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A 37-Year-Old Woman from Malawi With Haematemesis

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Clinical Presentation

History

A 37-year-old woman from the Lower Shire Valley in southern Malawi is referred from a clinic on one of the local sugar plantations to the district hospital. She has vomited blood three times over the past 24 hours. The blood is bright red in colour. There is no epigastric pain and no previous history of vomiting. There is no history of fever or abnormal bleeding and her stool has been normal in colour. Before the onset of symptoms, she was fine. She has not taken any regular painkillers and does not drink any alcohol.

Her past medical history is unremarkable. She lives and works on a large sugar plantation in the area. She is married with three children, all are well. An HIV test done 3 months previously was negative.

Clinical Findings

37-year-old woman who is slim but not wasted. Conjunctivae are slightly pale, but there are no subconjunctival effusions and she is not jaundiced. Her blood pressure is 90/60 mmHg, pulse 110 bpm, respiratory rate 28 breath cycles per minute, and she is afebrile.

On examination of the abdomen there is no abdominal distension and no tenderness. The spleen is palpable at 10 cm below the left costal margin. The liver is slightly enlarged but there are no stigmata of chronic liver disease. There is no shifting dullness and no peripheral oedema. Her lymph nodes are not enlarged. The rest of the physical examination is normal.

Laboratory Results

Her laboratory results on admission are shown in Table 57.1.

Questions

1. What is the most likely cause of her haematemesis?
2. What further investigations would you like to do to establish the diagnosis?

Discussion

A Malawian woman presents with a first episode of haematemesis. She has neither taken NSAIDs nor alcohol. On examination she is afebrile, slightly pale, shocked and has an enlarged spleen. Her abdomen is non-tender. Her full blood count shows pancytopenia with normocytic anaemia.

Answer to Question 1

What is the Most Likely Cause of Her Haematemesis?

Splenomegaly and pancytopenia point towards the presence of portal hypertension and she is most likely to bleed from gastro-oesophageal varices. In one series from Malawi, the presence of splenomegaly in patients with upper gastrointestinal bleeding was the single most specific clinical criterion to distinguish between a variceal bleed and a haemorrhage of other origin.

Furthermore, portal hypertension is the most common cause of upper gastrointestinal bleeding in parts of sub-Saharan Africa, accounting for more than 50% of bleeds in some series. The reason for this remains only partly understood. The prevalence of chronic viral hepatitis or alcohol abuse is not higher in the affected regions than elsewhere in the tropical world.

However, Malawi and other countries in the region are highly endemic for schistosomiasis. Virtually all water bodies

TABLE 57.1 Laboratory Results on Admission

Parameter	Patient	Reference Range
WBC ($\times 10^9$/L)	2.8	4–10
Haemoglobin (g/dL)	8.3	12–14
MCV (fL)	88	80–99
Platelets ($\times 10^9$/L)	130	150–400

in the country are infested with both *Schistosoma mansoni* and *S. haematobium* and it is likely that a high prevalence of hepatosplenic schistosomiasis with periportal fibrosis may explain why portal hypertension is so common in the region.

Little is known about causes of liver cirrhosis other than hepatitis B and C and their contribution to the burden of disease (e.g. autoimmune hepatitis, primary biliary cirrhosis, primary sclerosing cholangitis, haemochromatosis or Wilson's disease).

Other aetiologies of gastrointestinal bleeding, such as peptic ulcer disease, erosive gastritis or a bleeding tumour are less likely and would not explain her splenomegaly. Notably though, in sub-Saharan Africa oesophageal cancer is strikingly common in young adults in their third and fourth decade of life. Risk factors remain poorly understood. Progressive dysphagia rather than haematemesis is usually the presenting symptom.

Patients with visceral leishmaniasis (VL) can present with splenomegaly and pancytopenia. However, in VL there is usually a history of fever, and the patient's platelet count is only slightly diminished, which cannot explain her bleed. VL, furthermore, is uncommon in Southern Africa, even though few sporadic cases have been described from the region.

Answer to Question 2

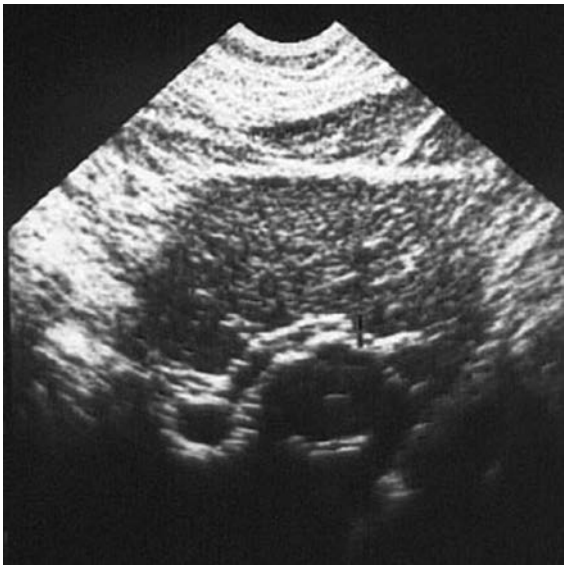
What Additional Investigations Would You Like to Do to Establish the Diagnosis?

In a resource-constrained setting, very limited options may be available to establish more than just a syndromic diagnosis.

The patient should be taken for gastroduodenoscopy without delay to detect and treat the source of bleeding. In case of oesophageal varices, an experienced ultrasonographer with a reasonable ultrasound machine would be able to distinguish between liver cirrhosis and periportal 'pipestem' fibrosis as seen in hepatic schistosomiasis (Fig. 57.1). Liver biopsy would be the most helpful tool to distinguish between cirrhosis and periportal fibrosis in case of schistosomiasis. It is usually unavailable, and it is risky in an environment where postinterventional monitoring is poor, and possible complications such as intra-abdominal bleeds are likely to go undetected.

Other investigations to diagnose schistosomiasis will lack sensitivity and/or specificity in the endemic setting. The demonstration of *Schistosoma* eggs in the stool is challenging in advanced disease and several samples would have to be examined which is commonly not feasible. The circulating cathodic antigen test (CCA) is a urine rapid diagnostic test based on antigens regurgitated by adult flukes. Its sensitivity decreases when the worm burden is low, as may be the case in advanced disease; but it is highly specific and – other than serology – the CCA test is able to distinguish between active and past infection.

Hepatitis B or C serologies should be done; other investigations for chronic liver disease are usually unavailable.



• **Fig. 57.1** Ultrasound of the liver showing pipestem fibrosis (Courtesy Prof. Joachim Richter).

The Case Continued…

The patient received IV fluids and was taken to the nearest central hospital for endoscopy. The presence of oesophageal varices was confirmed and banding was done. There was no other source of bleeding. Hepatitis B and C serologies were negative. One stool sample for *S. mansoni* ova was negative. However, on ultrasound of the liver a pattern typical of pipestem fibrosis was described. The patient received a single dose of praziquantel 40 mg/kg. She was discharged home.

SUMMARY BOX

Hepatosplenic Schistosomiasis

Hepatosplenic schistosomiasis is a complication of advanced infection with *Schistosoma mansoni, S. japonicum* or *S. mekongi.*

Only about 10% of people chronically infected with schistosomiasis develop late-stage disease. Risk factors for disease progression remain poorly understood. Apart from intensity and duration of infection, host genetic factors such as ethnic background and IFN-gamma polymorphism, variable degrees of semi-immunity and parasite strain differences may play a role.

Chronic infection with liver-pathogenic *Schistosoma* species results in periportal fibrosis and portal venous hypertension. One exception is *S. intercalatum,* which occurs focally in Central and West Africa and causes granulomatous inflammation of the liver without portal hypertension.

Patients may present with symptoms of hypersplenism, such as abdominal discomfort and fatigue secondary to progressive anaemia. Ascites is uncommon because of the preserved hepatocellular function but may occur in advanced disease or in coexisting liver cirrhosis.

The classical clinical signs of liver cirrhosis (e.g. gynaecomastia, palmar erythema, alterations in distribution of body hair) are absent in schistosomiasis.

A common, primary presenting sign of hepatosplenic schistosomiasis is upper gastrointestinal (GI) bleeding from gastro-oesophageal varices. In some endemic countries in

sub-Saharan Africa half or more of upper GI bleeds are caused by portal hypertension.

The demonstration of *Schistosoma* eggs in the stool can be challenging in advanced disease because the adult flukes may have long died and egg production may have stopped.

Experienced ultrasonographers may be able to detect the typical pattern of 'pipestem fibrosis'. The term refers to the macroscopic aspect of the liver which shows wide bands of fibrosis around portal tracts resembling the stems of a clay pipe. If available, liver biopsy may show ova of *Schistosoma* species along with proliferation of fibrous tissue in and around the portal tract.

Praziquantel may have an effect in treatment of early fibrosis, but has little role to play in advanced disease. Nevertheless, a single dose treatment with praziquantel 40 mg/kg may be given to stop the progression of fibrosis and reduce the worm burden and further egg production.

Treatment of variceal bleeding includes endoscopic sclerotherapy, band-ligation and devascularization surgery. Splenectomy is not recommended in tropical settings, because of the increased susceptibility to malaria and bacterial infections.

Studies on the use of non-selective beta-blockers for prophylaxis of upper GI bleed in hepatic schistosomiasis have yielded controversial results.

A few reports from resource-rich settings seem to indicate that the placement of a transjugular intrahepatic portovenous shunt (TIPS) may be beneficial for the prevention of variceal bleeding in hepatic schistosomiasis.

Further Reading

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