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Case Report

Schistosomiasis-Associated Pulmonary Hypertension cured with Praziquantel Therapy

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ABSTRACT

This is a case report of a 52-year-old female who immigrated from Africa and presented with impending right heart failure, characterized by shortness of breath and leg swelling for the past two months. In the emergency room (ER), a CT chest with a pulmonary embolism protocol was performed, which was negative for clots but revealed findings of cirrhosis, splenomegaly and pulmonary artery dilation. Further workup demonstrated iron deficiency anemia and severe pulmonary hypertension, confirmed via right heart catheterization. Treatment was initiated with diuretics and pulmonary vasodilators, eventually requiring triple drug therapy that included intravenous epoprostenol. Given her travel history, schistosomiasis was suspected and subsequently confirmed with antibody testing, indicating chronic schistosomiasis with hepatic involvement. She was treated with praziquantel but experienced a transient worsening of symptoms and hypoxia due to hypersensitivity pneumonitis likely form dying parasites. The patient responded well to a moderate dose of prednisone and was eventually weaned off oxygen and epoprostenol. A three-month follow-up right heart catheterization revealed near normalization of pulmonary pressures with significantly improved functional capacity. Six months later, she was successfully weaned off all pulmonary vasodilators.

Keywords: Impending right heart; Epoprostenol; Schistosomiasis; Pulmonary vasodilators

Introduction

Schistosomiasis is a parasitic infection caused by the genus Schistosoma, commonly endemic to tropical and subtropical regions. Among its severe complications is pulmonary hypertension (PH), which can lead to right heart failure and is typically associated with the hepato-splenic form of chronic schistosomiasis. The diagnosis is often challenging due to overlapping clinical features with conditions such as cirrhosis and iron deficiency anemia. Treatment typically involves a combination of antiparasitic agents like praziquantel and supportive therapy, including pulmonary vasodilators. This case highlights the clinical course and successful management of a patient with schistosomiasis-associated pulmonary hypertension (Sch-PAH), ultimately achieving resolution of pulmonary hypertension.

Case Presentation

A 52-year-old non-alcoholic female with a medical history of hypertension and type 2 diabetes presented with a two-month history of chronic pain, generalized weakness, difficulty sleeping, chest discomfort, progressive exertional shortness of breath and leg swelling. She reported a gradual decline in her functional capacity over this period, eventually becoming unable to perform activities of daily living. She also noted subjective evening fevers, grogginess and at least two near-

syncopal episodes prior to presentation. She denied hemoptysis, hematemesis, weight loss, convulsions, wheezing or changes in bowel or bladder habits.

The patient had immigrated to the United States two months earlier and had not yet established care with a primary physician. Upon arrival at the ER, her vital signs were as follows: blood pressure (BP) 90/65 mmHg, heart rate (HR) 112/min, respiratory rate (RR) 24/min and oxygen saturation (SpO2) 84%. Physical examination revealed a distended abdomen, dilated superficial veins, jugular venous distention (JVD) and pedal edema. Auscultation findings were unremarkable. Oxygen supplementation was initiated at 4 L/min. Troponin levels were negative for acute coronary syndrome, but an electrocardiogram (ECG) showed multiple premature ventricular contractions (PVCs) with a right bundle branch block (RBBB) pattern. A CT chest to rule out pulmonary embolism (PE) was negative for clots but demonstrated cirrhotic liver changes, splenomegaly and pulmonary artery dilation. See **(Figures 1,2 and 3)**.

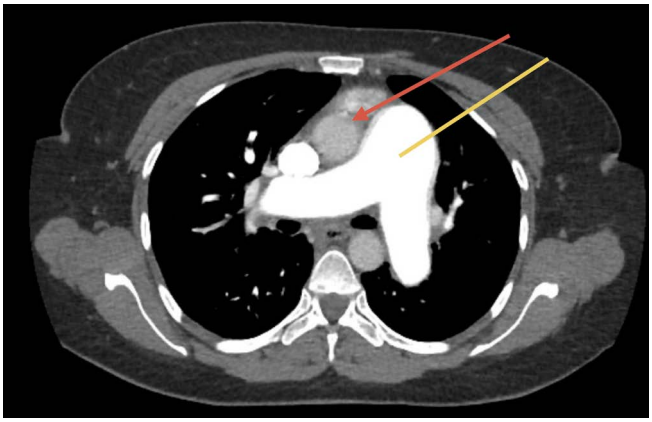

Figure 1: Ct Chest axial view, mediastinal window demonstrating enlarged pulmonary artery (yellow line) compared to aorta (red arrow) suggestive of pulmonary hypertension

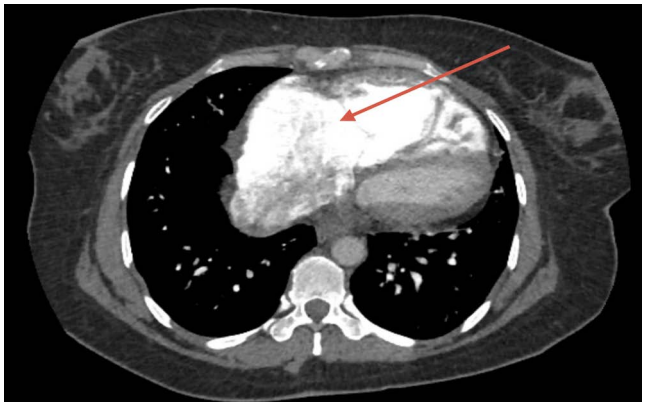

Figure 2: CT Chest Axial view, mediastinal window demonstrating dilated right ventricle (red arrow) suggestive of pulmonary hypertension

Laboratory results revealed a hemoglobin level of 7.6 g/dL, consistent with iron deficiency anemia; lactate level of 4.2 mmol/L; serum creatinine of 1.5 mg/dL; and brain natriuretic peptide (BNP) of 400 pg/mL. She was admitted to the cardiac intensive care unit (ICU) with a diagnosis of cardiogenic shock. Transthoracic echocardiography showed an estimated pulmonary artery systolic pressure (PASP) of 81 mmHg and severely reduced right ventricular systolic function, with a tricuspid annular plane systolic excursion (TAPSE) of 1.1 cm. See **(Figures 4 and 5)**.

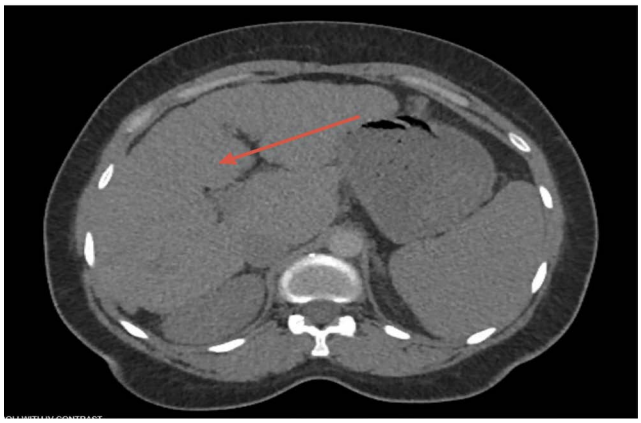

Figure 3: Ct Chest at the level of liver, abdominal window showing nodular configuration of Liver (red arrow) suggesting fibrosis

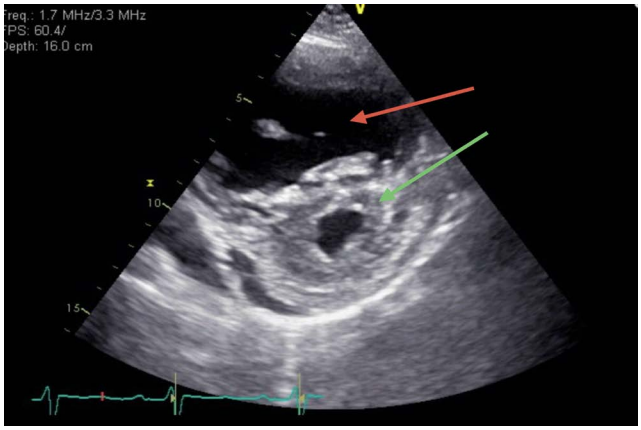

Figure 4: Echocardiogram: Parasternal short axis view demonstrating enlarged right ventricle (red arrow) compared to left ventricle (green arrow) suggestive of pulmonary hypertension

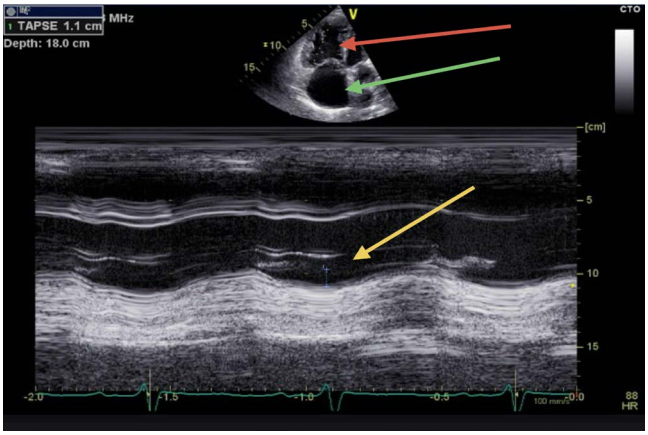

Figure 5: Echocardiogram Top figure: Apical four chamber view revealing enlargement of right atrium (green arrow) and right ventricle (red arrow). Bottom figure: M mode of the Tricuspid annulus. TAPSE (tricuspid annular plane systolic excursion) was noted to be 1.1 cm denoting severe decrease in right ventricular function. Normal TAPSE is > 1.7 cm Iron supplementation and gentle diuretics were administered.

A ventilation-perfusion (VQ) scan showed no mismatch. See **(Figure 6)**.

Right heart catheterization confirmed severe pulmonary arterial hypertension (PAH), with findings including a mean pulmonary artery pressure (mPAP) of 58 mmHg, pulmonary vascular resistance (PVR) of 16 Wood units (WU) and cardiac

index (CI) of 1.95 L/min/m² with central venous oxygen saturation of 35%. Vaso reactive testing showed a significant drop in systemic blood pressure and cardiac output. Key catheterization data:

- **Baseline:** RA 17 mmHg, PA 96/37/58 mmHg, PCW 4 mmHg, PVR 16 WU, CI 1.95 L/min/m²
- **Drug study:** RA 17 mmHg, PA 66/26/39 mmHg, PCW 4 mmHg, PVR 11 WU, CI 1.7 L/min/m²

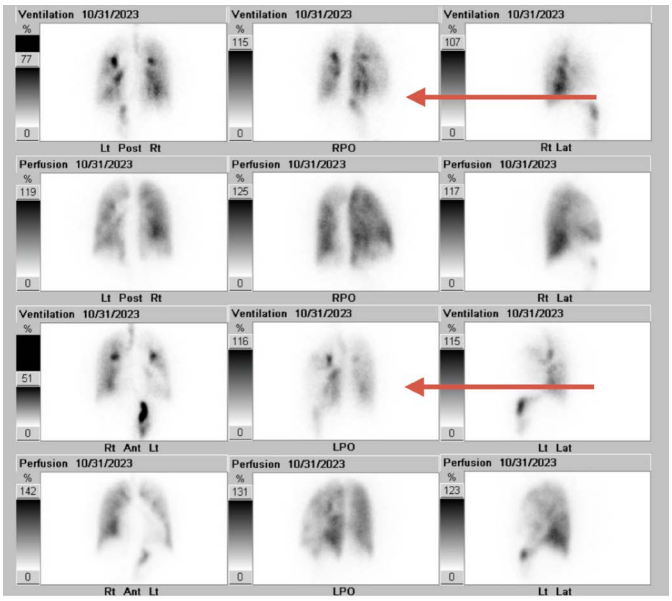

Figure 6: VQ Scan demonstrated mildly heterogenous perfusion and ventilatory radio-tracer distribution throughout both lungs (red arrows). No ventilation perfusion mismatch. Low probability of pulmonary embolism. Th over all pattern is suggestive of pulmonary hypertension

Treatment and Follow-Up: The patient was started on intravenous epoprostenol at 2 ng/kg/min, titrated to 10 ng/kg/min, with dose adjustments limited by side effects such as flushing. She was kept nil per os (NPO) pending an endoscopic evaluation. On day three, a gastroenterology consultation was obtained for cirrhosis, iron deficiency anemia and positive fecal occult blood tests.

Esophagogastroduodenoscopy (EGD) and colonoscopy revealed grade 1 esophageal varices, portal hypertensive gastropathy and multiple colonic ulcers, particularly in the sigmoid and rectum **(Figures 7 and 8)**.

Given her recent immigration from Africa, schistosomiasis was suspected. Testing confirmed chronic schistosomiasis with positive Schistosoma antibody and radiological evidence of cirrhosis, although stool ova and parasites were negative. Sildenafil was initiated on day four and uptitrated to 20 mg TID over 48 hours. On day six, macitentan 10 mg daily was added for triple therapy alongside epoprostenol and sildenafil, leading to significant improvements in functional status and hemodynamics. She was eventually weaned off oxygen. Praziquantel was administered in two doses, one week apart, as a part of anti-parasitic therapy. However, 24 hours after the first dose, the patient experienced worsening hypoxia and shortness of breath needing 4-6 l/min of Oxygen supplementation.

High-resolution CT chest revealed diffuse ground-glass opacities with mosaic attenuation and pulmonary nodules, suggestive of hypersensitivity pneumonitis. A short course of prednisone (40 mg daily for seven days) resolved her hypoxia within 48 hours. See **(Figure 9)**.

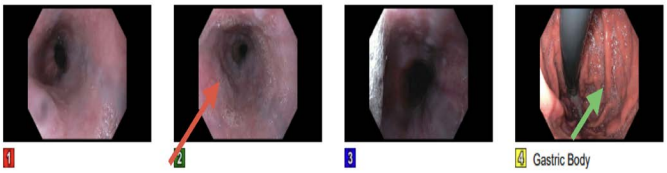

Figure 7: Upper GI endoscopy showed varices (red arrow) in the lower third of esophagus and moderate portal hypertensive gastropathy in the gastric body (green arrow). Duodenal biopsy revealed mildly increased intraepithelial lymphocytes and intact villous architecture.

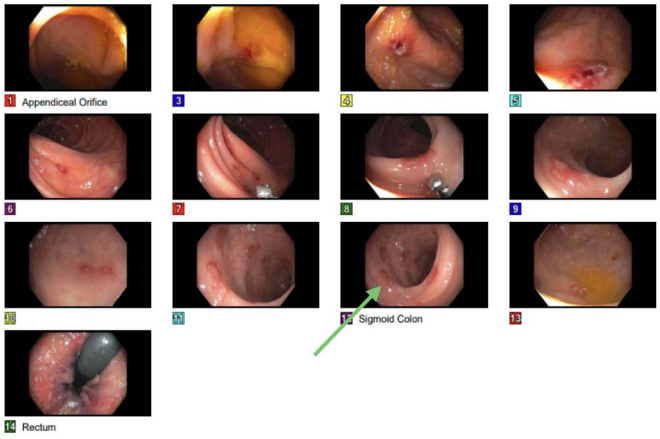

Figure 8: Colonoscopy showed multiple 2-6 mm ulcers in the entire colon with highest concentration in the sigmoid (green arrow) and rectum. Biopsy revealed active colitis with necro-inflammatory ulcer debris. There were no features of chronicity, granulomata or dysplasia. CMV immunostaining was negative. No viral inclusions noted. Serology was negative for Celiac disease and Inflammatory Bowel Disease (IBD).

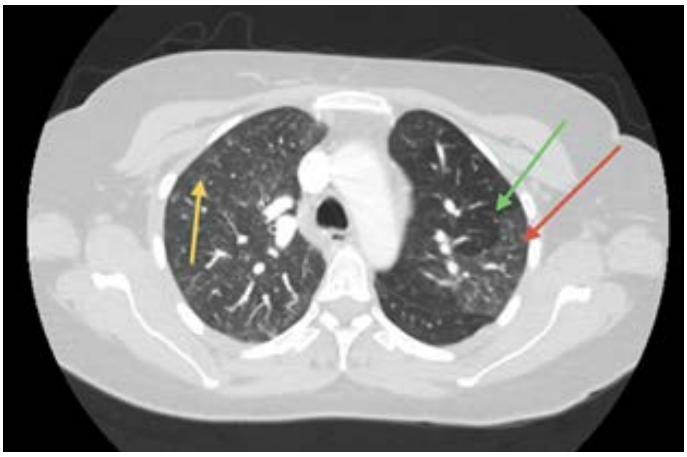

Figure 9: Hi-resolution T Chest, lung window at the level of aortic arch demonstrating diffuse ground glass opacities (red arrow) with mosaic attenuation (green arrow) and nodularity (yellow arrow) after receiving anti-parasitic therapy. Responded very well to steroids

By day ten, epoprostenol was gradually weaned off due to side effects of flushing and dizziness. The patient was maintained on sildenafil and macitentan. Follow-up right heart catheterization showed significant improvement, with a PA pressure of 29/48 mmHg, PVR of 6 WU and CI of 2.48 L/min/m² and hence prostaglandin therapy was not -re-initiated as there was good

response to dual therapy associated with rapid improvement in her walk distance. She was discharged on sildenafil 20 mg TID and macitentan 10 mg daily, with a six-minute walk distance (6MWD) of 200 meters.

At the three-month follow-up, her 6MWD had increased to 500 meters and BNP levels had normalized to 45 pg/mL. Repeat echocardiography revealed normalization of pulmonary pressures. A repeat right heart catheterization showed further improvements with a PA pressure of 32/16/21 mmHg, a PCWP of 8 and a PVR of 2.3 WU. As she was slightly hypotensive and complained of mild orthostatic dizziness, macitentan was discontinued and Sildenafil was continued. Six months later, her functional capacity and BNP levels continued to improve and she remained asymptomatic. Sildenafil was gradually tapered off and she remained stable. The patient remained asymptomatic and continued to perform all activities without limitations. A repeat echocardiogram in 1 month after the 6 months follow up confirmed complete normalization of pulmonary pressures.

Discussion

Schistosomiasis, caused by parasitic trematodes, remains a significant health concern in endemic regions such as Africa and Asia, affecting approximately 200 million individuals globally[1]. The chronic hepato-splenic form of the disease, a known risk factor for pulmonary arterial hypertension (PAH), accounts for severe morbidity and mortality[2]. This case presents a striking example of schistosomiasis-associated pulmonary arterial hypertension (Sch-PAH), a condition classified under World Health Organization (WHO) Group 1 PAH (1.4.5)[3]. The patient, a 52-year-old immigrant from Africa, presented with a constellation of symptoms, including exertional dyspnea, lower extremity edema, syncope and iron deficiency anemia. These features underscored the diagnostic challenge of Sch-PAH, often overshadowed by clinical features of portal hypertension, presumed liver cirrhosis and co-existing anemia.

The only clue to the diagnosis was the patient's residence in an endemic region. Clinical findings included progressive exertional dyspnea, lower extremity edema, jugular venous distention (JVD) and a loud S2, which is highly suggestive of pulmonary hypertension and right ventricular dysfunction[4]. Initial evaluation revealed evidence of right ventricular dysfunction (TAPSE < 1.7 cm) with an elevated pulmonary artery systolic pressure of 81 mmHg on echocardiography, alongside severe pulmonary hypertension confirmed via right heart catheterization (mean pulmonary artery pressure of 58 mmHg and pulmonary vascular resistance of 16 Wood units).

It is crucial to distinguish between pre-capillary and post-capillary forms of pulmonary arterial hypertension, as the management differs significantly. Pulmonary vasodilators are indicated in the former but contraindicated in the latter. Right heart catheterization helps make this distinction. Pre-capillary pulmonary hypertension is defined by a mean pulmonary artery pressure (mPAP) > 20 mmHg at rest, pulmonary vascular resistance (PVR) > 2 Wood units and pulmonary capillary wedge pressure (PCWP) ≤ 15 mmHg. In contrast, post-capillary pulmonary hypertension, often associated with left heart disease, is characterized by a PCWP > 15 mmHg [3]. The patient's immigration history and diagnostic imaging, which revealed cirrhosis and portal hypertension, raised suspicion for chronic schistosomiasis. Another important clue is the presence of eosinophilia in a patient with pulmonary arterial hypertension, which should prompt consideration of Sch-PAH[2].

The diagnosis of Sch-PAH in this case was supported by positive Schistosoma antibody testing, even though stool ova and parasites were negative-a finding not uncommon in chronic disease due to intermittent egg shedding[5]. This case highlights the importance of integrating travel history and risk factor assessment into the evaluation of pulmonary hypertension, particularly in immigrant populations.

The patient's management demonstrated a dual therapeutic approach targeting both the underlying parasitic infection and pulmonary arterial hypertension. Anti-parasitic therapy with praziquantel was initiated at 60 mg/kg, administered in two doses one week apart, targeting the adult Schistosoma parasites. Notably, the patient developed hypersensitivity pneumonitis after the first dose, presenting as worsening hypoxia and ground-glass opacities with mosaic attenuation on computed tomography (CT). This inflammatory reaction, likely due to antigen release from dying parasites, was successfully managed with a short course of corticosteroids[2].

Pulmonary arterial hypertension was treated with specific vasodilatory therapy. Initial management included intravenous epoprostenol, titrated according to tolerance, followed by the introduction of sildenafil and macitentan as part of a triple therapy protocol, given the high-risk assessment of her pulmonary hypertension. This combination led to marked clinical and hemodynamic improvement, with a reduction in PVR from 16 to 6 Wood units and normalization of pulmonary pressures over three months. The patient's six-minute walk distance improved from 200 to 500 meters, reflecting a significant recovery in functional capacity.

Although many patients with Sch-PAH remain dependent on pulmonary vasodilator therapy indefinitely, this patient demonstrated significant hemodynamic improvement with anti-parasitic therapy[6-9].

This suggests that some cases of Sch-PAH may be reversible. Therefore, patients with positive Schistosoma antibodies and no prior anti-parasitic treatment should be given the opportunity for therapy[8,9].

The patient's clinical course exemplifies the potential for substantial improvement in Sch-PAH with early and targeted interventions. Despite her initial presentation in cardiogenic shock, the combination of anti-parasitic therapy and pulmonary vasodilators resulted in sustained remission. Follow-up evaluations at six months revealed continued functional and hemodynamic improvement, normalization of biomarkers such as BNP (20 pg/mL) and cessation of vasodilator therapy without recurrence of symptoms.

Conclusion

This case underscores the complex interplay between schistosomiasis, cirrhosis and pulmonary hypertension. Early diagnosis and appropriate treatment with praziquantel, combined with pulmonary hypertension-targeted therapy, led to remarkable clinical improvement. Close monitoring and management of treatment-related side effects, such as hypersensitivity pneumonitis, were critical in ensuring a positive outcome. This case also illustrates the potential for complete reversal of Sch-PAH with early treatment of schistosomiasis. The use of the

REVEAL score to guide risk stratification and management was essential for tracking the patient's progress and optimizing her treatment regimen.

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