

A rare cause of acute mesenteric ischemia: JAK2 positivity and chronic active hepatitis B

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ABSTRACT

Short bowel syndrome occurs as a result of insufficiency in the total length of the small intestine to provide adequate supply of nutrients. Seventy-five percent of cases are due to massive intestinal resection. A 35-year-old male complaining of abdominal pain was admitted to the gastroenterology department. A CT scan was performed, showing total occlusion of the portal vein and superior mesenteric vein. During the operation, widespread edema and necrosis of the small intestine were found. The necrotic segments of the small intestine were resected. The spleen was larger than normal and, in some parts, infarcts were evident, thus splenectomy was also performed during surgery. A second-look procedure was performed 24 hours later, and an additional 10 cm jejunal resection and anastomosis was performed. His further evaluations revealed myeloproliferative disease and chronic active hepatitis B leading to thrombosis. Essential thrombocytosis and portal vein thrombosis are common in hepatitis B infection. Patients with complaints of abdominal pain in the context of essential thrombocytosis and hepatitis B should be handled with caution as they are at risk of developing portal vein thrombosis.

Key Words: Portal vein thrombosis, myeloproliferative disease, hepatitis B

INTRODUCTION

Short bowel syndrome results from the inability of the total length of the small intestine to provide adequate nutritional support. Seventy-five percent of cases are caused by massive intestinal resection (1). In adults, mesenteric occlusion, midgut volvulus and traumatic avulsion of superior mesenteric veins are the most common causes (1). In this article, an atypical short bowel syndrome resulting from portal vein thrombosis that occurred in a patient with JAK 2 positivity and chronic active hepatitis B, will be presented.

CASE PRESENTATION

A 35-year old male patient complaining of abdominal pain was admitted to the gastroenterology clinic. After obtaining informed consent from the patient examinations were started. The WBC: $8.87 \times 10^3/\mu\text{L}$, hemoglobin 14 g/dL, and platelet were $626 \times 10^3/\mu\text{L}$. The abdominal ultrasonography revealed splenomegaly and portal vein thrombosis, and the upper gastrointestinal endoscopy identified prominent esophageal submucosal veins. The abdominal computed tomography showed complete occlusion of the portal and superior mesenteric veins and splenomegaly (Figure 1). Due to aggravation of the patient's abdominal pain and increasing white blood cell count ($22 \times 10^3/\mu\text{L}$) an emergency operation was decided. During the operation, generalized edema and necrosis in the small bowel loops, from 55 cm. distal to the ligament of Treitz up to 35 cm proximal to the ileocecal valve was identified (Figure 1). The necrotic bowel segment was resected. The spleen was larger than normal and contained areas of infarction, thus splenectomy was performed. Since there were concerns regarding the viability of the remaining small intestine, a second-look operation was planned and the patient was transferred to the General Surgery Intensive Care Unit. The second-look operation was done 12 hours later. An additional ischemic segment was observed in a 10 cm segment of jejunum and resection and anastomosis was performed. Etiologic evaluations were started. On peripheral blood smear, hypersegmentation, thrombocytosis and erythroblasts were seen. Genetic tests revealed positivity for JAK-2 V617F gene mutation. Hematology consultation was requested for essential thrombocytosis. Bone marrow aspiration biopsy and flow cytometry studies were performed. Results were consistent with myeloproliferative disease. It was found out that the patient had previously been diagnosed with chronic hepatitis B, but he did not receive any treatment. The serologic tests showed HBs Ag (including verification): 5968 (positive) and HBV-DNA: 1.14×100.000 IU/mL (positive) (test measuring range 20 IU/mL-170 million IU/mL). The patient

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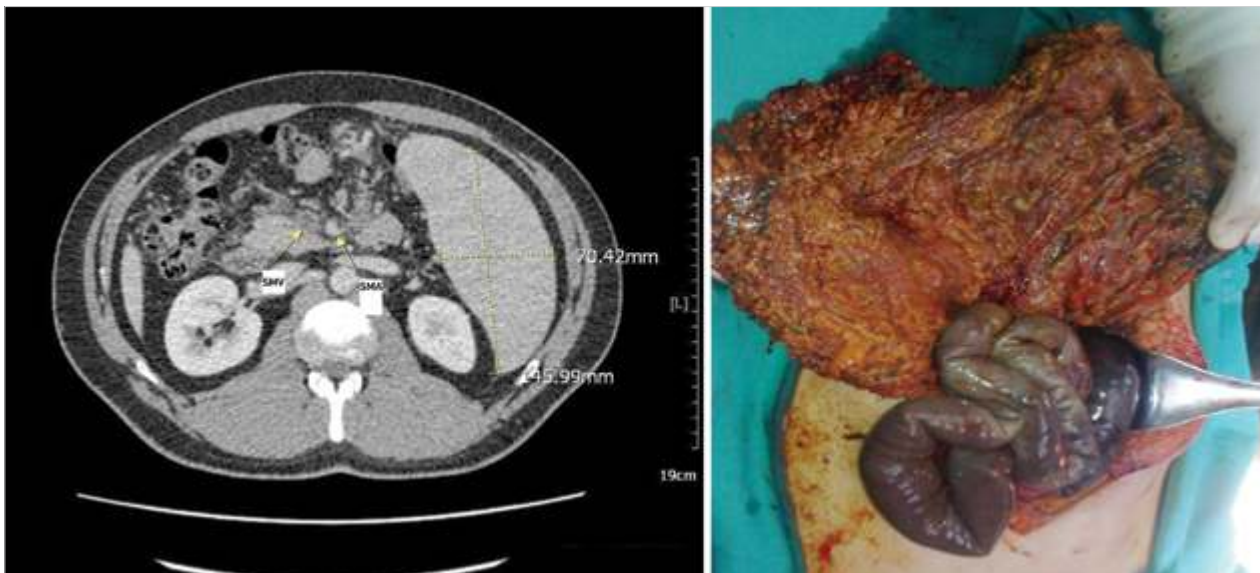


Figure 1. On CT the superior mesenteric vein is completely thrombosed and splenomegaly is detected. The photo image of operative findings: Intestinal necrosis and edema

was started on 500 mg hydroxyurea (capsule) twice daily, and bid 100 mg lamivudine (tablets). He was discharged and planned for outpatient clinic follow-up.

DISCUSSION

Mesenteric and portal vein occlusion is a frequent clinical presentation to general surgery and gastroenterology clinics. Essential thrombocytosis is a disease characterized by increased numbers of platelets within circulation. Hematopoietic cells become more sensitive to erythropoietin and thrombopoietin due to a phenylalanine to valine substitution mutation in the important signal transduction protein JAK 2 V617F (2). Up to 50 to 60% of essential thrombocytosis cases are due to mutations in the JAK 2 (3). Mesenteric and portal vein thrombosis is a rare complication of myeloproliferative diseases. In patients with JAK 2 V617F mutation and venous thrombosis, the splenic vein is effected at rates ranging from 12-74% (4, 5). Determining the incidence of acute mesenteric occlusion is difficult due to lack of data. Nevertheless, Acosta et al. (6) reported this rate as 12.9 in 100.000. This rate will increase with the increase in the elderly population (7). The early diagnosis of acute mesenteric ischemia increases survival (8). The main reasons for delay in diagnosis and treatment are delayed presentation to hospitals, and lack of disease-specific symptoms, physical examination and laboratory findings. Akyüz et al. (9), from our department, found that mortality significantly increased in patients who presented with a more than 24-hour delay between onset of complaints and hospital admission. In our patient, the diagnosis was made within 24 hours and surgery was performed in time, which might have contributed to the favorable outcome. Different options are available in the surgical treatment of acute mesenteric ischemia. Resection and anastomosis, resection and stoma formation and additional second-look operations may be used. Type of surgical procedures by should be decided according to the patient's clinical status and co-morbidities. Second look operation 24-48 hours after the initial operation, to observe the viability of bowel movements was first proposed in 1965, by Shaw. This

method enables the assessment of bowel viability and the effect of heparinization (10). We had doubts about the viability of remaining small bowel in our patient that is why we felt the need to make a second look. In this way, we were able to identify an additional 10-cm segment of progressive necrosis and perform a resection.

Hepatitis B virus infection is reported to be associated with portal venous thrombosis in Southeast Asian countries, in hepatic cirrhosis and carcinoma (11). In a series of 467 patients, thrombosis in several veins has been compared and portal vein thrombosis has been found in 194 (41%) patients. The most common causes of this thrombosis were malignancy in 126 patients (hepatocellular carcinoma, cholangiocellular carcinoma), and infection in 98 patients (hepatitis B, hepatitis C) (11). In the same study, it has been shown that hepatitis C resulted in a higher rate of portal venous thrombosis than hepatitis B (11). In that particular study, only one patient was reported to have essential thrombocytosis and portal vein thrombosis (11). This might set an example for the interesting nature of our case. Chronic hepatitis B virus increases coagulation via anticardiolipin antibodies (ACA) and increases thrombosis formation in the portal vein (12). Portal vein thrombosis was observed in 8 out of 11 patients (72%) with hepatitis B-related hepatocellular carcinoma and ACA (+), whereas out of 54 hepatitis B-related hepatocellular carcinoma and ACA (-), portal vein thrombosis was observed in 14 patients (26%) (11). In our case ACA was (-).

CONCLUSION

Concomitant essential thrombocytosis and hepatitis B infection increase tendency for portal vein thrombosis. In patients with essential thrombocytosis and hepatitis B infection, portal vein thrombosis must be considered as part of differential diagnosis in the evaluation of abdominal pain.

Informed Consent: Written informed consent was obtained from patients who participated in this case.

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REFERENCES

1. Scolapi JS, Flemming CR. Short bowel syndrome. *Gastrointestinal Clin North Am* 1998; 27: 467-479. [\[CrossRef\]](#)
2. McLornan D, Percy M, McMullin MF. JAK2 V617F: a single mutation in the myeloproliferative group of disorders. *Ulster Med J* 2006; 75: 112-119.
3. Sokołowska B, Nowaczyńska A, Bykowska K, Chocholska S, Wejksza K, Walter-Croneck A, et al. JAK2 mutation status, hemostatic risk factors and thrombophilic factors in essential thrombocythemia (ET) patients. *Folia Histochem Cytobiol* 2011; 49: 267-271. [\[CrossRef\]](#)
4. P'ng S, Carnley B, Baker R, Kontorinis N, Cheng W. Undiagnosed myeloproliferative disease in cases of intra-abdominal thrombosis: the utility of the JAK2 617F mutation. *Clin Gastroenterol Hepatol* 2008; 6: 472-475. [\[CrossRef\]](#)
5. Brandt LJ, Boley SJ. AGA technical review on intestinal ischemia. *American Gastrointestinal Association. Gastroenterology* 2000; 118: 954-968. [\[CrossRef\]](#)
6. Acosta S. Epidemiology of mesenteric vascular disease: clinical implications. *Semin Vasc Surg* 2010; 23: 4-8. [\[CrossRef\]](#)
7. Florian A, Jurcut R, Lupescu I, Grasu M, Croitoru M, Ginghina C. Mesenteric ischaemia-a complex disease requiring a multidisciplinary approach. A review of the current literature. *Roman J Intern Med* 2010; 48: 207-222.
8. Yasuhara H. Acute mesenteric ischemia: The challenge of gastroenterology. *Surg Today* 2005; 35: 185-195. [\[CrossRef\]](#)
9. Akyüz M, Sözüer E, Akyıldız H, Akcan A, Küçük C, Poyrazoğlu B. Surgical results in acute mesenteric ischemia. *Kolon Rektum Hast Derg* 2010; 20: 121-126.
10. Shaw R. The "second look" after superior mesenteric embolectomy or reconstruction for mesenteric infarction. *Curr Surg Manage* 1965: 509.
11. Lertpipopmetha K, Auewarakul CU. High incidence of hepatitis B infection-associated cirrhosis and hepatocellular carcinoma in the Southeast Asian patients with portal vein thrombosis. *BMC Gastroenterol* 2011; 11: 66. [\[CrossRef\]](#)
12. Elefsiniotis IS, Diamantis ID, Dourakis SP, Kafiri G, Pantazis K, Mavrogiannis C. Anticardiolipin antibodies in chronic hepatitis B and chronic hepatitis D infection, and hepatitis B-related hepatocellular carcinoma. Relationship with portal vein thrombosis. *Eur J Gastroenterol Hepatol* 2003; 15: 721-726. [\[CrossRef\]](#)