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Instructive Case

13-Year Old Boy from Northern Samar with Fever and Cardiomegaly

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A 13-year old male from Northern Samar was admitted for recurrent episodes of fever. Since 3 years prior to admission, the patient had monthly bouts of high grade fever and chills, lasting for about 1 week, treated with Paracetamol and other unrecalled medications. The patient was then brought to Metro Manila to seek consult. Two years prior to admission, the patient was placed in a center for street children due to financial difficulties. At this time the frequency of the fevers decreased, but the patient complained of occasional chest pain and non-productive cough. One year prior to the admission, the patient again had moderate grade fever, pallor and easy fatigability. He was then brought to a private hospital wherein laboratory exams showed anemia on CBC, with normal stool and urinary examinations. A chest radiograph showed enlarged right atrium and ventricle with signs of pulmonary congestion. The patient was assessed to have congenital heart disease. The patient was transfused with packed RBC, and was sent home on iron supplements, Digoxin, INH, Rifampicin and Pen VK. He was advised to consult at a tertiary government hospital for further management and work-up.

Eight months prior to admission, the patient consulted at the Out Patient Department of the tertiary government and physical examination revealed normal vital signs, with sallow skin, neck vein engorgement, with distinct heart sounds, gr. 3 holosystolic murmur at the left parasternal border, with liver edge 3 to 4 cm below the right coastal margin, and marked splenomegaly. An electrocardiogram showed tachycardia, regular rhythm, right axis deviation and right bundle branch block. Accentuation of pulmonary vascular markings and prominent right atrium and ventricle was seen on chest x-ray. A 2-d echocardiogram showed right atrium enlargement, right ventricular enlargement, dilated main pulmonary artery, mild mitral stenosis, and moderate to severe tricuspid regurgitation. The patient was assessed to have rheumatic heart disease and pulmonary tuberculosis. The patient was started on benzathine penicillin, which was to be given every 21 days, INH and Rifampicin were continued. On subsequent follow-up a hemogram stilled showed anemia and eosinophilia (hemoglobin = 84, eosinophils = 30) with an elevated sedimentation rate of 70. The ASO was normal, but the C-reactive protein levels was 12. The patient was lost to follow-up but was subsequently admitted on 2 other occasions at another government hospital for fever and chills. At both admissions the patient was assessed to have typhoid fever and was treated accordingly.

Due to another recurrence of fever, chills, productive cough and chest pain, the patient was subsequently admitted. At this time, the patient had a temperature of 39°C, with distended neck veins, a grade 3/6 holosystolic murmur at the left parasternal border, with hepatosplenomegaly. Chest x-ray showed right axis deviation and right ventricular hypertrophy. A repeat echocardiogram showed severe pulmonary hypertension. What is your diagnosis?

*For **DENOUEMENT** see page 36...*

Keywords: *pulmonary schistosomiasis*

We have a 13-year old male from Samar manifesting symptoms of heart disease (right-sided congestion and cardiomegaly, chest pain). Congenital heart disease is not likely because of the patient's age, he is already 13 years old and he did not exhibit any symptoms until he was 10 years old. Rheumatic fever eventually developing into rheumatic heart disease is a consideration due to the patient's recurrent episodes of fever. But strictly speaking, the patient did not fulfill the other elements of the Jones' criteria (arthritis, erythema marginatum, chorea). Also, it is more common for a patient with rheumatic heart disease to develop left ventricular hypertrophy because the most common lesions of RHD affect the mitral valve.

The patient has lived in Samar until he was 11 years old when he was brought to Manila and also presented with eosinophilia. One would strongly suspect that his clinical signs and symptoms may be due to Schistosomiasis. During his last admission, this was also suspected thus the following laboratory examinations were performed:

CBC Hb: 60 WBC = 5.7, Seg = 39, Eo = 36, Platelet = 166
Peripheral blood smear = hyhpo++, poikilo = +, ESR = 32
ECG = Right axis deviation, right ventricular hypertrophy, regular sinus rhythm
Kato-Katz = (+)ascaris ova, (+)trichuris ova, (+)hookworm ova
COPT= (+)
Rectal imprint = for Schistosoma eggs
Chest CT scan = fibroid densities in the posterior segment of the right upper lobe, prominent pulmonary artery

Schistosomiasis (bilharziasis) is a chronic infection of the circulatory system caused by several species of blood flukes. In the Philippines, along with other Southeast Asian countries, China and Japan, it is *Schistosoma japonicum* which is dominant. In the Philippines, it is endemic in 24 provinces with 5.1 million people at risk affecting mostly farmers and their families.[1]

The infection is acquired by exposure to water containing cercariae released by infected snails which are the intermediate host for these trematodes. There are three clinical syndromes associated with schistosomiasis: 1. Cercarial penetration – into unbroken skin which sometimes cause intense itching. Pruritic papular lesion or Swimmer's itch may be seen. 2. Oviposition (after 2-3 weeks) – called Katayama fever wherein the most common findings are fever, diarrhea, arthralgias, anorexia, malaise, hepatosplenomegaly and is associated with leukocytosis and eosinophilia. The symptoms may gradually abate without therapy usually within 3-4 months after exposure. 3. Chronic schistosomiasis – due to the deposition of eggs leading to inflammation of the intestines, fibrosis of the liver and portal hypertension. The clinical manifestations shall depend in the organs involved which may include the intestines, liver, spleen, heart, lungs or brain.

Pulmonary manifestations of schistosomiasis may also present as two forms. In the acute phase, the schistosomulae pass through the lungs and there may be an acute febrile illness with eosonophilia and diffuse pulmonary nodules on chest x-ray.[2] The illness and radiographic picture resemble miliary tuberculosis or influenza pneumonia. Clinical and radiographic resolution of this phase is usually rapid and complete even without specific therapy. In chronic schistosomiasis, embolization of ova to small arteries in the lungs results in multiple pseudotubercles. The egg deposition causes a granulomatous reaction around them, thus obliterating these vessels and subsequently cause pulmonary hypertension. About 1/4 of the patients with hepatosplenic schistosomiasis with portal hypertension may have clinical evidence of pulmonary disease but less than 5% would progress to true cor pulmonale, which was seen in this patient. Symptoms include dyspnea, and when cor pulmonale has developed the patient may complain of chest pain.

The most common radiographic finding is a diffuse fine nodular pattern similar to that seen with miliary tuberculosis. Other findings would be those of focal opacities of varying size which may appear singly or in groups resembling a honeycomb, a spider web or the pulmonary vasculature may be accentuated. Either of these pictures may predominate without correlation to the severity of cardiac manifestations. In a pathologic review of pulmonary schistosomiasis, the most consistent findings in patients with cor pulmonale was diffuse interalveolar septal thickening. Other findings included obliterative endarteritis and medial hyperplasia.[3]

A presumptive diagnosis of this can be made based on the geographical history in a patient with hepatosplenomegaly. In chronic cases, no viable worms may be seen in sputum examinations because the average life span of the schistosomes is 4-5 years. Demonstration of the ovas may be done by rectal biopsy. Serologic tests for schistosoma antibodies are available and this includes the circumoval preciptin test. This test has the advantage of having a high sensitivity (94-97%) and high specificity but unfortunately it does give any indication of the intensity of infection and it can not distinguish past and present infections.[4]

Medical treatment of chronic pulmonary schistosomiasis is unsatisfactory because granulomatous changes are already irreversible. Thus treatment is limited to prevent further damage caused by additional emboli of the eggs. Praziquantel is the drug of choice[5] and it is administered at 20 mg/kg/dose for three doses at 4-6 hour intervals. It is well-tolerated and well suited for mass treatment control programs. Vaccine models employing crude schistosome extracts, purified antigens or anti-idiotypes are currently being investigated.

Prologue: The patient was treated with Praziquantel and on subsequent follow-up still had dyspnea only during vigorous activity. Repeat blood counts showed a decrease of the eosinophilia to normal levels. Fever episodes did not recur.

REFERENCES

1. Department of Health, Philippine Schistomiasis Control Service Annual Report, 1995.
2. Barett Connor E, 1982. Parasitic Pulmonary Disease. Am Rev Respir Dis 126: 558-563.
3. Ostrea EM, Marcelo FB. 1965. Relationship between pulmonary schistosoma japonicum and cor pulmonale. Acta Medica Philippina Vol. 2, Sept.-Dec.
4. Garcia EG, 1981. Attempts to standardize the COPT for schistosomiasis japonica. Southeast Asian J Trop Med. 12:3.
5. WHO Drug Information Vol 11, No. 2, 1997.
6. Arcy PF, Harron DWG. 1983. Shistosomiasis. Pharmacy International. January, pp 16-20.

"Warmth, water and poverty are the basic ingredients of tropical life: add a few snails and a dash of feces or urine, and you have schistosomiasis; the recipe is simple and will serve any number."

E. Chernin (1978) in Tropical Medicine: From Romance to Reality, Academic Press, London