Case report

A 29-year-old multigravid woman at 33 5/7 weeks of gestation presented with shortness of breath and abdominal pain. She was an outpatient in the gastrointestinal department due to recurrent jaundice and had a history of the Kasai procedure.

On clinical examination, the patient was drowsy and had an oxygen saturation level of 89% on 15 L/min of oxygen. Chest radiography revealed total opacification of the right hemithorax with a right-to-left mediastinal shift (Fig. 1). Thoracentesis was performed, and 50 mL was drained from the hemothorax. The non-stress test simultaneously showed late deceleration with a fetal heart rate of 60 beats/min; thus, an emergency cesarean section was performed. Chest and abdominal computed tomography were performed immediately after the delivery. The right hemothorax was assumed to be secondary to bleeding at the variceal dilatation of the right inferior phrenic vein, with extravasation around the right hemidiaphragm (Fig. 2).

For bedside closed thoracostomy, a 16F trocar catheter was inserted without complications, and 900 mL of blood was confirmed in the chest cavity. However, the chest radiograph showed no change in the right total opacification. Therefore, an emergency thoracotomy was performed for hematoma evacuation and active bleeding control.

Approximately 2,500 mL of unclotted blood and hematoma were found in the postero-inferior pleural cavity. A thin (presumed diameter, 5–8 mm), dilated, engorged phrenic vein on the anterolateral diaphragmatic surface and a ruptured distal phrenic vein were found in the surgical field (Fig. 3). The bleeding vein was clamped with Kelly...
forceps, and interrupted sutures were subsequently performed with a small Teflon (Dacron) pledget (4-0 Prolene). After applying surgical materials, including TachoComb (CSL Behring, Tokyo, Japan), Surgicel (Johnson & Johnson, Wokingham, UK), and Neoveil (Gunze, Kyoto, Japan), a 24F thoracic catheter was inserted.

The patient had an excellent recovery and was discharged on postoperative day 7. On short-term follow-up, chest radiography findings had improved to slight right costophrenic angle blunting.

The approval by the institutional review board (IRB) of Inje University Busan Paik Hospital was obtained (IRB file no., 2021-12-091), and exemption of consent was accepted.

Discussion

Hemothorax is defined as a collection of fluid in the pleural space with a hematocrit level >50% of the serum hematocrit. It is mostly caused by trauma, neoplasm, coagulopathy, or iatrogenic factors.

Spontaneous hemothorax due to sudden vascular rupture, including phrenic vein varices, is far less common [1]. In this case, the patient had undergone the Kasai procedure due to biliary atresia and multiple episodes of cholangitis at 1 year of age. The patient’s engorged phrenic vein seen in the surgical field led us to focus on ectopic variceal hemorrhage, a complication of portal hypertension that can result from chronic hepatobiliary disease.

The Kasai procedure, hepatico-portoenterostomy, is a first-line treatment for biliary atresia to restore bile flow. A systematic review found that 60.5% of patients who underwent the Kasai procedure during early infancy eventually experienced progressive liver-related complications [2]. One study reported that esophageal varices after a successful Kasai procedure developed in one-fourth of patients, and half of these progressed to hemorrhages [3]. Varices associated with portal hypertension most often occur in the esophagus and stomach. Ectopic varices, which frequently happen in the duodenum, jejenum, ileum, colon, rectum, and peritoneum, represent up to 5% of cases of variceal bleeding [4,5]. This underscores the rarity of phrenic vein varix rupture.

Our patient presumably had varices for a lifetime, and the rupture may have been caused by the increased blood volume [6] and intra-abdominal pressure during pregnancy [7].

The management of spontaneous hemothorax is the same as that of typical hemothorax; the first-line treatment involves closed thoracostomy and drainage. If the drained blood volume exceeds expectations (>1,500 mL initially or 200 mL/hr), as in our patient, hemostatic surgery must be considered.

To the best of our knowledge, this is the first reported case of a massive spontaneous hemothorax that affected vital signs due to ectopic variceal hemorrhage. Although spontaneous hemothorax is rare, a precise diagnosis and appropriate treatment are important. Early surgical management, when applicable, is also critical.

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Conflict of interest

No potential conflict of interest relevant to this article was reported.

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