Abstract

Hepatic portal venous gas (HPVG) is the existence of air in the hepatic portal venous system. It was considered very rare in the past literatures before the introduction of computed tomographic (CT) scan. With special image feature in the abdominal CT scan, HPVG has been well known to be associated with high mortality even while urgent surgical intervention is performed for acute mesenteric infarction. In this paper, we present a 55 year-old woman who was admitted due to dialytic fluid related peritonitis with the manifestations of severe abdominal distension, severe diarrhea, fever, and septic shock. HPVG formation was detected in the CT scan of abdomen, as well as pneumatosis intestinalis (PI) and severe aorta atherosclerosis, implying small bowel necrosis, which was possibly originated from acute mesenteric infarction. Operation was not performed due to severely deteriorated condition. The patient rapidly died despite of antibiotic combination therapy and intensive hemodynamic support. Early recognition of the HPVG and PI combination signs is crucial for prompt diagnosis and management for acute mesenteric infarction. (J Intern Med Taiwan 2007; 18: 146-151)

Key Words: Hepatic portal venous gas, Pneumatosis intestinalis
Introduction

Hepatic portal venous gas (HPVG) is an impressive image finding that air exists within hepatic portal venous but not in the biliary system. With the wide use of computed tomographic (CT) scan and ultrasonography (US) in the patient with acute abdomen, early detection of HPVG is possible and more and more HPVG with various causes are recognized. This image finding highly associates with pneumatosis intestinalis (PI), which may result from several conditions such as acute mesenteric ischemia, intra-abdominal infection, blunt abdominal trauma, and intestinal obstruction. Although patients with HPVG were considered to have high mortality rate, some cases with benign courses were reported in recent years. Most survivors did not have transmural infarction or necrosis of the intestine in the image. Therefore, HPVG formation coexistent with PI in the abdominal CT scan could be used as prognostic indicator for patients with acute abdomen.

Case Report

A 55-year-old woman was hospitalized due to shortness of breath and fever. Her medical history included hypertension without regular medication control for ten years; and end stage renal disease with continuous ambulatory peritoneal dialysis (CAPD) for 3 years. Two month earlier before this admission, sudden onset of cardiac arrest due to acute myocardial infarction of inferior wall was noted. She remained vegetative state because of hypoxic encephalopathy post cardiopulmonary resuscitation since then. After a tracheostomy was created, she was transferred to a local respiratory care ward (RCW). The patient had only one abdominal surgery for the implantation of CAPD tube three years ago. She did not use alcohol, and was reported no known allergies by her family.

For recent ten days, the patient had been noticed to be febrile off and on which was accompanied with severe watery diarrhea in the RCW. Empirical antibiotic was prescribed without improvement. Shortness of breath deteriorated, so she was brought to the emergency department of this hospital.

On examination, the patient appeared persistent vegetative state. Her temperature was 36.3 °C. Her pulse was regular at 128 beats per minute, blood pressure, 123/76 mmHg; and respiratory rate, 26 breaths per minute. Her neck was supple with a tracheostomy and there was no palpable cervical lymphadenopathy. Her chest was clear to auscultation and there was no heart murmur. Her abdomen was distended. Other tests showed no remarkable abnormalities.

Laboratory data were as follows. The electrocardiograph revealed a sinus tachycardia. Chest radiography showed elevated diaphragm, probably due to obesity stature, and increased lung markings without clinical significance. Her white cell count was 11,900 cells/μL and C-reactive protein was 30.20 mg/dL. Biochemical laboratory tests included a serum sodium level of 131.1 mmol/L; potassium, of 3.17 mmol/L; and glucose, 195 mg/dL. Specimens of urine, blood, and sputum were obtained for culture. Empirical antimicrobial therapy with 1200 mg of amoxicillin/clavulanate was administered intravenously every 8 hours.

On the second hospital day, the patient remained febrile, unstable blood pressure, and persistent shortness of breath. Coffee ground substance drained from nasogastric tube was noted. Continuous intravenous infusion of dopamine was needed to maintain adequate mean blood pressure despite normal saline resuscitation about two liters. She was transferred to medical intensive care unit with an impression of septic shock and assisted with mechanical ventilation via tracheostomy. The patient was placed a central venous catheter via right jugular vein. After transfer to medical intensive care unit, she was changed CAPD fluid and found pus-like appearance CAPD fluid in the next several times. Her CAPD fluid analysis re-
revealed a white cell count of 161,865 cells/μL with neutrophil 91%. Because of the above findings, she was suggested abdominal CT scan for possible intraabdominal lesions workup. CT scan of the abdomen without contrast media revealed severe abdominal aorta atherosclerosis, intrahepatic portal venous gas accumulation (Fig. 1), dilated small bowel with air-fluid level (Fig. 2), implying transmural infarction of the small bowel (Fig. 3).

Diagnosis of small bowel necrosis was made.

Fig. 1. Hepatic portal venous gas (white arrow) in the branching pattern within the portal vein and severe abdominal aorta atherosclerosis (white arrowhead) shown in an abdominal computed tomographic scan.

Discussion

Hepatic portal venous gas (HPVG) is an image finding due to the presence of gas in the portal vein system, as branching radiolucency extending to the periphery of the liver. It was first described in six infants with necrotizing enteritis to have this image features by Wolfe and Evans in 1955 and was considered as pediatric disease. In 1960, Susman and Senturia reported the first adult case associated with small bowel infarction. Surgical implications and clinical significance in adult patients were analysed in a retrospective study with 64 cases by Liebman et al. in 1978. The major cause was bowel necrosis (72%), followed by intraabdominal abscess (6%), gastric ulcers bleeding (3%), bowel obstruction (3%),

Fig. 2. Circumferential mixed bubble-like pneumatosis intestinalis (white arrow) and dilated small bowel with air-fluid level (white arrowhead).

Fig. 3. Transmural small bowel infarction (white arrowhead): circumferential band-like small bowel wall.
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and barium enemas in patients with stable ulcerative colitis (8%). Because of its high mortality up to 75%, Liebman et al. suggested urgent abdominal exploration in all patients of HPVG but not in ulcerative colitis which had a benign course.

Before the introduction of ultrasonography (US) and abdominal computed tomography (CT), HPVG was detected in large gas amount at late stage by plain abdomen. The radiographic criteria for HPVG was a branching radiolucency extending to within 2 cm of the liver capsule. Plain films were most revealing of HPVG when taken in the left lateral decubitus position of the patient. In the radiography, difficult differential diagnosis included necrotic metastasis lesion, air embolism in biliary tree, and gas-forming intrahepatic abscess.

With the development of other image study modalities, resolution was better by US than abdominal radiography. US detected early and small amount of HPVG before any radiographic abnormalities and was considered of great value in the early evaluation. Gosink used diagnostic US to examine twelve patients with intrahepatic gas and a useful differential diagnosis was obtained. Because of the simplicity, higher sensitivity and specificity in the abdomen, US was once suggested in emergency department for screening of the ominous prognostic image sign in bowel necrosis from mesenteric arterial occlusion. However, diagnosis of parenchymal abnormalities would be missed due to the prominent shadowing of intrahepatic gas in abdominal US causing disruption of the normal hepatic parenchymal pattern.

The CT scan of the abdomen is suggested as good as abdominal US for HPVG detection and considered extremely helpful now in localizing extraluminal gas to the liver and differentiating between air in the biliary tree and hepatic portal venous gas. After the introduction of abdominal CT scan, precise differential diagnosis from biliary tree air embolism, intrahepatic abscess, metastatic lesion, and parenchymal liver diseases are possible without the drawback of abdominal US, as well as detection of extrahepatic lesions.

HPVG and pneumatosis intestinalis (PI) are generally considered as separate entities separately in the radiologic findings. Many causes are associated with PI, such as infectious and inflammatory abdominal diseases, interventional procedures, trauma, and transplantation. The prognosis resulted from the above diseases is benign and conservative therapy is commonly required. However, the most common cause resulting to PI is acute mesenteric ischemia, and it is most frequently observed as extrahepatic air accumulation with HPVG, accounting of 50-75% of cases. Significant association between HPVG and PI was established in the era of abdominal CT scan.

Many cases were detected either by the US or CT scan of the abdomen after mid-1980s. More patients receiving prompt surgical intervention after earlier diagnosis of HPVG survived. The absolute indication for exploratory laparotomy is questioned because some benign courses or non-occlusive arterial ischemia are noted. Although mortality is still high, some survivors without surgical treatment were reported in the literatures, depending on the causes of the event and the degree and extent of ischemic damage of the bowel wall.

The pathogenesis of HPVG is not yet well understood. Liebman et al proposed that possible factors predisposing to HPVG are mucosal damage, bowel distention, and sepsis by their clinical observation. Many patients show two of these findings (50%) and some show three (35%) in their study. Although HPVG is rare in the living people, it is commonly found in postmortem CT scan performed immediately in patients with non-traumatic death after cardiopulmonary resuscitation, which is considered compatible with mucosal damage and bowel distention.

In those surviving patients with benign course, air existing in the mesenteric circulation is readily absorbed within one day. This is possibly because of
non-transmural bowel infarction and recovery of the bowel from ischemic event. Walter and Bharti suggested a three-stage theory combined with the findings of abdominal CT scan. Stage-I and -II diseases represent partial mural ischemia of the bowel. Stage I is called reversible ischemic enterocolitis. Mucosal necrosis, erosions and ulcerations, with/without hemorrhage are characteristic in this stage. Fibrotic strictures stemming from local reparative changes were found in stage II with the development of the deep submucosal and muscular layers necrosis as the infarction area more deeply into the bowel wall. Distinct from stages I and II, stage-III disease is defined as transmural necrosis of the bowel wall. It requires immediate surgical intervention but still has a high mortality rate. Therefore, the HPVG appearance in the CT scan of the abdomen should not be recognized as a sole prognostic sign but warrant further workup of possible acute mesenteric infarction and its severity.

Our patient presented typical signs in portomesenteric gas formation. The CT scans of the abdomen showed branching hypoattenuating HPVG extending into the periphery of both liver lobes. Dilated small bowel with air-fluid level in the lumen, massive circumferential, mixed bubble-like and band-like pneumatosis in multiple necrotic loops, consisted with diffuse transmural infarction of the small bowel. The patient did not receive any operation because refractory shock and severe metabolic acidosis. The rapid fatality of the clinical course highlights the importance of early detection for the signs of hepatic portal venous gas and pneumatosis intestinalis.

References
在一持續性攜帶式腹膜透析末期腎病病人的肝門靜脈積氣和腸壁積氣

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摘 要

肝門靜脈積氣，指的是肝門靜脈裡面出現氣體堆積。當電腦斷層影像尚未出現之前，在過去的文獻資料中它被視為是非常罕見的情形。它的電腦斷層影像特徵很特別，若病人出現肝門靜脈積氣，就算緊急開刀處理急性腸壁膜動脈梗塞，仍舊有相當高的死亡率。本文報告一名55歲女性病人以重度腹瀉、嚴重腹瀉、發燒和敗血性休克為症狀的腹膜透析液腹膜炎而住院。肝門靜脈積氣以及腸壁積氣的影像出現在她的腹部電腦斷層上，並且加上重度主動脈硬化，可能是由於腸壁膜動脈梗塞的腸壞死。然病人病情不佳而無法開刀，儘管合併多種抗生素使用和重症積極治療，病人依然不治。故我們認為倘早發現肝門靜脈積氣和腸壁積氣這兩種影像學特徵並做出診斷以及處理腸壁膜動脈梗塞是相當重要的。