Diagnostic Challenge

There will be an answer; let it bleed

Brian Deady, MD*; Brock Pullen, MD†; David Hodges, MD‡

CASE HISTORY

A 19-year-old African male, a recent immigrant from Sudan, presents to the emergency department after midnight complaining of nausea and two episodes of painless vomiting of bright red blood. The patient describes black stools for 3 days. Through a translator, it is learned that there is a past history of mild asthma; no other medical conditions are described. He has not used acetylsalicylic acid or nonsteroidal antiinflammatory drugs.

On examination, his vital signs are as follows: blood pressure 128/81 mm Hg, pulse 102 beats/min, temperature 36.6°C (97.8°F), and respiration 16 breaths/min. There is no scleral icterus or obvious jaundice. A chest examination reveals good bilateral air entry with normal heart sounds. The abdomen is mildly to moderately distended and nontender. A digital rectal examination reveals black stool, occult blood positive.

Bloodwork is requested, and a large-bore intravenous (IV) drip is initiated. His nausea is treated with IV metoclopramide and diphenhydramine. His initial hemoglobin is 114 g/L, white blood cell count is 17 $\times$ 10^9, and platelets are 249 $\times$ 10^9. His international normalized ratio is 1.6, and liver enzymes are within normal limits.

During the course of emergency department treatment, he has two more moderate episodes of hematemesis. Pantoprazole is initiated, a type and cross-match for blood are requested, and he is moved to a monitored area. He remains hemodynamically stable throughout the duration of the night.

Later in the day, following admission to hospital, computed tomography CT of the abdomen is performed (Figure 1 and Figure 2).

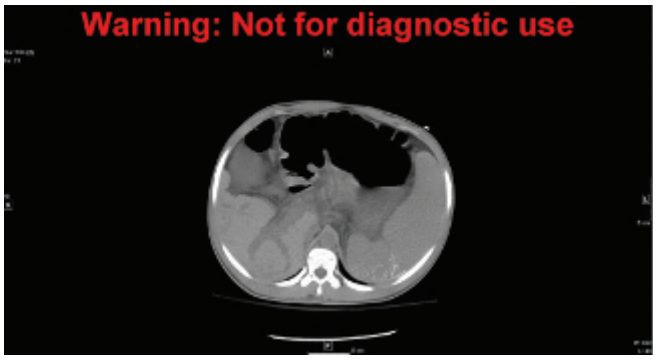

Figure 1. Noncontrast axial computed tomographic scan of the abdomen.

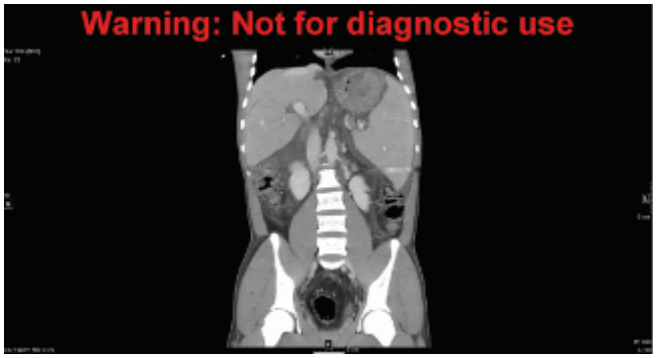

Figure 2. Noncontrast coronal computed tomographic scan of the abdomen.

QUESTION

What is the most likely diagnosis?

a) Esophageal varices
b) Mallory-Weiss tear
c) Peptic ulcer
d) Dieulafoy lesion

For the answer to this challenge, see page 161.

From the Departments of *Emergency Medicine, †Internal Medicine (retired), and ‡Medical Imaging, The Royal Columbian Hospital, New Westminster, BC.

Correspondence to: Dr. Brian Deady, Department of Emergency Medicine, The Royal Columbian Hospital, 330 East Columbia Street, New Westminster, BC V3L 3W7; brian.deady@hotmail.com.

This article has not been peer reviewed.

Diagnostic Challenge

There will be an answer; let it bleed

Brian Deady, MD*; Brock Pullen, MD†; David Hodges, MD‡

ANSWER

The most likely diagnosis is esophageal varices. The patient had presinusoidal portal hypertension secondary to schistosomiasis.

Mallory-Weiss syndrome (MWS) is a relatively common condition with a prevalence of approximately 2 to 10% of patients who present with upper gastrointestinal bleeding.[1–4] A sudden increase in intra-abdominal pressure with retching produces a longitudinal tear in the mucosa and submucosa at or near the gastroesophageal junction. Further vomiting is then associated with hematemesis, although in up to 50% of patients, blood is noted with the first episode of vomiting.[5] Blood loss is usually self-limited, but massive hemorrhage leading to death has been described.[6] Although up to 40 to 70% of patients with active upper gastrointestinal bleeding due to MWS may require blood transfusion, most tears heal spontaneously.[7,8] Endoscopy is the diagnostic procedure of choice.[9]

Peptic ulcer is the most common cause of acute hemorrhage in the upper gastrointestinal tract, accounting for about 50% of cases.[10] About 20% of patients who have bleeding ulcers present with melena, 30% with hematemesis, and 50% with both.[11] As many as 5% of patients with bleeding ulcers present with hematochezia.[11,12]

Dieulafoy lesion is an uncommon cause of major gastrointestinal bleeding,[13] where a large tortuous arteriole erodes through the proximal gastric submucosa and bleeds in the absence of a primary ulcer.[14] Typically, the lesion lies within 6 cm of the gastroesophageal junction on the lesser curve of the stomach, although similar lesions have been described in the intestine, colon, and rectum.[14–16] Awareness of the condition and experience in endoscopy are the mainstay of diagnosis as both diagnosis and treatment can be challenging.[17]

COMMENTARY

Schistosomiasis is a tropical parasitic disease caused by infection with blood flukes known as schistosomes.[18,19] Geographically, schistosomiasis is prevalent in tropical and subtropical regions, particularly sub-Saharan Africa, but also the Middle East, South America, the Caribbean, the Philippines, Thailand, and Indonesia. According to the World Health Organization, 200 million people are infected worldwide, and the infection rates are higher in children than in adults; 200,000 deaths annually are attributable to schistosomiasis.[18–20]

Acute schistosomiasis (AS), or Katayama fever, is seen mostly in travelers after primary infection. Typically, the illness is a result of a systemic hypersensitivity reaction against the migrating schistosomulae, occurring a few weeks to months after a primary infection. The disease starts abruptly with fever, fatigue, myalgia, malaise, nonproductive cough, eosinophilia, and patchy infiltrates on chest radiography.[18,19,21,22]

In chronic schistosomiasis, immunopathologic reactions against schistosome eggs trapped in the tissues lead to inflammatory and obstructive disease in the urinary system (*Schistosoma haematobium*) or intestinal disease, hepatosplenic inflammation, and liver fibrosis (*Schistosoma mansoni*, *Schistosoma japonicum*).[18,19] The eggs secrete proteolytic enzymes that provoke typical eosinophilic inflammatory and granulomatous reactions, which are progressively replaced by fibrotic deposits.[18,19,23]

Chronic schistosomal disease mainly affects individuals with long-standing infections in poor rural areas.

From the Departments of *Emergency Medicine, †Internal Medicine (retired), and ‡Medical Imaging, The Royal Columbian Hospital, New Westminster, BC.

Correspondence to: Dr. Brian Deady, Department of Emergency Medicine, The Royal Columbian Hospital, 330 East Columbia Street, New Westminster, BC V3L 3W7; brian.deady@hotmail.com.

This article has not been peer reviewed.

Hepatic schistosomiasis can lead to two separate clinical scenarios. Inflammatory hepatic schistosomiasis causes hepatosplenomegaly in children and adolescents; disease severity is proportional to the intensity of egg infestation. Chronic hepatic schistosomiasis develops after a longer duration of intense infection. Diffuse collagen deposits in the periportal spaces lead to periportal fibrosis—so-called Symmer pipestem fibrosis—with accompanying splenomegaly and portal hypertension. Interestingly, hepatocellular function commonly remains normal. Morbidity and mortality are a function of ascites and esophageal varices.[18,19] If suspected, ultrasonography is useful in confirming the presence of ascites.[24,25]

The diagnostic standard is microscopic demonstration of eggs in the excreta. Praziquantel, effective against mature worms to stop egg production, is the drug treatment of choice for chronic schistosomiasis. Vaccines are not yet available.[18,19]

Patients with liver disease secondary to schistosomiasis have significant reductions in vitamin K–dependent factors, which can lead to a coagulopathy. Vitamin K administration is advised.[26]

The treatment of AS differs in that the illness is due to an immune response to schistosomulae migration and responds to the combined use of praziquantel and steroids.[27,28] Praziquantel is ineffective against eggs and immature worms[18,19] and may worsen the symptoms of AS if employed without steroids.[29]

CLINICAL COURSE

The next morning in the endoscopy suite, prior to gastroscopy, the patient began vomiting massive amounts of blood and clots; he fell off the stretcher and collapsed to the floor. He remained conscious and had a palpable femoral pulse but was tachycardic, hypotensive, and hypoxic. His hemoglobin had dropped to 65 g/L from the initial value of 114 g/L and was 85 g/L posttransfusion.

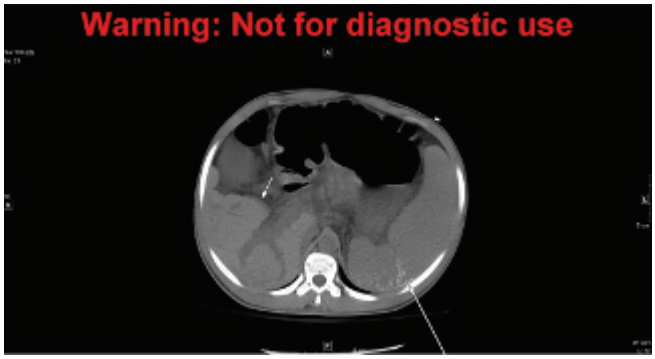

Figure 3. Noncontrast axial computed tomography image through upper abdomen showing small liver with nodular contour, consistent with cirrhosis (*short arrow*), and enlarged spleen containing both fine parenchymal and linear capsular calcifications (*long arrow*).

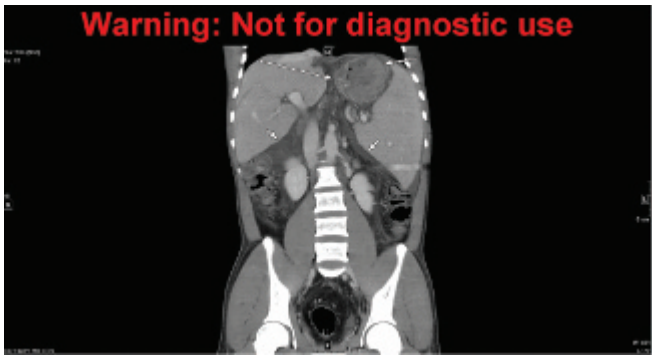

Figure 4. Coronal computed tomography image post contrast during portal venous phase demonstrating ascites (*short arrows*), gastro-esophageal junction varices (*long arrow*), blood in gastric lumen (*medium arrow*), and enlarged spleen.

Gastroscopy identified multiple varices throughout the esophagus. Fourteen bands were applied to the varices. No gastric or duodenal ulcers were visualized. Adequate hemostasis was achieved, and the gastroscope was withdrawn.

Computed tomography of the abdomen and pelvis documented a small liver consistent with cirrhosis, massive splenomegaly, severe portal hypertension with ascites, and varices (Figure 3 and Figure 4). The patient underwent liver biopsy, which demonstrated portal and periportal fibrosis and foreign bodies consistent with *S. mansoni* eggs. No eggs were identified in stool samples. He was treated with praziquantel orally. A decision was made to proceed with a mesocaval shunt to control the severe portal hypertension. He was discharged from hospital and continues to live in the area.

CONCLUSION

Significant and life-threatening hemorrhage secondary to esophageal varices is possible in young adults who fall outside the much more common cohort of older, chronic alcoholic patients. Presinusoidal portal hypertension manifested by esophageal varices secondary to chronic schistosomiasis should be considered in recent immigrants from endemic areas who present with massive upper gastrointestinal bleeding.

Competing interests: None declared.

REFERENCES

1. Weaver DH, Maxwell JG, Castleton KB. Mallory-Weiss syndrome. Am J Surg 1969;118:887-92, doi:10.1016/0002-9610(69)90252-9.
2. Watts HD, Admirand WH. Mallory-Weiss syndrome. A reappraisal. JAMA 1974;230:1674-5, doi:10.1001/jama.1974.03240120042018.
3. Michel L, Serrano A, Malt RA. Mallory-Weiss syndrome. Evolution of diagnostic and therapeutic patterns over two decades. Ann Surg 1980;192:716-21, doi:10.1097/00000658-198012000-00004.
4. Sugawa C, Benishek D, Walt AJ. Mallory-Weiss syndrome. A study of 224 patients. Am J Surg 1983;145:30-3, doi:10.1016/0002-9610(83)90162-9.
5. Harris JM, DiPalma JA. Clinical significance of Mallory-Weiss tears. Am J Gastroenterol 1993;88:2056-8.
6. Skok P. Fatal hemorrhage from a giant Mallory-Weiss tear. Endoscopy 2003;35:635, doi:10.1055/s-2003-40214.
7. Knauer CM, Mallory-Weiss syndrome. Characterization of 75 Mallory-Weiss lacerations in 528 patients with upper gastrointestinal hemorrhage. Gastroenterology 1976;71:5-8.
8. Kovacs TO, Jensen DM. Endoscopic diagnosis and treatment of bleeding Mallory-Weiss tears. Gastrointest Endosc Clin North Am 1991;1:387.
9. Bharucha AE, Gostout CJ, Balm RK. Clinical and endoscopic risk factors in the Mallory-Weiss syndrome. Am J Gastroenterol 1997;92:805-8.
10. Laine L, Peterson WL. Bleeding peptic ulcer. N Engl J Med 1994;331:717-27, doi:10.1056/NEJM199409153311107.
11. Wara P, Stodkilde H. Bleeding pattern before admission as guideline for emergency endoscopy. Scand J Gastroenterol 1985;20:72-8, doi:10.3109/00365528509089635.
12. Jensen DM, Machicado GA. Diagnosis and treatment of severe hematochezia: the role of urgent colonoscopy after purge. Gastroenterology 1988;95:1569-74.
13. Mumtaz R, Shaukat M, Ramirez FC. Outcomes of endoscopic treatment of gastroduodenal Dieulafoy's lesion with rubber band ligation and thermal/injection therapy. J Clin Gastroenterol 2003;36:310-4, doi:10.1097/00004836-200304000-00006.
14. Lee YT, Walmsley RS, Leong RW, et al. Dieulafoy's lesion. Gastrointest Endosc 1993;58:236-43, doi:10.1067/mge.2003.328.
15. Pollack R, Lipsky H, Goldberg RI. Duodenal Dieulafoy's lesion. Gastrointest Endosc 1993;39:820-2, doi:10.1016/S0016-5107(93)70276-X.
16. Anireddy D, Timberlake G, Seibert D. Dieulafoy's lesion of the esophagus. Gastrointest Endosc 1993;39:604, doi:10.1016/S0016-5107(93)70198-4.
17. Linhares MM, Filho BH, Schraibman V, et al. Dieulafoy lesion: endoscopic and surgical management. Surg Laparosc Endosc Percutan Tech 2006;16:1-3, doi:10.1097/01.sle.0000202191.59322.5f.
18. Gryseels B. Schistosomiasis. Infect Dis Clin North Am 2012;26:383-97, doi:10.1016/j.idc.2012.03.004.
19. Gryseels B, Polman K, Clerinx J, et al. Human schistosomiasis. Lancet 2006;368:1106-18, doi:10.1016/S0140-6736(06)69440-3.
20. Chitsulo L, Engels D, Montresor A, et al. The global status of schistosomiasis and its control. Acta Trop 2000;77:41-51, doi:10.1016/S0001-706X(00)00122-4.
21. Bottieau E, Clerinx J, De Vega MR, et al. Imported Katayama fever: clinical and biological features at presentation and during treatment. J Infect 2006;52:339-45, doi:10.1016/j.jinf.2005.07.022.
22. Lambertucci JR. Acute schistosomiasis: clinical, diagnostic and therapeutic features. Rev Inst Med Trop Sao Paulo 1993;35:399-404, doi:10.1590/S0036-46651993000500003.
23. Cheever AW, Hoffmann KF, Wynn TA. Immunopathology of schistosomiasis mansoni in mice and men. Immunol Today 2000;21:46566, doi:10.1016/S0167-5699(00)01626-1.
24. Thoeni RF. The role of imaging in patients with ascites. AJR Am J Roentgenol 1995;165:16-8.
25. Cattau EL, Benjamin SB, Knuff TE, et al. The accuracy of the physical examination in the diagnosis of suspected ascites. JAMA 1982;247:1164-6, doi:10.1001/jama.1982.03320330060027.
26. Amin HM, Omran SA, el-Bassuoni NE, et al. Assessment of factors II, VII, IX, X and protein C in hepatosplenic schistosomiasis. Haemostasis 1994;24:22-6.
27. Bottieau E, Clerinx J. Imported Katayama fever: clinical and biological features at presentation and during treatment. J Infect 2006;52:339-45, doi:10.1016/j.jinf.2005.07.022.
28. Ross AG, Vickers D. Katayama syndrome. Lancet Infect Dis 2007;7:218-24, doi:10.1016/S1473-3099(07)70053-1.
29. Jauréguiberry S, Paris L, Caumes E. Acute schistosomiasis, a diagnostic and therapeutic challenge. Clin Microbiol Infect 2010;16:225-31, doi:10.1111/j.1469-0691.2009.03131.x.

For the challenge, see page 160.