Clinical Features and Management of Hepatic Portal Venous Gas

Four Case Reports and Cumulative Review of the Literature

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Hypothesis: Hepatic portal venous gas (HPVG) has been considered a rare entity associated with a grave prognosis. Since 1978, when Liebman et al reviewed 64 cases of HPVG and reported a mortality of 75%, the number of reported cases has been increasing.

Design: Case series.

Patients and Methods: We reviewed the literature on 182 cases of HPVG in adults, including 4 of our patients, (transplantation and abdominal trauma cases were excluded) and analyzed the cause, pathogenesis, and clinical features.

Results: In this series, the underlying clinical events associated with HPVG were bowel necrosis (43%), digestive tract dilatation (12%), intraperitoneal abscess (11%), ulcerative colitis (4%), gastric ulcer (4%), Crohn disease (4%), complications of endoscopic procedures (4%), intraperitoneal tumor (3%), and other (15%). The overall mortality was 39% but varied depending on the underlying disease.

Conclusions: Hepatic portal venous gas is a lethal or curable entity caused by various diseases. The underlying disease associated with HPVG determines the clinical features and prognosis of the patients. The treatment of patients with HPVG should be directed to the underlying disease.

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Hepatic portal venous gas (HPVG) was first described by Wolf and Evans1 in 1955 in infants with necrotizing enterocolitis. In 1960, Susman and Senturia2 reported the first adult case, a patient critically ill with small bowel infarction. Thereafter, many authors have reported HPVG. Liebman et al3 reviewed 64 cases of HPVG in 1978 and reported a mortality of 75%. Since then, the number of reported cases has increased because of improvements in diagnostic methods, including computed tomography (CT) and ultrasonography, and the overall mortality rate has decreased because of adequate therapy. This study reviews the clinical data from 182 cases of this entity in adults from the literature, including 4 patients we treated, and discusses the management of HPVG relative to the underlying disease.

REPORT OF CASES

CASE 1

A 66-year-old man was referred to Wakayama Medical University Hospital, Wakayama, Japan, because of a fever spiking higher than 40°C and chills. He had a history of chronic hepatitis C, liver cirrhosis, and diabetes mellitus. The abdomen was soft and slightly distended without tenderness. Blood analysis showed a white blood cell count of 6.4 × 10^9/L, C-reactive protein value of 0.16 g/L, platelet count of 35 × 10^9/L, and prothrombin time of 12.7 seconds. Abdominal CT demonstrated splenomegaly, splenic abscess, and a tumor of the descending colon, which penetrated to the spleen. In addition, there were branching radiolucencies in the peripheries of the liver and no enhancement of the portal vein by intravenous administration of contrast medium (Figure 1).

Emergency laparotomy was carried out under the diagnosis of cancer of the descending colon invaded to the spleen. Cancer of the splenic flexure with narrowing of the descending colon and infiltration into the spleen was found, and left hemicolectomy and splenectomy were performed, with construction of a stoma with transverse colon. Microscopically, the tumor exposed the serosa and penetrated to the spleen, with formation of abscess. The
patient had liver abscess postoperatively, and percutaneous transhepatic abscess drainage was performed on the seventh day after the operation. The patient was treated for giant liver abscess at another hospital. Two months after the operation, he was discharged, and no recurrence has been noted.

CASE 2

A 57-year-old man was referred to our hospital with chief complaints of severe abdominal pain with involuntary guarding and rebound tenderness. The patient had had distal gastrectomy for a gastric peptic ulcer at age 47 years. Laboratory data revealed a white blood cell count of 14.7 × 10^9/L, prothrombin time of 14.6 seconds, glutamic oxaloacetic transaminase level of 361 U/L, lactate dehydrogenase level of 411 U/L, serum urea nitrogen level of 24 mg/dL (8.6 mmol/L), and C-reactive protein level of 0.12 g/L. Blood gas analysis at room temperature demonstrated: pH, 7.11; PaCO₂, 85 mm Hg; PaO₂, 22.2 mm Hg; bicarbonate, 6.7 mmol/L; and base excess, −21.3 mmol/L, which indicated metabolic acidosis.

Plain abdominal radiography and CT showed an abundance of free intraperitoneal air and multiple branching radiolucencies to the liver (Figure 2). An emergency operation was performed because of the diagnosis of peritonitis and portal venous gas because of bowel necrosis. At laparotomy, an extensive necrotic change was seen from the stomach to the sigmoid colon. Air bubbles were seen in all the mesenteric veins, and pulsation was absent in the celiac axis and superior mesenteric artery. The patient died 3 hours after the operation. No postmortem examination was performed.

CASE 3

A 51-year-old woman with progressive systemic sclerosis was referred to our hospital because of a continuous fever and abdominal distension. Nine days before admission, she had had a partial resection of the ileum for intestinal necrosis in another hospital. Her blood pressure was 139/75 mm Hg, pulse rate was 120/min, the abdomen was distended with decreased bowel sounds, and involuntary guarding was shown with rebounding tenderness. Laboratory data indicated a white blood cell count of 24.1 × 10^9/L, prothrombin time of 19.6 seconds, creatine kinase level of 211 U/L, lactate dehydrogenase level of 411 U/L, serum urea nitrogen level of 24 mg/dL (8.6 mmol/L), and C-reactive protein level of 0.12 g/L. Blood gas analysis at room temperature demonstrated: pH, 7.11; PaCO₂, 85 mm Hg; PaO₂, 22.2 mm Hg; bicarbonate, 6.7 mmol/L; and base excess, −21.3 mmol/L, which indicated metabolic acidosis.

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CASE 4

A 57-year-old man was admitted to our hospital with disturbed consciousness, hypotension at 60 mm Hg, and tachycardia at 141/min. The patient had a history of alcohol abuse, chronic hepatitis C, liver cirrhosis, esophageal varices, and diabetes mellitus. Physical examination revealed general jaundice, abdominal involuntary guarding, and an absence of bowel sounds. Laboratory data showed leukocytopenia, thrombocytopenia, and severe liver and kidney dysfunction. The arterial pH was 7.10; PaCO₂, 50 mm Hg; bicarbonate, 12.9 mmol/L; and base excess, −13.3 mmol/L. Abdominal CT showed multiple small tubular lucencies in the periphery of the liver, in addition to intraperitoneal effusion and splenomegaly.

The patient was moved to the intensive care unit to improve his general condition. Nevertheless, he died 4 hours after admission. An autopsy confirmed diffuse bowel necrosis, including the stomach to the sigmoid colon, secondary to obstruction of the superior mesenteric artery.

Figure 1. A, Air is present in the peripheral branches of the portal vein (arrows) in patient 1. B, Computed tomogram shows gas within the superior mesenteric vein (long arrow) and distal splenic vein (short arrow).
The clinical features of these 4 cases are summarized in Table 1. Three patients who had bowel necrosis presented with severe metabolic acidosis, and 2 of them died after their operation or during treatment. The patient with HPVG associated with colon cancer underwent an appropriate operation immediately and survived. Analysis of our surviving cases indicated that an appropriate operation was required if HPVG was caused by bowel necrosis, abscess, or massive damage to the mucosa of the alimentary canal.

**Table 1. Summary of the Clinical Features of 4 Case Reports**

<table>
<thead>
<tr>
<th>Case No./Age/Sex</th>
<th>Underlying Disease</th>
<th>Degree of Bowel Necrosis</th>
<th>Blood Pressure, mm Hg</th>
<th>Base Excess, mmol/L</th>
<th>Operation</th>
<th>Complications</th>
<th>Clinical Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/66/M</td>
<td>Colon cancer</td>
<td>None</td>
<td>104/67</td>
<td>4.0</td>
<td>Left hemicolecotomy and splenectomy</td>
<td>Liver abscess</td>
<td>Alive</td>
</tr>
<tr>
<td>2/57/M</td>
<td>Bowel necrosis</td>
<td>Jejunum to the middle of the ascending colon</td>
<td>97/57</td>
<td>-11.5</td>
<td>Resection of small intestine and colon</td>
<td>Short-bowel syndrome</td>
<td>Alive</td>
</tr>
<tr>
<td>3/51/F</td>
<td>Bowel necrosis</td>
<td>Stomach to sigmoid colon</td>
<td>139/75</td>
<td>-21.3</td>
<td>Exploratory laparotomy</td>
<td>Multiple organ failure</td>
<td>Dead</td>
</tr>
<tr>
<td>4/57/M</td>
<td>Bowel necrosis</td>
<td>Stomach to sigmoid colon</td>
<td>60/34</td>
<td>-13.3</td>
<td>None</td>
<td>Multiple organ failure</td>
<td>Dead</td>
</tr>
</tbody>
</table>
CUMULATIVE REVIEW OF THE LITERATURE

We summarized the clinical data from 182 cases of HPVG, including the present cases, in Table 2. The mean age of patients with this entity was 55 years, and 16 patients manifested signs of shock on admission. Patient management included an operation in 83 cases (46%) and conservative therapy in 79 (43%). The overall mortality was 39%. Hepatic portal venous gas occurred in different clinical scenarios (Table 3), including bowel necrosis (43%), digestive tract dilatation (12%), intraperitoneal abscess (11%), ulcerative colitis (4%), gastric ulcer (4%), Crohn disease (4%), complications of endoscopic procedures (4%), intraperitoneal tumor (3%), and other (15%).

The mortality rate varied based on the underlying disease. The mortality was high in cases of HPVG associated with bowel necrosis (75%), and there was a tendency for the degree of bowel necrosis to affect mortality (Table 4). On the other hand, there were no lethal cases of HPVG associated with ulcerative colitis, intraperitoneal tumor, Crohn disease, cholangitis, pancreatitis, or complications of endoscopic procedures.

Table 2. Clinical Characteristics of 182 Cases*

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex</td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>84 (62)</td>
</tr>
<tr>
<td>Female</td>
<td>51 (38)</td>
</tr>
<tr>
<td>Age, range (mean), y</td>
<td>13-85 (55)</td>
</tr>
<tr>
<td>Shock on admission, No.†</td>
<td>16</td>
</tr>
<tr>
<td>Management</td>
<td></td>
</tr>
<tr>
<td>Operation</td>
<td>83 (51)</td>
</tr>
<tr>
<td>Conservative</td>
<td>79 (49)</td>
</tr>
<tr>
<td>Prognosis</td>
<td></td>
</tr>
<tr>
<td>Alive</td>
<td>99 (61)</td>
</tr>
<tr>
<td>Dead</td>
<td>63 (39)</td>
</tr>
</tbody>
</table>

*Data are given as number (percentage) unless otherwise indicated. Categories do not sum to 182 because of missing data.
†The criteria used to diagnose shock were those of the Committee on Trauma, American College of Surgeons.

COMMENT

Hepatic portal venous gas was first described in 1955 by Wolf and Evans.¹ The pathogenesis of HPVG is not fully understood. Two sources of its origin have been proposed: (1) an escape of gas from increased pressure in the bowel lumen or in an abscess and then circulation into the liver or (2) the presence of gas-forming bacteria in the portal venous system and passage of gas into the circulation. Both findings were seen in many cases of HPVG.²,⁵,¹⁰

Hepatic portal venous gas is recognized not as a specific disease entity but as a diagnostic clue in patients with acute abdominal pathology. The diagnosis of HPVG is usually made by plain abdominal radiography, CT, or ultrasonography. It is diagnosed by the appearance of branching lucencies within 2 cm of the liver capsule on plain abdominal radiography or CT, whereas biliary gas usually is found within the central portion of the liver more than 2 cm from the liver capsule. Gas in the portal venous system is likely transported to the small peripheral branches in the liver by the centrifugal flow of portal venous blood, whereas gas in the biliary tree is prevented from migrating peripherally by the centripetal flow of bile.

Computed tomography has a higher sensitivity for the detection of HPVG than plain radiography.³,¹⁰ The number of cases of HPVG diagnosed in the United States has been steadily increasing.¹¹⁻¹⁴ The ultrasonographic features of HPVG have been reported as (1) echogenic particles flowing within the portal vein or (2) poorly defined, echogenic patches within the hepatic parenchyma, especially in the nondependent part.¹⁵ Schulze et al⁶ reported that color Doppler sonograms showed bright spots moving with the bloodstream.

Most cases of HPVG are caused by mesenteric vascular occlusion and subsequent bowel necrosis. However, HPVG has occurred with various conditions, some of which were cured with conservative management alone, including digestive tract dilatation,⁷⁻¹⁰,¹²⁻¹⁶ gastric ulcer,⁸⁻¹⁰,¹²⁻¹⁶,¹¹⁻¹³,¹⁷ ulcerative colitis,⁸⁻¹⁰,¹²⁻¹⁶,¹¹⁻¹³,¹⁷ Crohn disease,⁸⁻¹⁰,¹²⁻¹⁶,¹¹⁻¹³,¹⁷ and complications of endoscopic procedures.¹⁸⁻²⁰ However, other cases were potentially lethal, and an emergency operation has been recommended in conditions such as abscesses⁹⁻¹³ and tumors.⁸⁻¹³⁻¹⁴,²¹⁻²³,⁴¹ All 4 of our patients required an emergency operation. From our surviving cases, it appeared that survival in potentially lethal cases is dependent on adequate treatment of the underlying disease associated with HPVG. Statistical analysis did not show a correlation between age, heart rate, or base excess on admission and survival rate.
However, relative to the high mortality rate of HPVG associated with bowel necrosis, the underlying disease might be an important factor contributing to patient survival. There was a tendency toward association of degree of bowel necrosis with mortality.

Hepatic portal venous gas has been considered an ominous sign, with an overall mortality higher than 75%. Because of poor clinical outcomes, exploratory laparotomy is recommended. In reviewing the cases in the literature, the overall mortality was 39%, but this was mitigated by a large number of patients with HPVG with underlying diseases other than bowel necrosis, which was associated with a 75% mortality. Operations were not performed in patients with HPVG associated with digestive tract dilatation, gastric ulcer, ulcerative colitis, Crohn disease, or complications of endoscopic procedures, because these conditions are curable with conservative management alone. The mortality was 38% in operated cases and 39% in nonoperated cases. There was no statistical difference in mortality rates between the operated and nonoperated cases in the cumulative review.

In conclusion, HPVG is not a specific disease entity but rather a diagnostic clue in patients with acute abdominal pathologic conditions. The treatment of patients with HPVG should be directed to the underlying disease, and the indications for an emergency operation should be based on the primary disease, with consideration of the high mortality rate of HPVG associated with bowel necrosis.

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REFERENCES


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