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Case Report

Ectopic Varices in a Right Diaphragm that Ruptured into the Pleural Cavity

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The term ectopic varices is used to describe dilated portosystemic collateral veins in unusual locations other than the gastroesophageal region. We recently experienced a rare case of ectopic varices that developed in the right diaphragm and ruptured into the pleural cavity. A 68-year-old female with hepatocellular carcinoma complicated with liver cirrhosis was admitted due to an acute onset of dyspnea and right bloody pleural effusion. Because of the patient's advanced hepatocellular carcinoma and poor condition, conservative therapies such as hemostats and blood transfusion were selected. Even though the bleeding to the pleural cavity stopped spontaneously, the patient died due to a progression of liver failure. Autopsy revealed a huge collateral vein in the right diaphragm. The etiology, prevalence, relationship with portal hypertension, and treatment of ectopic varices are discussed herein.

Key words: ectopic varices, portal hypertension, collateral vein, diaphragm

Ectopic or aberrant varices are dilated portosystemic venous collaterals with unusual locations, and they are typically associated with portal hypertension. Ectopic varices have been reported to develop in various organs such as the duodenum, colon, gall bladder, uterus, urinary bladder, and abdominal stomas [1–8]. However, varices other than gastroesophageal or rectal are rare entities [3]. We report herein a patient with hepatocellular carcinoma (HCC) complicated with ectopic varices that ruptured into the right pleural cavity.

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Case Report

A 68-year-old woman had been treated with HCC complicated with liver cirrhosis due to hepatitis C virus infection at Tone Chuo Hospital, Numata, Japan. She had been diagnosed with liver cirrhosis in 1985 by her primary physician. She developed HCC at 1992 and thus came to our hospital. Her previous history other than the liver cirrhosis and family history were unremarkable. We treated her HCC with transcatheter arterial embolization (TAE) and percutaneous ethanol injection (PEIT) therapies for over 4 years. She had esophageal varices that were obliterated by repeated endoscopic ligation and sclerotherapy. A complete eradication of the esopha-

geal varices was noted after endoscopic treatment. However, new HCC lesions appeared in the liver and progressed. Finally, 4 years after the first treatment, the HCC could no longer be controlled. Computed tomography (CT) showed a huge tumor in the liver with a longest diameter of 14 cm that showed central necrosis compatible with HCC (Fig. 1A, 1B). She was therefore treated with supportive care at our outpatient clinic. One day, she suddenly experienced dyspnea and thus was admitted to our hospital. Physical examination revealed anemia, icterus, tachycardia, ascites, and decreased respiratory sounds in the right lower lung fields, although her blood pressure and consciousness were normal. Laboratory data at admission were as follows: hemoglobin 7.9 g/dl, platelets 45,000 /mm^3, total protein 6.0 g/dl, serum albumin 2.4 g/dl, blood urea nitrogen 25 mg/dl, creatinine 1.1 mg/dl, total bilirubin 3.5 mg/dl, direct bilirubin 2.0 mg/dl, aspartate aminotransferase 90 IU/l, alanine aminotransferase 55 IU/l, lactate dehydrogenase 420 IU/l, alkaline phosphatase 410 IU/l, gamma-glutamyltranspeptidase 101 IU/l, cholinesterase 1.9 IU/ml, total cholesterol 154 mg/dl, prothrombin time 42.4%, alpha-fetoprotein 707,995 ng/ml, and protein induced by vitamin K absence or antagonist-II (PIVKA-II) 1,520 AU/ml. Hepatitis B surface antigen was negative and hepatitis C virus antibody was positive. A chest X-ray (Fig. 2) showed the right pleural effusion. Thoracentesis revealed bloody pleural effusion, and the hemoglobin of the effusion was 2.6 g/dl. Because of the sudden onset, bloody pleural effusion, and huge HCC, we presumed that there was bleeding from the HCC and did no further examinations to diagnose the bloody effusion at that time. Because of the presence of the advanced stage of HCC, we performed supportive care with thoracentesis, hemostats, and blood transfusion. Bleeding to the pleural cavity was spontaneously stopped by conservative therapy. However, she died due to liver failure 40 days after the onset of these symptoms. Autopsy revealed a huge liver tumor, as previously identified by the CT findings and histological findings, which was diagnosed to be moderately differentiated HCC and liver cirrhosis. Dilated collateral vessels attached to adipose tissue were observed on the right diaphragm (Fig. 3A, 3B). The histological findings of the vessels revealed a dilated vein containing red blood cells inside the

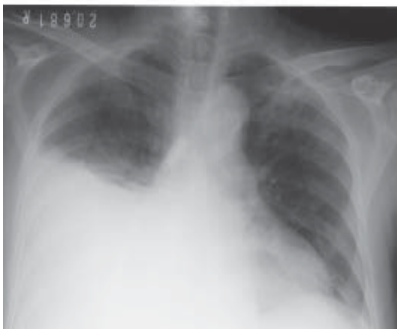

Fig. 2 A chest X-ray showed right pleural effusion, and thoracentesis revealed bloody pleural effusion.

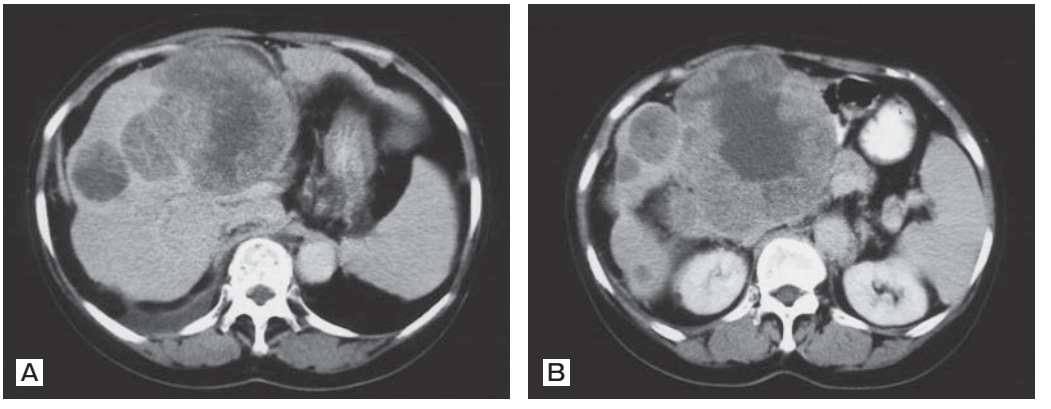

Fig. 1 Computed tomography (CT) showed a huge tumor in the liver with a longest diameter of 14 cm that showed central necrosis compatible with hepatocellular carcinoma.

lumen (Fig. 3C). Because thin-slice CT scanning was not performed while the patient was alive, the precise status of the collateral veins was not revealed by CT scans before autopsy. Finally, we diagnosed the patient as having ectopic varices that developed on the right diaphragm and then ruptured into the pleural cavity.

Discussion

The recognition of varices at unusual sites has long been described in the literature, since Alberti *et al.* [9] described duodenal varices in 1931. Ectopic varices have been reported in the duodenum, ileum, cecum, ascending, descending and rectosigmoid colon, gall bladder, uterus, vagina, urinary bladder, and abdominal stomas [2–10]. Although ectopic varices can occur at several sites, bleeding ectopic varices are most commonly found in the duodenum and at sites of previous bowel surgery, including stomas [3]. In a review of 169 cases of bleeding ectopic varices, 26% of the cases occurred in the stoma, followed by 17% in the duodenum, 17% in the jejunum or ileum, 14% in the colon, 8% in the rectum, and 9% in the peritoneum [3]. Cardiophrenic varices, particularly on the right side, have also sometimes been observed in patients with portal hypertension [11]. Our case may be classified as a variation of cardiophrenic varices. However, such varices are usually located at a cardiophrenic angle, and rupture is rare. Huge varices on the diaphragm complicated with a rupture, as seen in our case, are rare. The origins of varices in

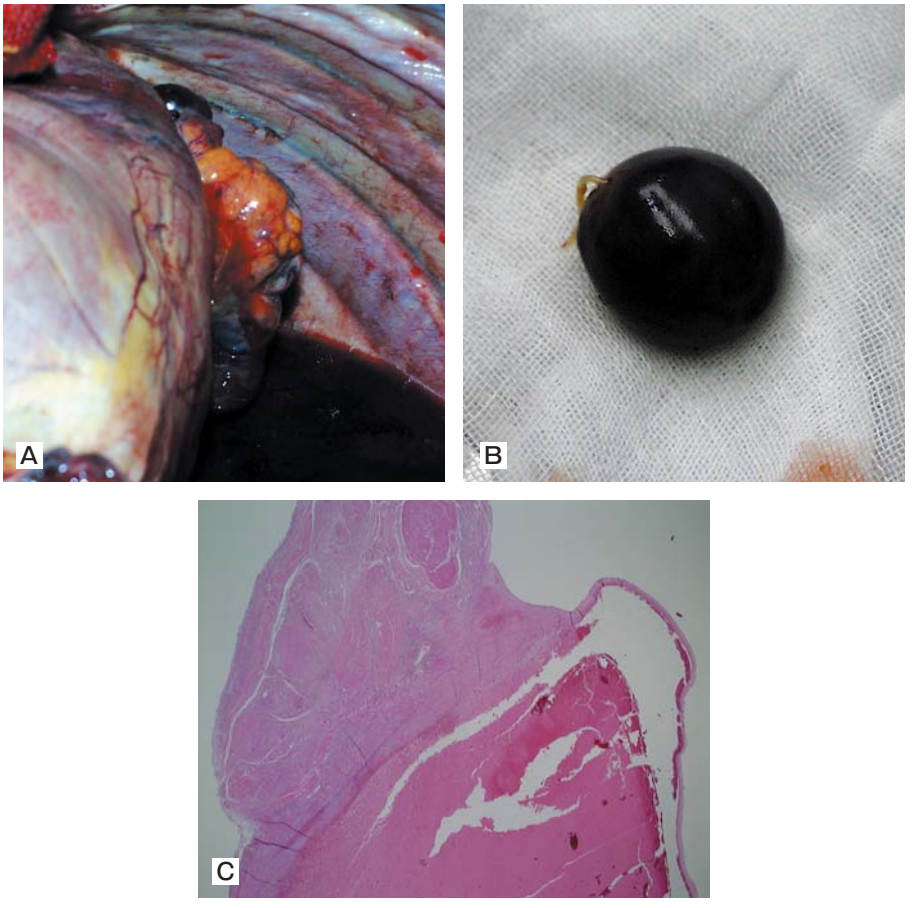

Fig. 3 **A**, Dilated collateral vessels attached to adipose tissue were observed on the right diaphragm at autopsy; **B**, Part of the dilated vessel; **C**, The histological findings revealed a dilated vein containing red blood cells inside the lumen.

this case were collaterals from the paraumbilical vein. Our case was complicated with huge HCC, and it invaded large vessels such as the hepatic and portal vein. Such invasion to large vessels may thus have contributed to the development of these unusual varices.

Uncontrollable bleeding from gastroesophageal varices has sometimes caused death in patients with portal hypertension in the past decade [1]. Recently, endoscopic or interventional treatments for gastroesophageal varices have improved due to such treatment modalities as endoscopic ligation therapy, sclerotherapy, and balloon-occluded retrograde transvenous obliteration (BRTO) [1]. The survival and prognosis of patients with bleeding from gastroesophageal varices have improved, and ectopic varices are also sometimes observed after treatment of gastroesophageal varices. In our case, the patient had undergone endoscopic therapy for esophageal varices, and a complete eradication of these varices was noted. A complete eradication of esophageal varices may be one reason for the development of the ectopic varices.

The management of ectopic varices is frequently difficult and controversial. Bleeding from ectopic varices is rare and accounts for only between 1% and 5% of all variceal bleeding [3]. However, once they start to bleed, they are difficult to control and sometimes fatal. In our case, the progression of HCC and the patient's poor condition led us to select conservative treatment. A surgical resection or interventional embolization of varices is sometimes useful when the varices are localized. Somatostatin analog or β-blocker has also been used to control the bleeding from varices [3]. However, surgical options such as a portosystemic shunt or variceal ligation are limited to selected patients. Unfortunately, many patients are not good operative candidates for such treatment modalities. The transjugular intrahepatic portosystemic shunt (TIPS) procedure is an effective modality in therapy for cirrhotic patients with bleeding from ectopic varices unresponsive to conservative management [12–14].

In conclusion, we experienced a rare case of ectopic varices at the right diaphragm that ruptured into the right pleural cavity. Although they are rare, we should consider ectopic varices as a possible cause of hemothorax when bloody pleural effusion suddenly appears in cirrhotic patients.

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