

A patient from Limpopo province presenting with haematuria: what is the diagnosis?

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Keywords: schistosomiasis, computed tomography

We present a quiz-case of a 28-year-old female patient, from Limpopo province, who presented with haematuria, hepatosplenomegaly and caput medusae. Computed tomography (CT) imaging findings are provided. The diagnosis is provided in the discussion below.

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South Afr J Epidemiol Infect 2012;27(2):84-86

Case study

A 28-year-old female patient, from Limpopo province, presented with haematuria. On clinical examination, her vital signs were normal. She had signs of pulmonary hypertension, with a loud P2. No murmurs were present. Respiratory and neurological evaluation was normal. She had hepatosplenomegaly and caput medusae. There was no history of malena stool or haematemesis. No significant lymph nodes were present. Biochemical evaluation, that was

pertinent, included microcytic anaemia and elevated D Dimers of 1.4. Urine microscopy results are withheld at this point, for the purpose of this quiz.

Contrast-enhanced computed tomography (CT) pulmonary angiography was performed as part of the patient's investigation. Selected images (Figures 1-8) showing pulmonary and cardiovascular changes that are relevant to the diagnosis are presented below.



Figure 1: Axial contrast-enhanced computed tomography, demonstrating dilatation of the central pulmonary arteries, with a mural thrombus (arrow) in the right main pulmonary artery

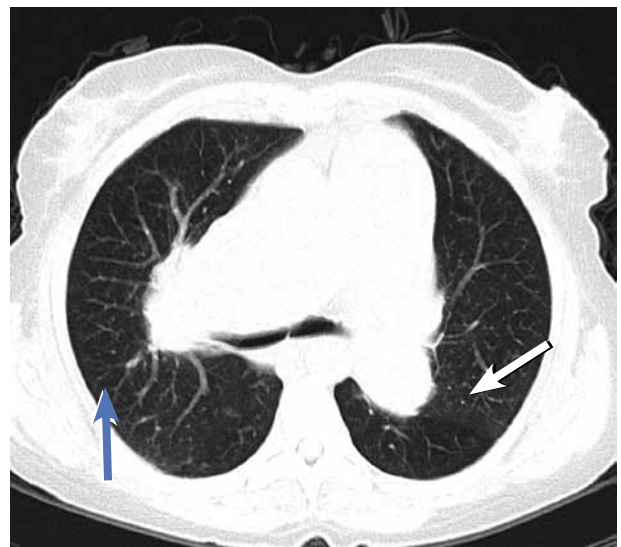


Figure 2: Axial non-enhanced computed tomography, in a lung window, which shows nodules (white arrow) and interstitial thickening in the lungs (blue arrow)

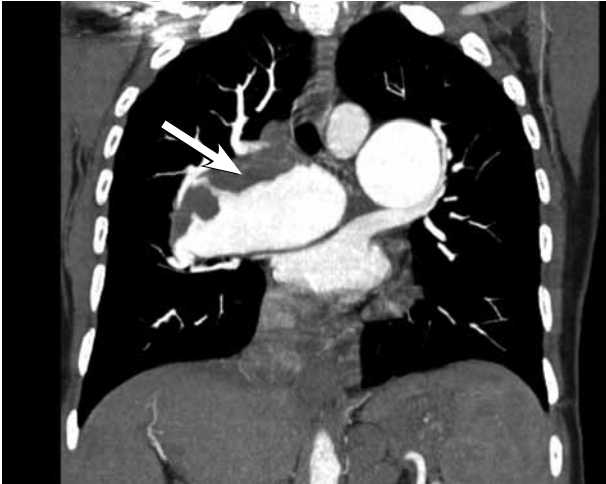


Figure 3: Coronal view which demonstrates a dilated right main pulmonary artery, with a mural thrombus (arrow)

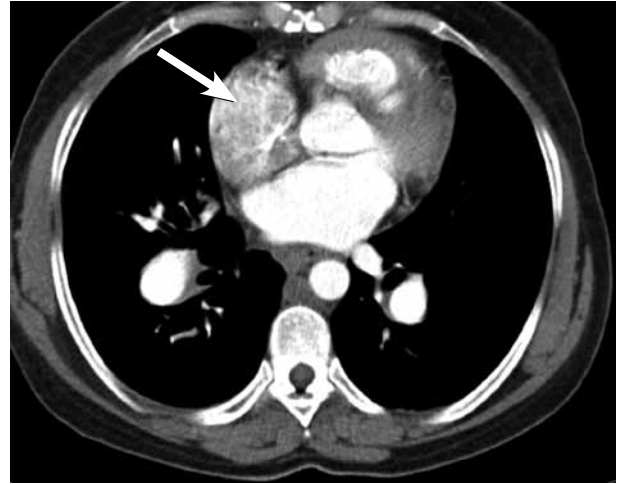


Figure 4: Axial contrast-enhanced computed tomography, showing dilatation of the right atrium



Figure 5: Contrast-enhanced computed tomography demonstrating enlargement of the right ventricle



Figure 6: Axial contrast-enhanced computed tomography, which shows dilatation of the azygos vein (due to portal hypertension) (arrow)

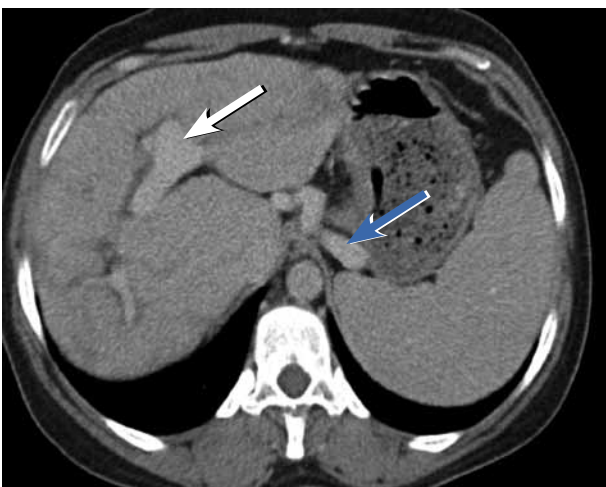


Figure 7: Axial contrast enhanced CT of the abdomen which shows an enlarged portal vein (white arrow) and splenic vein (blue arrow) in keeping with portal hypertension

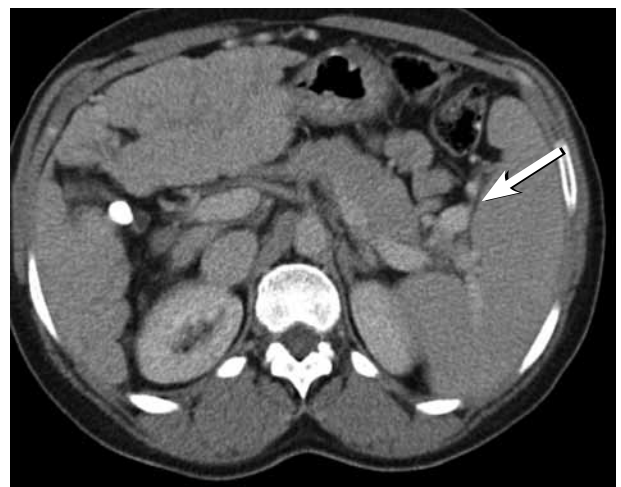


Figure 8: Axial contrast-enhanced computed tomography of the abdomen, which demonstrates varices in the splenic hilum (arrow)

Diagnosis

The diagnosis is cardiopulmonary schistosomiasis on computed tomography.

Schistosomiasis is a common parasitic infection, with over 200 million people infected worldwide.¹ The disease can present acutely, with cutaneous inflammation at the site of parasitic penetration, followed by a sub-acute hypersensitivity immune-mediated response, as the parasite passes through the lungs.¹ This can present with influenza-like symptoms, including a cough.² Chronic infection is characterised by granulomatous inflammation around ova deposition in the tissues.

The disease is characterised by portal hypertension, with peri-portal hepatic fibrosis. Other findings include gradually worsening hepatosplenomegaly, dyspnea, a cough, chest pain, severe hypoxaemia, cyanosis, and digital clubbing.

The organism that is mainly responsible for this condition is *Schistosoma mansoni*, with *S. japonicum* and *S. haematobium* less commonly involved. *S. mansoni* is endemic in the Middle East, Africa, and the Atlantic coast of South America and the Caribbean.³

The female parasite discharges eggs into vesicular venules. These eggs then erode the bladder mucosa, and are excreted with the urine and the faeces. In fresh water, the eggs hatch into larval miracidia. The larvae invade snails (the intermediate host) of genus *Bulinus*, *Biomphalaria* and *Oncomelania*. The resulting daughter sporocysts develop into cercariae, and pass into the surrounding body of water. The cercariae then penetrate human skin (usually through the feet) and pass into the lymphatics.

S. mansoni settles in the portal venous system, and causes pre-sinusoidal portal hypertension and porto-systemic shunts.⁴ Portal hypertension facilitates the shunting of ova to the pulmonary arteries.¹ Eggs lodge in pulmonary muscular arteries and arterioles, which are 50-150 µm in diameter. The trapped eggs are antigenic, and incite an obliterative

endarteritis, due to delayed host hypersensitivity. Therefore, cardiopulmonary schistosomiasis is regarded as a form of parasitic embolism, and patients with more than five years of continuous ova excretion are at risk.

Extensive remodelling of the pulmonary vasculature occurs with the development of intra and perivascular granulomas, intimal hyperplasia, medial hypertrophy, concentric collagen deposition, fibrosis of vessel walls, and localised alveolitis, with eosinophilic infiltration and pulmonary infarction. Crosby et al also found that in mice infected with a higher egg burden, perivascular inflammation and plexiform-like lesions developed in the pulmonary vasculature.⁵

On computed tomography, patients present with nodules, interstitial thickening (Figure 1); dilatation of the right atrium, right ventricle and central pulmonary arteries (Figures 2-5); as well as the azygous vein from portal hypertension (Figure 6); and features of portal hypertension, including oesophageal varices, hepatosplenomegaly and splenic vein varices (Figures 7 and 8).

Other systemic manifestations that may be present include bladder-wall calcification, distal ureteral calcification, and honeycombed calcification of the seminal vesicles.

Conclusion

Computed tomography is a useful adjunct in the evaluation of cardio-pulmonary, and other, systemic manifestations of Schistosomiasis.

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