Case Report

Delayed development of portal vein thrombosis in a patient initially detected with portal venous gas and pneumatosis intestinalis: a case report

Masatoku Arai, Shiei Kim, Hiromoto Ishii, Jun Hagiwara, Toru Takiguchi, Yoshito Ishiki, and Hiroyuki Yokota

Department of Emergency & Critical Care Medicine, Nippon Medical School, Bunkyo-ku, Tokyo, Japan

Background: Portal venous gas (PVG) and pneumatosis intestinalis (PI) are rare pathologic findings, and a delayed appearance of portal vein thrombosis (PVT) in such patients is extremely rare.

Case Presentation: A 51-year-old man complaining of epigastric pain was referred to our hospital. Computed tomography (CT) at admission revealed massive PVG and extensive PI, but no PVT. Emergency laparotomy was carried out, but bowel resection was unnecessary. On follow-up CT on postoperative day 5, thrombosis was noted in the portal venous system, and anticoagulant was started immediately. This patient was discharged and continued to take the anticoagulant. Seven months after discharge, PVT had disappeared on CT without any thromboembolic complications.

Conclusion: If acute PVT is detected, anticoagulant is needed to prevent bowel ischemia and/or portal hypertension due to the growth of the thrombus. Clinicians should be aware of the potential for such a complication, and make their best efforts to exclude this entity using CT or sonography.

Key words: Pneumatosis intestinalis, portal vein thrombosis, portal venous gas

BACKGROUND

PORTAL VENOUS GAS (PVG) and pneumatosis intestinalis (PI) are rare radiographic findings indicating underlying intra-abdominal pathology, such as mechanical causes, acute mesenteric ischemia, benign idiopathic, and abdominal infection, the clinical significance of which ranges from benign to catastrophic.[1–3] Portal vein thrombosis (PVT) is also a rare occurrence with numerous potential causes. Cirrhosis and cancer are the leading causes of PVT;[4,5] however, 10% of cases of PVT are attributed to major abdominal infectious or inflammatory disease.[5] Portal venous gas and/or PI are reported to be sometimes associated with acute PVT;[6–8] however, a delayed appearance of PVT not detected on initial computed tomography (CT) at admission in patients with PVG and PI is rarely reported.

We herein report a case of acute PVT appearing late in a patient with PVG and PI on follow-up CT.

Corresponding: Masatoku Arai, MD, PhD, Department of Emergency & Critical Care Medicine, Nippon Medical School, 1-1-5 Sendagi, Bunkyo-ku 113-8603 Tokyo, Japan. E-mail: amasatoku@nms.ac.jp.

Received 25 Mar, 2019; accepted 22 Jul, 2019; online publication 8 Aug, 2019

Funding Information

No funding information provided.

CASE

A 51 -year-old man complaining of epigastric pain, vomiting, and diarrhea was found to have PVG and PI on CT without contrast in another hospital and was referred to our hospital. His medical history did not indicate liver cirrhosis, malignancy, or predisposing procoagulant state.

The patient had a blood pressure of 146/92 mmHg, a heart rate of 157 b.p.m., and a body temperature of 37.2°C. A physical examination showed a distended abdomen and tenderness over the whole abdomen with slight rebound tenderness. The laboratory data on admission showed a white blood cell count of 7,200/mm^3 (normal range, 4,000–8,000/mm^3) with 78.8% segmented neutrophils (normal range, <70%), C-reactive protein 24.59 mg/dL (normal range, <0.3 mg/dL), procalcitonin 12.65 ng/mL (normal range, <0.05 ng/mL), blood urea nitrogen 63.9 mg/dL (normal range, 7–20 mg/dL), creatinine 5.08 mg/dL (normal

range, 0.5–1.0 mg/dL), prothrombin time 88.1% (normal range, 70–130%), activated partial thromboplastin time 35.8 s (normal range, 24–37 s), fibrinogen/fibrin degradation products 132.1 μg/mL (normal range, <10 μg/mL), D-dimer 51.4 μg/mL (normal range, <0.5 μg/mL), antithrombin III 97.8% (normal range, 80–120%), and lactate 4.9 mmol/L (normal range, 0.56–1.39 mmol/L). Blood cultures were positive for coagulase-negative staphylococci.

Abdominal X-ray showed a distended small bowel loop with remarkable PI (Fig. 1A), and CT with contrast in our hospital revealed a massive amount of air in the portal venous system and extensively throughout the small intestinal wall; however, portomesenteric vein thrombosis was not seen. Although vessel contrast enhancement was intact on CT, the partial attenuation of the contrast-induced enhancement of the bowel wall, bowel wall edema, and distension were identified, and bowel ischemia/necrosis due to non-occlusive mesenteric ischemia was suspected (Figs. 1B, 2A, B).

Emergency laparotomy was carried out, and a small amount of serous ascites was seen. Approximately 3 m of the small intestine from the ligament of Treitz appeared to show slight ischemic changes and extensive dilatation, however, immediate bowel resection was deemed unnecessary. Postoperative management included broad-spectrum antibiotics and continuous hemodiafiltration for the treatment of acute kidney injury. The next day, second-look laparotomy was carried out. As no bowel necrosis was evident, the abdomen was closed. Our final diagnosis was gastroenteritis, however, we could not identify the causative pathogen.

After surgery, the postoperative course was uneventful. Computed tomography was undertaken for follow-up after laparotomy on postoperative day (POD) 5, and thrombosis was noted in the right portal vein and the umbilical portion of the left portal vein (Fig. 2C, D). Heparinization was started on the same day and then changed to the oral direct factor Xa inhibitor edoxaban at 60 mg/day. On follow-up CT on POD 39, thrombosis in the right portal vein had disappeared completely, but left PVT showed spreading (Fig. 2E, F). As neither abnormal laboratory data findings nor complaints were seen, this patient was discharged and continued to take the oral anticoagulant. On follow-up CT at our outpatient clinic on POD 74, left PVT was still noted; however, the size had been reduced. Seven months after discharge, it had disappeared on follow-up CT without any thromboembolic complications, and edoxaban was discontinued.

DISCUSSION

PORTAL VENOUS GAS and PI are rare pathologic conditions. In approximately 50% of reported cases, PVG presented with PI,[1] with the clinical significance ranging from benign to catastrophic.[1–3] The indications of surgical intervention remain controversial.[1–3] However, a recent study recommended that surgical exploration should be considered for PI patients presenting with elevated lactate levels and peritonitis[3] because of the strong suspicion of bowel necrosis. In the present case, we carried out emergency

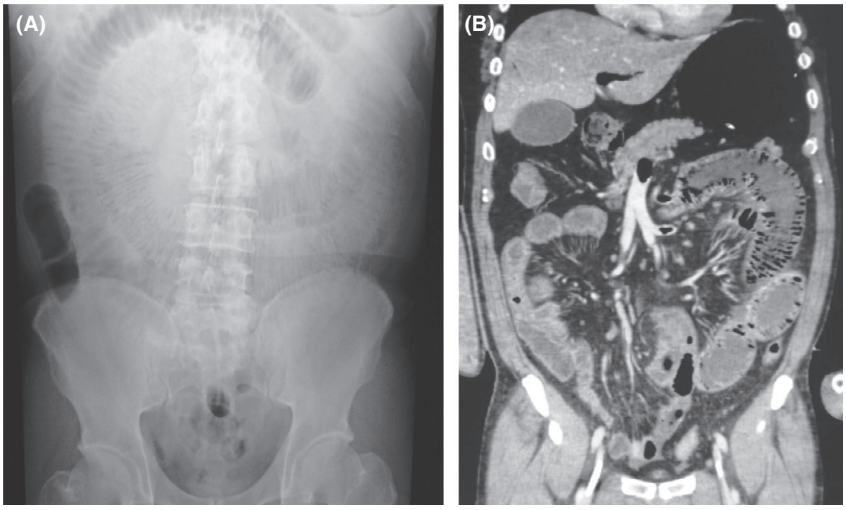

Fig. 1. Abdominal X-ray and computed tomography scan (coronal view) on admission to our hospital of a 51-year-old man with epigastric pain, vomiting, and diarrhea. A, Abdominal X-ray showed a distended small bowel loop with remarkable pneumatosis intestinalis. B, Coronal-view computed tomography scan with contrast revealed air in the portal venous system and extensively throughout the small intestinal wall; however, portal venous thrombosis was not seen.

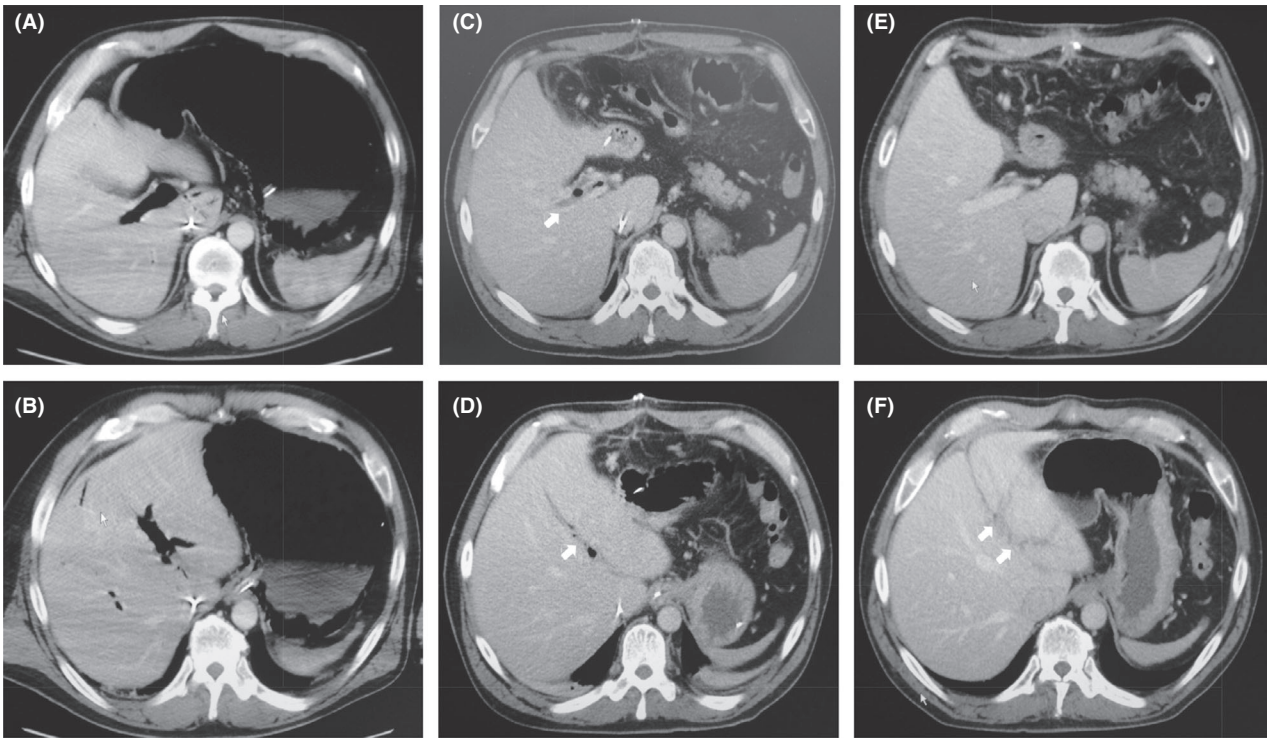

Fig. 2. Changes in portal venous gas and portal vein thrombosis (PVT) on computed tomography (CT) with contrast (axial view) in a 51-year-old man with epigastric pain, vomiting, and diarrhea. A, B, Portal venous gas was seen in the right (A) and left (B) portal vein on abdominal CT at admission; however, PVT was not yet seen. C, D, On postoperative day 5, thrombosis was noted in the right portal vein (C) and the umbilical portion of the left portal vein (D) on abdominal CT (white arrow). A small amount of air still remained. E, F, On follow-up CT on postoperative day 39, thrombosis in the right portal vein had disappeared completely (E), but left PVT showed spreading (F) (white arrows).

laparotomy, but there was no need for resection. Abdominal surgery has been shown to be involved in the formation of PVT. However, such occurrence involves the direct injury or manipulation of the portal vein.[9] Although emergency laparotomy was carried out in the present study, it was only a probe laparotomy and a second look without direct injury or manipulation of the portal vein, suggesting that the main cause of the formation of PVT might have been abdominal infection with PI/PVG rather than laparotomy.

Portal vein thrombosis is also rare, with numerous causes. Although the most common predisposing conditions for PVT are cirrhosis and cancer, major infectious or inflammatory abdominal disease has been reported to account for 10% of cases, according to the results of a large population study based on consecutive autopsies.[5] Portal venous gas and/or PI was shown to be associated with acute PVT in previous studies;[6–8,10,11] however, the delayed appearance of PVT, which was not noted on initial CT at admission, in a patient with PVG and PI is rarely reported. Indeed, we undertook a detailed search of works published in English using the PubMed database using the keywords "portal vein thrombosis" and "portal venous gas" or "portal vein thrombosis" and "pneumatosis intestinalis" in adults, and the full texts were examined. Only three cases of the delayed appearance of PVT with PVG and/or PI in adults have been reported to date.[8,10,11]

Once PVT has been diagnosed, not only broad-spectrum antibiotics but also anticoagulants are recommended in order to prevent bowel ischemia and/or portal hypertension due to the growth of the thrombus.[4–6] Anticoagulant therapy is recommended to be continued for at least 3 months in all patients with acute PVT according to the guidelines.[4] As the PVT was still present on follow-up CT in the present patient, edoxaban was continued for 7 months after discharge.

In the present study, the formation of PVT might have been the result of hypercoagulability due to bacteremia. Indeed, the laboratory data on admission showed increased levels of C-reactive protein and procalcitonin, and increases of fibrinogen/fibrin degradation products and D-dimer were supposed to be due to a hyperfibrinolysis state. Although initial CT imaging with contrast on admission did not detect

evident PVT, the microthrombus formation might have already begun.

It can be difficult to determine why patients suffer from PVT. Indeed, it was previously reported that no predisposing factors were detected in 14% of patients with PVT.[5] It is caused by a combination of general and local risk factors.[4] General risk factors, such as an inherited or acquired prothrombotic condition, have been found in many patients with PVT.[4] In the present study, although not all general risk factors were detected, no suspected procoagulant episodes were observed throughout the course of the patient's life. In contrast, among local risk factors, as cancer, cirrhosis, and injury to the portal vein had been excluded, the most likely local risk factor seemed to be abdominal infection due to gastroenteritis with PVG and PI, which can result in vascular endothelial cell injury and the subsequent formation of a microthrombus. In addition, we speculated that PVT might have developed due to venous congestion caused by extensive massive PVG. Taken together, these findings suggest that abdominal infection and massive PVG might have influenced the vascular endothelial cell injury and subsequent formation and development of PVT in the present case.

As there is no established evidence concerning whether or not the prophylactic use of anticoagulant therapy will be effective in this situation, follow-up CT was considered important for the prompt diagnosis of the late formation of PVT in the present case, however, the ideal timing of follow-up CT remains unclear. According to the guideline of acute PVT, if PVT is suspected, CT with contrast should be undertaken for the prompt confirmation or exclusion of this diagnosis.[4] If CT is not readily available, Doppler sonography is a reasonable alternative approach, and if the findings suggest acute PVT, CT should be undertaken rapidly.[4] Follow-up CT or Doppler sonography is thus suggested to be extremely important for the prompt detection of the late formation of PVT in patients with PI and/or PVG.

CONCLUSION

WE ENCOUNTERED A case of the late appearance of acute PVT in a patient with PVG and PI. If PVT is detected, anticoagulation therapy should be begun in order to prevent bowel ischemia and/or portal hypertension due to the growth of the thrombus. However, as no specific recommendations exist concerning the prophylactic use of anticoagulants in this situation, the prompt diagnosis of PVT seems important for preventing these disastrous complications. Clinicians should be aware of the potential for such complications and make their best efforts to exclude this entity using CT or sonography.

DISCLOSURE

Approval of the research protocol: Ethical approval to report this case was not required.
Informed consent: Written, informed consent was obtained from the patient for publication of this case report and any accompanying images.
Registry and registration no. of the study/trial: N/A.
Animal studies: N/A.
Conflict of interest: None declared.

REFERENCES

1 Nelson AL, Millington TM, Sahani D *et al*. Hepatic portal venous gas: the ABCs of management. Arch. Surg. 2009; 144: 575–81.
2 Wayne E, Ough M, Wu A *et al*. Management algorithm for pneumatosis intestinalis and portal venous gas: treatment and outcome of 88 consecutive cases. J. Gastrointest. Surg. 2010; 14: 437–48.
3 Ferrada P, Callcut R, Bauza G *et al*. Pneumatosis Intestinalis Predictive Evaluation Study: a multicenter epidemiologic study of the American Association for the Surgery of Trauma. J. Trauma Acute Care Surg. 2017;82:451–60.
4 DeLeve LD, Valla DC, Garcia-Tsao G; American Association for the Study Liver Diseases. Vascular disorders of the liver. Hepatology 2009; 49: 1729–64.
5 Ogren M, Bergqvist D, Björck M *et al*. Portal vein thrombosis: prevalence, patient characteristics and lifetime risk: a population study based on 23,796 consecutive autopsies. World J. Gastroenterol. 2006; 12: 2115–9.
6 Hong D, Lovrics P. Soft-tissue images. Portal vein thrombosis and gas formation: unusual presentation of colon cancer. Can. J. Surg. 2001; 44: 90–1.
7 Ng SS, Yiu RY, Lee JF *et al*. Portal venous gas and thrombosis in a Chinese patient with fulminant Crohn's colitis: a case report with literature review. World J. Gastroenterol. 2006; 12: 5582–6.
8 Wakisaka M, Mori H, Kiyosue H *et al*. Septic thrombosis of the portal vein due to peripancreatic ligamental abscess. Eur. Radiol. 1999; 9: 90–2.
9 Thomas RM, Ahmad SA. Management of acute post-operative portal venous thrombosis. J. Gastrointest. Surg. 2010; 14: 570–7.
10 Al-Jundi W, Shebl A. Emphysematous gastritis: case report and literature review. Int. J. Surg. 2008; 6: e63–6.
11 Ma AS, Ewing I, Murray CD *et al*. Hepatic portal venous gas and portal venous thrombosis following colonoscopy in a patient with terminal ileal Crohn's disease. BMJ Case Rep. 2015; 4: 1–3.