and schistosomal cor pulmonale. If the granulomatous response is both widespread and vigorous, mechanical obstruction of the vascular bed follows resulting in pulmonary hypertension and cor pulmonale[5]. The index case had clinical evidence of pulmonary hypertension and cor-pulmonale. In less than 5% of infections, schistosomal egg obstruction of the lung vasculature results in pulmonary hypertension and cor pulmonale. Limited data suggests that cardiopulmonary schistosomiasis is seen most often in S. mansoni infections and is rare in S. haematobium infections. Hepatic fibrosis and portal hypertension appear to be a prerequisite to the development of schistosomal cor pulmonale caused by S. mansoni. The premortem diagnosis of cardiopulmonary

schistosomiasis depends on the detection of viable schistosomal ova in stool or urine along with evidence of hepatic fibrosis and pulmonary hypertension. Although treatment with praziquantel can effectively eradicate all schistosomal infections with minimal toxicity, cardiopulmonary manifestations are not likely to be reversible given the chronic fibrotic tissue changes that are present[6, 7].

In patients presenting with obstructive uropathy, portal hypertension, pulmonary hypertension or cor pulmonale; chronic schistosomiasis should always be ruled out if they give history of past travel to endemic areas of this disease.

REFERENCES

1. Whitty CJM, Mabey DC, Armstrong M, et al. Presentation and outcome of 1107 cases of schistosomiasis from Africa diagnosed in a non-endemic country Trans R Soc Trop Med Hyg 2000; 94: 531-4.
2. King CH. Disease in schistosomiasis haematobia. In: Mahmoud AAF, ed. Schistosomiasis. London: Imperial College Press, 2001:265-95.
3. Smith J, Hyg M, von Lichtenberg F. Parasitic diseases of the genitourinary system. In: Walsh P, Retnik A, Vaughan E, Wein A, eds. Campbell's Urology. 7th ed. St Louis: WB Saunders Company; 1998:734-57.
4. Bethlem EP, Schettino GP, Carvalho CR. Pulmonary schistosomiasis. Curr Opin Pulm Med. 1997; 3: 5: 361-5.
5. Robotham JL. Schistosomal Cor pulmonale: A Fluke in the Fas Lane?. Respiration 2003; 70: 569-571.
6. Morris W, Knauer CM. Cardiopulmonary manifestations of schistosomiasis. Semin Respir Infect. 1997; 12: 2: 159-70.
7. Waldman AD, Day JH, Shaw P, Bryceson AD. Subacute pulmonary granulomatous schistosomiasis: high resolution CT appearances--another cause of the halo sign. Br J Radiol. 2001; 74: 887: 1052-5.