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The White Nipple Sign: Please Do Not Disturb

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Key Words

White nipple · Variceal hemorrhage · Mount St. Helens' sign

Abstract

Blood spurting or oozing from a varix confirms the diagnosis of variceal hemorrhage. In most cases of variceal hemorrhage, however, the bleeding has ceased by the time endoscopy is performed. Endoscopists rely on identification of stigmata of recent hemorrhage to determine whether varices are the cause of bleeding and to predict the likelihood of rebleeding. Most of the attention has focused on red color signs, such as red wale markings, described by Beppu et al. [Gastrointest Endosc 1981;27:213–218] and well known to endoscopists. Here we describe our experience with a less recognized stigma of variceal hemorrhage known as the 'white nipple sign', which resulted in active hemorrhage when manipulated.

Introduction

Portal hypertension is the most common complication of chronic liver disease, leading to the development of ascites, portal-systemic encephalopathy, and gastroesophageal varices. Hemorrhage from gastroesophageal varices is among the most lethal complications of cirrhosis and portal hypertension [1]. Esophageal varices develop at a rate of 5% per year in patients with cirrhosis and portal hypertension [1, 2]. In patients with esophageal varices, hemorrhage occurs at a yearly rate of 5–15% [1, 2]. Esophageal variceal bleeding is associated with a 15–20% early mortality [1, 2]. In patients who survive the initial hemorrhage, the risk of rebleeding is as high as 60% within 2 years, with a 33% mortality [1].

Because of the high mortality associated with variceal hemorrhage, as well as the high likelihood of rebleeding after a first episode, the treatment of portal hypertension includes attempts at preventing the first bleed (primary prophylaxis), management of acute variceal hemorrhage, and the prevention of rebleeding (secondary prophylaxis) [1, 2]. Primary prophylaxis constitutes treatment with a non-selective beta-blocker to reduce

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portal pressure. For those who cannot tolerate beta-blockade, endoscopic therapy in the form of endoscopic variceal band ligation can be performed and repeated until the varices are obliterated [2]. In those patients who have recovered from acute esophageal variceal hemorrhage, secondary prophylaxis or prevention of further variceal bleeding should be performed in the form of endoscopic variceal ligation, in addition to portal pressure-reducing medication [2].

The treatment of choice for bleeding esophageal varices is a combination of pharmacologic therapy with somatostatin and its analogues (such as octreotide) or terlipressin, and endoscopic therapy, preferably variceal ligation [1, 2]. Blood spurting or oozing from a varix confirms the diagnosis of variceal hemorrhage, however in most cases the bleeding has ceased by the time endoscopy is performed. It is therefore important for the endoscopist to identify and recognize signs of recent variceal hemorrhage. The most commonly seen stigmata are red color signs, including red wale marks (longitudinal red streaks on varices that resemble red corduroy wales), cherry red spots, and hematocystic spots (raised discrete red spots on the surface of varices that appear as blood blisters) [1, 3, 4]. A less commonly recognized stigma of recent variceal hemorrhage is a white nipple-like projection from a varix into the esophageal lumen [1, 2, 4–6]. This platelet-fibrin plug, also referred to as the ‘white nipple sign’, is indicative of a site of recent bleeding [1, 4–6]. Recognition of this stigma of recent hemorrhage is necessary so that endoscopic therapy may be performed in order to prevent further variceal bleeding [4–6]. Here we describe our encounter with the white nipple sign.

Case Report

A 53-year-old male with a past medical history of alcoholic cirrhosis, hypertension, and depression presented to the emergency department with a history of hematemesis of frank red blood and clots. Admission vital signs were: heart rate 83, respiratory rate 18 with SpO₂ 94% and blood pressure 188/99 mm Hg. Physical examination revealed no jugular venous distention. Mild gynecomastia was present. Respiratory and cardiovascular examinations were normal. The abdomen was protuberant with the presence of ascites. There was appreciable hepatomegaly with a palpable left lobe of the liver. Admission laboratory data included INR 1.3, WBC 4,400/μl, hemoglobin 12.2 g/dl, and platelets $51 \times 10^6/\mu\text{l}$. His basic metabolic panel revealed serum sodium 143 mmol/l, potassium 3.6 mmol/l, chloride 103 mmol/l, bicarbonate 28 mmol/l, BUN 6 mg/dl and creatinine 0.6 mg/dl. AST and ALT were 60 and 154 U/l respectively, alkaline phosphatase 201 U/l, total bilirubin 1.6 mg/dl, total protein 7.8 g/dl and albumin 3.3 g/dl.

In the emergency department, he was started on octreotide infusion and given intravenous pantoprazole. Emergent endoscopy revealed grade II–III esophageal varices without any active bleeding. There were no red wale markings or other red color signs. However, one of the larger varices had a protuberant white spot (fig. 1). The spot was initially irrigated with water and was resistant to dislodgement. It was then inadvertently dislodged with gentle suctioning, resulting in brisk hemorrhage (fig. 2). Variceal band ligation was promptly performed with successful control of bleeding (fig. 3). The patient was monitored in the intensive care unit post-procedure and was maintained on octreotide infusion for 96 h. Antibiotic prophylaxis with intravenous piperacillin-tazobactam was initiated. He continued to demonstrate hemodynamic stability without evidence of recurrent bleeding and was transferred out of the intensive care unit 2 days later. The remainder of the hospitalization was uneventful and the patient was successfully discharged home 6 days after admission on beta-blocker therapy. He was scheduled for repeat endoscopy and variceal ligation the following week.

Discussion

Though not widely mentioned as a stigma of recent variceal hemorrhage, this finding was first reported by Chung and Lewis in 1984 [4]. They described it as a white nipple on top of a varix protruding into the lumen and noted its presence in up to 5% of cases of variceal bleeding. It was thought to represent a platelet-fibrin plug at the site of recent variceal rupture. They noted that dislodgement would be accompanied by jet-like bleeding and called it ‘Mount St. Helens’ sign’ due its volcanic-like eruption.

Few reports have commented on the significance or prognostic value of this finding. Two prospective studies have evaluated the finding of the ‘white nipple sign’ as a diagnostic sign of recent hemorrhage. Siringo et al. [5] evaluated the white nipple sign prospectively in 203 separate admissions for esophageal variceal bleeding in 145 cirrhotic patients. They identified a white nipple at endoscopy in 9% of cases. Endoscopic therapy was not initially delivered to these patients in order to evaluate the risk of rebleeding. They felt that the white nipple sign was diagnostic of a varix that had recently bled but found that it had no adverse prognostic significance. In another study done in Taiwan [6], 166 patients with recent variceal hemorrhage were prospectively studied with respect to presence or absence of the white nipple sign and to risk of rebleeding following endoscopic therapy. The authors noted a white nipple sign in 21% of their cases of variceal hemorrhage. They concluded that the white nipple sign suggested recent variceal hemorrhage and was more likely to be seen if endoscopy was performed soon after the bleeding episode.

Our case reaffirms the finding of a white nipple sign as a definite stigma of recent variceal hemorrhage. It is likely that endoscopists are not as familiar with this sign as they are with red color signs. As Chung and Lewis [4] initially suggested, it should ‘alert the endoscopist to take urgent measures to avert a disaster’. Recognizing it as such, attempts to dislodge the lesion must be avoided and endoscopic therapy should be undertaken.

Fig. 1. Endoscopic image of the platelet-fibrin plug protruding into the esophageal lumen from an esophageal varix. It is also referred to as the ‘white nipple sign’.

Fig. 2. Endoscopic image of the brisk hemorrhage that resulted after inadvertent dislodgement of the platelet-fibrin plug.

Fig. 3. Endoscopic image displaying successful control of variceal hemorrhage after endoscopic variceal band ligation.

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