Transient Budd-Chiari syndrome as an unpredictable complication of supradiaphragmatic inferior vena cava reconstruction after blunt thoracic trauma

A case report
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Abstract

Rationale: Supradiaphragmatic inferior vena cava (IVC) injury due to blunt thoracic trauma is extremely rare. Budd-Chiari syndrome (BCS) is also rare and presents with ascites, abdominal pain, hepatomegaly, leg swelling, and jaundice. Its etiology is diverse, and it is rarely caused by trauma.

Patient concerns: A 36-year-old man with blunt trauma from a traffic accident presented with chest pain. Chest computed (CT) and emergency surgery with CPB revealed completely transected supradiaphragmatic inferior vena cava (IVC), which reconstruction was essential.

Diagnoses: BCS caused by impaired hepatic venous drainage through a reconstructed neo-IVC after severe blunt trauma injury to the supradiaphragmatic IVC was diagnosed.

Intervention: Hepatic failure, ascites, leg swelling, and jaundice were resolved post-insertion of a veno-venous extracorporeal membrane oxygenator (V-V ECMO) for hepatic venous drainage, but these clinical symptoms reappeared after ECMO removal.

Outcome: The patient died from rapidly progressing sepsis, pneumonia, and acute renal failure during repeated insertion of ECMO and weaning off ECMO.

Lessons: Reconstructing and improving the patency of the supradiaphragmatic IVC is essential for successful hepatic venous drainage. Additionally, a surgical strategy focused on graft selection can prevent kinking stenosis, and possibly BCS, especially in emergency surgeries. A ring-supported synthetic graft should be considered an alternative to improve long-term patency and survival rate.

Abbreviations: ALT = alanine aminotransferase, AST = aspartate aminotransferase, BCS = Budd-Chiari syndrome, CPB = cardiopulmonary bypass, CRRT = continuous renal replacement therapy, CT = computed tomography, DM = diabetes mellitus, ECMO = extracorporeal membrane oxygenator, HCA = hypothermic circulatory arrest, IVC = inferior vena cava, RA = right atrium, SVC = superior vena cava, TTE = transthoracic echocardiogram, V-V ECMO = veno-venous extracorporeal membrane oxygenator.

Keywords: Budd-Chiari syndrome, inferior vena cava transection, ring-supported synthetic graft, supradiaphragmatic IVC

1. Introduction

The location of inferior vena cava (IVC) injury due to trauma is one of the most important factors that influence mortality. Particularly, suprahepatic IVC injuries and supradiaphragmatic IVC injuries caused by blunt trauma although extremely rare, are fatal because of massive bleeding and the difficulty associated with its repair.\textsuperscript{1} Emergent surgical management including primary repair, patch repair, or reconstruction with various grafts, is used to manage IVC injuries, but the optimal strategy for surgery remains controversial.\textsuperscript{2,3} Immediate reconstruction of IVC using a graft is required to restore hepatic venous drainage and survival in cases of severe supradiaphragmatic IVC injury such as a transection. Prevention of complications, graft infection, or thrombosis after reconstruction is important to improve patency and survival rate.\textsuperscript{4} In the present case, Budd-Chiari syndrome (BCS), which may result from impaired hepatic venous drainage, developed without evidence of mechanical graft failure, such as infection or thrombosis, after successful reconstruction surgery.
with a pulse rate of 106 beats/min. The patient had an unremarkable medical history except for diabetes mellitus (DM). He complained of chest pain alone. Physical findings were normal except for a 2 x 2-cm sized focal abrasion on the sternum.

Chest radiography showed an enlarged cardiac silhouette. Chest computed tomography (CT) showed pericardial effusion and multiple rib fractures with no other organ injuries or fluid collection. A lesion on IVC could be seen on the preoperative abdominal CT scan, and the infradiaphragmatic IVC and no other organ were damaged. (Fig. 1) Transthoracic echocardiogram (TTE) revealed pericardial effusion, IVC plethora without inspiratory or expiratory changes in diameter, right atrium (RA) systolic collapse, and inversion of the RA free wall.

Emergency median sternotomy was performed. The pericardium was intact with no laceration. Pericardiotomy revealed a large hematoma in front of RA and around IVC. Massive hemorrhage from the infracardiac IVC area started immediately after the removal of the hematoma overlying IVC. The hemorrhage was stopped using gauze packing and digital compression. Compression was maintained until cardiopulmonary bypass (CPB) could be performed. A cannula was inserted into the ascending aorta for arterial perfusion, and 2 venous cannulas were required for CPB. One cannula was inserted into the superior vena cava (SVC) and the other was inserted into the left femoral vein. Once CPB had been established, blood loss reduced, and a clear view of the area was secured.

The IVC wall between the heart and diaphragm, including a part of RA, was absent. IVC was almost completely transected at the caval opening in the diaphragm; only ragged, retracted IVC remnant tissue was present, which was unsuitable for anastomosis. A hole was noted at the atrio caval junction and was lacerated towards the coronary sinus. Anastomosis using the existing hole was impossible. We made a new entrance to RA with a clean incision and reinforced it with multiple pledgeted prolene 4-0 sutures to make the edge durable. The caval opening in the diaphragm was also reconstructed by suturing the diaphragm and the remaining shredded tissue using multiple pledgeted prolene 4-0 sutures.

The lost IVC segment and part of the RA wall were successfully reconstructed using a 28-mm Gelweave graft (Vascutek Ltd., Inchinnan, Scotland, UK), which was very short at approximately 4 cm, under hypothermic circulatory arrest (HCA) and CPB. A 44-minute period of HCA was essential to secure a clear view for distal anastomosis of the new IVC. The patient was easily weaned from CPB and diffuse bleeding was corrected by the administration of blood products, including 16 units of red blood cells, 22 fresh frozen plasma units, and 3 platelet concentrate units, intraoperatively.

The patient was transferred to the ICU and the immediate postoperative course was uneventful. On postoperative day 3, hypoxemia, hypotension, and hepatic insufficiency developed. The patient was hemodynamically unstable with no response to fluid loading or to the administration of inotropic agents or vasopressors. Aspartate aminotransferase (AST) increased to 16,880 U/L, and alanine aminotransferase (ALT) increased to 4273 U/L. Extracorporeal membrane oxygenator (ECMO) was established immediately, and the patient became hemodynamically stable. AST and ALT decreased to 12,094 U/L and 3216 U/L, approximately 12 hours after ECMO insertion and gradually, they nearly normalized to 70 U/L and 48 U/L, respectively.

Coagulation ability normalized in 7 days. Weaning off ECMO was uneventful. However, on the 3rd day after ECMO removal, hypoxemia and hypotension reappeared despite full ventilator support and the administration of inotropic agents and vasopressors. V-V ECMO was re-established, and the patient became hemodynamically stable.

The cause of hepatic failure was uncertain. Leg swelling and severe jaundice developed. Limited examination could be performed due to the marginal hemodynamic condition, but a CT scan and TTE images were reviewed before ECMO insertion. These revealed diffuse ascites (Fig. 2) and mottling in the liver (Fig. 3) without thrombi or obstacles throughout the length of
IVC, and no external compressive structures were noted around the graft. TTE showed narrowing of IVC, sudden flow acceleration through the short graft segment, but no obstruction was noted in IVC. However, hepatic failure including jaundice, leg swelling, ascites, and mottling in the liver corresponded to BCS. This complication was responsive to V-V ECMO insertion alone, that is, by securing hepatic venous drainage outflow.

The patient’s condition worsened due to rapidly progressing sepsis and acute renal failure after the reestablishment of ECMO, and the hepatic failure showed no response to treatment, including antibiotics, vasopressors, inotropes, continuous renal replacement therapy (CRRT), and ECMO. The patient died on the 26th postoperative day after withdrawal of life support.

The Human Research Ethics committee of Yeungnam University Hospital waived the requirement of ethical approval, as this is a single case report. The patient provided written informed consent for publication of the clinical details and images pertinent to this case.

3. Discussion

This report describes a severe complication, Budd-Chiari syndrome, after successful IVC reconstruction using a synthetic graft. Budd-Chiari syndrome is a rare condition that is associated with hepatic failure, showing hepatomegaly, ascites, jaundice, mottling in the liver, swelling of both legs, and abdominal pain. This disastrous syndrome is due to impaired hepatic venous drainage and congestion and is fatal, if left untreated. BCS has multiple causes including myeloproliferative disorders, malignancy, infection, benign hepatic lesions, pregnancy, thrombophilia, toxins, Behcet syndrome, and chronic inflammatory disease; it is rarely caused by trauma.[5]

Only a few case reports have documented posttraumatic BCS, involving 2 different disruptions to hepatic venous drainage: intravascular obstruction and extravascular compression. In a report, IVC thrombosis due to endothelial injury led to BCS and the patient was treated successfully via thrombolysis and angioplasty.[6] Another published report described extrinsic compression of IVC due to obstacles around the vessel. The patient in this report was treated by the removal of the surrounding hematoma, which was compressing the vessel.[7]

In the present case, no thrombus was detected inside the vessel, and no other structures were compressing the vessel externally. However, the patient developed ascites, transaminitis, and lower extremity edema. Contrast CT scan of the abdomen showed hepatomegaly, mottling in the liver, and diffuse ascites, which is consistent with BCS diagnosis. TTE showed that the replaced graft was patent with focal narrowing, and that flow was accelerated only through this segment. We reached a tentative conclusion that the cause of BCS was kinking stenosis or twisting of the reconstructed graft.

The choice of an ideal graft for IVC reconstruction remains controversial. Currently, 2 kinds of grafts are used for IVC reconstruction. One is composed of biologic materials including
autologous pericardium and porcine and bovine pericardia; the other employs synthetic materials such as expanded polytetrafluoroethylene. 

Biological graft material allows ease of handling, long-term patency without coagulation, and lower rate of graft infection. Use of an autologous pericardium demonstrates better fibrinolytic activity, less endothelial lining contraction, and subendothelial fibrosis. Using an autologous pericardium results in low thrombogenicity and a lower risk of narrowing due to contraction. Endothelialized autologous grafts can be an optimal choice for a low-pressure and low-flow large vessel, such as the IVC. However, one crucial disadvantage is its availability. An intact autologous pericardium may not be available in cases of trauma and may be more time-consuming, which can be critical in emergency situations.

Despite disadvantages, such as the need for life-long anticoagulant therapy and the risk of infection, synthetic grafts are more commonly used, as they are more readily available, and in various diameters and lengths. Cumulative 5-year patency for IVC graft replacement was good, and was reported to be between 89% and 100%. The critical merit of synthetic materials is less time-consumption, which is essential, especially in emergency surgeries.

In the present case, a synthetic graft was employed. Kinking of the graft resulted in impaired hepatic venous drainage and BCS. Externally, a ring-supported synthetic graft can avoid graft collapse and kinking stenosis. Early diagnosis and early interventions, such as angioplasty using an endovascular stent, can prevent severe complications.

This extremely rare and severe IVC injury requires rapid and appropriate surgical therapy. However, the ideal strategy for emergency surgery remains controversial due to the rarity of the condition. Rapid and accurate preoperative diagnosis is essential, and HCA and CPB should be prepared for severe IVC injuries. Reconstruction using an optimal graft should be considered. Moreover, a ringed graft is a synthetic graft with benefits such as availability and good patency. Although, synthetic grafts are not optimal for IVC reconstruction, ring-supported synthetic graft can be the first choice for supradiaphragmatic IVC reconstruction in emergency situations. We speculate that good patency of the reconstructed IVC can prevent complications like BCS. However, graft failure can also cause BCS. Altogether, there is a need to research and improve pre-hospital systems and surgical techniques to successfully manage such cases.

Author contributions

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