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CASE REPORT | ENDOSCOPY

Duodenocaval Fistula: Unmasking the Rare Culprit of Gastrointestinal Hemorrhage

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ABSTRACT

Duodenocaval fistula is an extremely rare and life-threatening cause of gastrointestinal hemorrhage and septicemia. Diagnosing this condition is challenging due to its nonspecific symptoms, leading to significant delays in diagnosis and contributing to its remarkably high mortality rate. We present a unique case of duodenocaval fistula associated with prior radiation, peptic ulcer disease, and antiangiogenic cancer therapy, nearly resulting in the death of a young patient.

KEYWORDS: Duodenocaval Fistula; Duodeno-caval fistula; Gastrointestinal Bleeding; Gastrointestinal Hemorrhage; Hematemesis; Upper gastrointestinal bleeding

INTRODUCTION

Duodenocaval fistula (DCF) is a rare pathologic connection between the inferior vena cava (IVC) and duodenum. Despite identification in 1974, few cases exist.[1] Symptoms are often nonspecific, including fever, septic or hypovolemic shock, and gastrointestinal bleeding. With a mortality rate up to 40%, the elusive nature of DCF frequently leads to devastating outcomes unveiled only during emergent laparotomy or autopsy.[2] Computed tomography only identifies DCF in approximately 50% of cases.[2,3] Endoscopy may reveal duodenal ulcers, but the depth of penetration can be underestimated.[2] Timely surgical intervention is imperative upon diagnosis to circumvent high mortality rates. We present a case of DCF manifesting as gastrointestinal hemorrhage and sepsis in a young patient that highlights the importance of early recognition of DCF and sheds light on associated risk factors.

CASE REPORT

An 18-year-old man presented with 5 days of fever and 1 day of hematemesis. History was notable for metastatic osteosarcoma of the femur with spinal metastases diagnosed 2 years prior. He underwent chemotherapy, partial surgical resection of the femur and first lumbar spine (L1), and radiation to L1. Alongside maintenance oral chemotherapy with lenvatinib, he was taking high-dose nonsteroidal anti-inflammatory drugs (NSAIDs).

On presentation, he was febrile and tachycardic. Hemoglobin (Hgb) was 10.8 g/dL (reference range, 13.3–17.3 g/dL), and blood cultures grew *Lactobacillus rhamnosus*. Intravenous pantoprazole and meropenem were initiated. Esophagogastroduodenoscopy showed multiple Forrest III duodenal ulcers and a large 2–3 cm Forrest IIb ulcer with contained perforation at the junction of the second and third portions of the duodenum. Endoscopic interventions were deferred given inactive bleeding, and empiric gastroduodenal artery/coil embolization was performed.

Ten days later, he redeveloped hematemesis and Hgb declined to 6.8 g/dL. Computed tomography angiography revealed near-occlusive thrombus with scattered foci of gas in the IVC, suggesting a fistulous connection to the duodenum due to the proximity of the ulcerated IVC (Figure 1). Repeat endoscopy demonstrated the Forrest IIb ulcer squirting blood from the ulcer base (Figure 1). Endoscopic hemostasis with bipolar cauterization and hemostatic clip placement was unsuccessful. During the procedure, he developed

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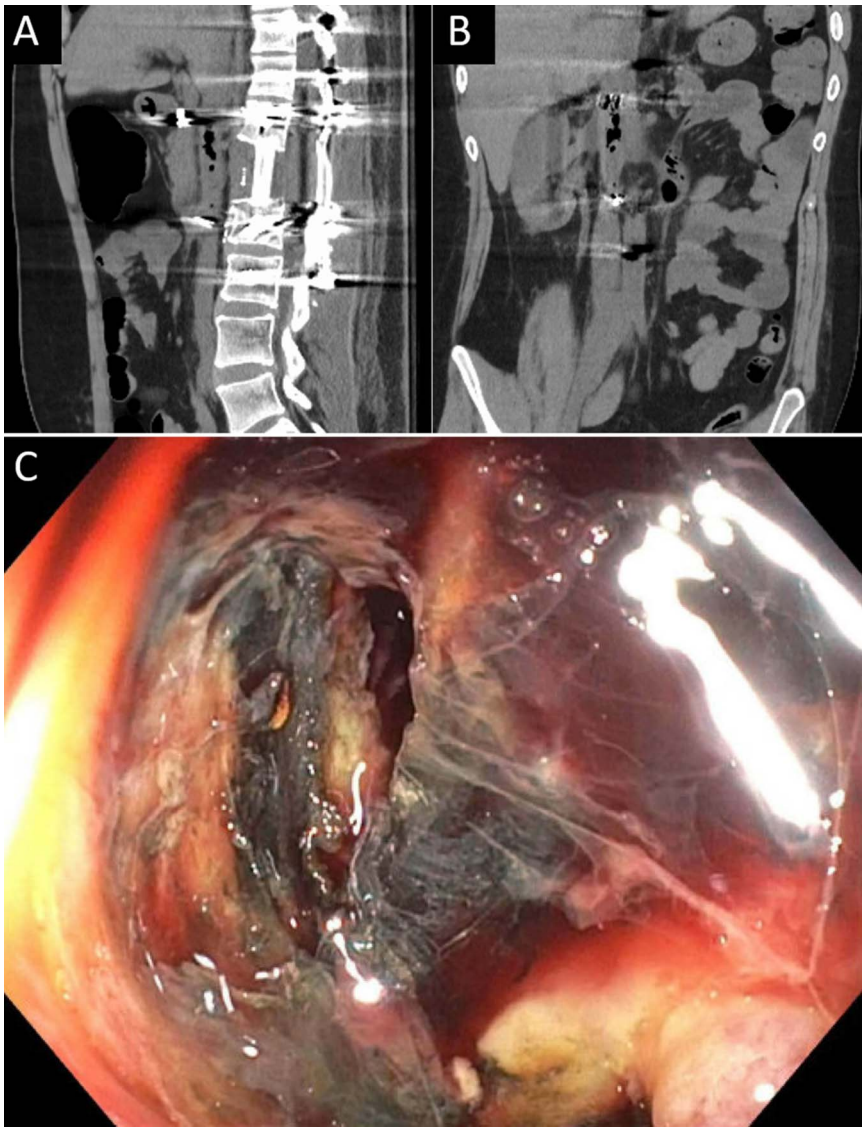

Figure 1. (A and B) Computed tomography imaging demonstrates multiple foci of gas within the inferior vena cava extending 6.8 cm craniocaudally from the level of the third portion of the duodenum to the level of gastroduodenal artery embolization consistent with duodenocaval fistula in sagittal (A) and coronal (B) views. (C) Endoscopic view of Forrest IIb ulcer with a suspected contained perforated ulcer base located at the junction of the second and third portions of the duodenum.

hemorrhagic shock requiring blood transfusions. Vascular surgery and general surgery were consulted. However, given DCF is exceedingly rare, surgical specialties thought septic phlebitis without concomitant DCF was more likely. Nonoperative management was favored over exploratory laparotomy given the elevated risk due to prior radiation and surgery.

His condition stabilized temporarily until developing repeat large-volume hematemesis with worsening hemodynamic instability a week later. Emergent esophagogastroduodenoscopy performed showed the Forrest IIb ulcer with fresh blood clots and active oozing. Attempts at hemostasis remained unsuccessful requiring emergent laparotomy, which confirmed DCF and suppurative IVC thrombophlebitis. Vascular surgeons performed an IVC thrombectomy and reconstruction with bovine patch angioplasty while general surgeons repaired the perforated ulcer and performed a Roux-en-Y. The IVC clot grew *Lactobacillus*. Overall, he was hospitalized for 35 days and received 24 units of blood. Postoperative course was complicated only by self-limited delayed gastric emptying requiring brief nasogastric tube feeding. Repeat endoscopy a month later revealed a healing ulcer a quarter of its original size; bleeding did not recur.

DISCUSSION

DCF is commonly seen in middle-aged men in association with traumatic injuries including penetrating abdominal traumas, IVC filter migration, and foreign body ingestion.[4] Remarkably, this patient was the youngest ever diagnosed with DCF, likely

playing a role in his survival. This patient's etiology of DCF appears multifactorial, including prior retroperitoneal radiation, peptic ulcer disease (PUD), and vascular endothelial growth factor (VEGF) inhibitor therapy.

Associations between DCF and retroperitoneal cancers have been documented.[5,6] His history of metastatic osteosarcoma to L1 likely contributed to his development of DCF. Surgical resection of L1 was followed by local radiation, aligning with the location of the duodenum in respect to the spine (L1-L3). Postradiation mucosal damage and fibrosis, leading to adherence to the IVC and duodenum, are identified as potential contributors to the ulceration and fistula formation seen in DCF.[5] A prior literature review found the time between radiation and initial presentation for DCF to be on average 32 months, while this patient presented within 7 months.[7]

PUD is implicated in nearly 20% of DCF cases, with over 60% of these proving lethal.[4] Literature suggests PUD-associated DCF cases typically exhibit symptoms for 1–4 months prior and occur with large ulcers ranging in size from 1.5 to 9 cm.[4,8]

While 4 reported cases associate DCF with the antiangiogenic bevacizumab, this case marks the first associated with lenvatinib.[4,9–11] These 2 drugs share similar mechanisms, with bevacizumab directly inhibiting VEGF and lenvatinib indirectly inhibiting VEGF through its tyrosine kinase receptor. The SELECT trial demonstrated 1.5% of patients on lenvatinib developed a gastrointestinal fistula or perforation.[12] The proposed mechanism includes platelet-endothelial cell homeostasis disruption with resultant submucosal inflammation/ulceration, diminished healing capacity, and thrombosis and/or vasoconstriction-related mesenteric ischemia.[11] Lenvatinib was continued for 2 weeks before being identified as a potential contributing factor.

This case highlights the need for a high index of suspicion for DCF and consideration for preventative strategies in at-risk patients. Risk factors in this patient included radiation at the level where the IVC and duodenum are situated adjacent to each other, antiangiogenic therapy associated with fistula formation, and NSAID use contributing to PUD. In similar patients, the use of proton pump inhibitors or H2 blockers empirically while on lenvatinib and avoidance of NSAIDs may lower the risk of DCF. Findings of duodenal ulceration with contained perforation of the second and third portions of the duodenum on endoscopy and air within the IVC on imaging should raise suspicion for DCF. Although gastroduodenal artery embolization and endoscopic interventions were attempted in this case, definitive management of DCF nearly always requires surgical intervention, which when performed early, has been associated with improved outcomes. Surgical management typically involves division of the fistulous connection, repair of the IVC and duodenum, and a diverting procedure such as a Roux-en-Y as was seen in this patient.[13] Despite its rarity, the consequences of delayed intervention can be severe, emphasizing the need for increased awareness and proactive management of suspected DCF.

DISCLOSURES

Author contributions: A. Fiedler is responsible for the overall composition, edits, and submission and is the article guarantor. B. Dhindsa critically revised the manuscript for important intellectual content. S. Singh was directly involved in the care of the patient and helped manage the clinical case. The authors reviewed and approved the final version of the manuscript.

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