Primary pancreatic lymphoma: A rare cause of massive upper gastrointestinal hemorrhage

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CASE PRESENTATION

A 68-year-old man presented with abdominal pain radiating to the back associated with a 25 kg weight loss. Abdominal ultrasound and computed tomography (CT) revealed an 8 cm partially cystic mass in the pancreatic head that encased the aorta and inferior vena cava (IVC) (Figure 1). A CT-guided fine-needle aspiration (FNA) was performed and confirmed a diagnosis of diffuse large B cell lymphoma. Subsequent staging CT demonstrated peripancreatic lymphadenopathy and air within the tumour, suggesting a possible communication between the intestine and the pancreatic mass. Direct visualization with gastroscopy confirmed a large penetrating ulcer between the second portion of the duodenum and the pancreatic mass. Chemotherapy was delayed while pyloric exclusion surgery was performed to reduce the risk of chemotherapy-related uncontained perforation of the duodenum. The postoperative clinical course was complicated by a slowly declining hemoglobin level, followed by cardiopulmonary arrest in concert with a sudden drop in hemoglobin level to 50 g/L. An emergent upper endoscopy revealed copious amounts of blood within the stomach. The grave clinical deterioration precluded further resuscitative efforts, and the patient died from a massive upper gastrointestinal hemorrhage.

A postmortem examination confirmed an 8 cm, extensively necrotic mass in the head of the pancreas. Significant peripancreatic lymphadenopathy was present; however, all nonregional lymph nodes were of normal size. The pancreatic head mass extended laterally and eroded into the adjacent duodenal wall producing the large mucosal erosion identified endoscopically. Posteriorly, the mass encased the aorta and the IVC, producing an irregular ulceration of the endoluminal surface of the IVC (Figure 3). This ulceration of the IVC resulted in a fistulous tract between the IVC and the duodenum via an intermediary of the pancreatic head mass. Histological examination revealed a poorly differentiated malignant neoplasm, which by immunohistochemical studies confirmed the original diagnosis of a diffuse large B cell lymphoma.

DISCUSSION

Primary pancreatic lymphomas (PPLs) are rare, accounting for less than 1.5% of primary pancreatic malignant neoplasms (1). Secondary pancreatic involvement of systemic non-Hodgkins’ lymphoma is far more common (2). The typical clinical presentation of a PPL is characterized by nonspecific symptoms that mimic pancreatic adenocarcinoma: painless jaundice, weight loss, nausea and vomiting, and abdominal/back discomfort. The diagnosis can only be confirmed by tissue sampling because imaging is usually nonspecific, identifying a pancreatic head mass with peripancreatic adenopathy in most cases. Minimally invasive tissue sampling techniques, such as endoscopic ultrasound-guided FNA and CT-guided FNA, have good test performance characteristics and are emerging as the methods of choice in the evaluation of pancreatic masses (3). These modalities play an important role in the triage of patients toward appropriate medical or surgical treatment. However, surgery may still be required when these minimally invasive diagnostic techniques fail to yield a definitive diagnosis. Medical management with chemotherapy is the mainstay of treatment for PPL. The role of surgical intervention beyond tissue biopsy for diagnosis and palliation remains controversial (4).

Upper gastrointestinal hemorrhages have a wide spectrum of presentation and etiologies. Relatively uncommon entities that can lead to massive upper gastrointestinal hemorrhage include anomalous communications between the luminal gastrointestinal tract and nearby...
vessels, examples of which include aortoenteric (5) and duodenocaval (6) fistulas. Vascular-enteric communications can occur as complications of previous medical and surgical interventions including surgical repair of abdominal aortic aneurysms, retroperitoneal irradiation and endovascular filter migration (7). Rare reports describe vascular-enteric communications occurring secondary to natural disease process (8,9).

Fewer than 40 cases of duodenocaval fistulas have been reported in the literature, with an estimated mortality rate of 40%. The presenting signs and symptoms are variable, ranging from weight loss, abdominal pain and fever of unknown origin, to life-threatening sepsis and fatal gastrointestinal hemorrhage. An unusual presentation of pulmonary embolization of intestinal contents has also been reported (9). Recognized risk factors leading to the development of duodenocaval fistulas include trauma, ingested foreign bodies, peptic duodenal ulceration, irradiation, migrating IVC filter and resection of retroperitoneal tumours (eg, renal cell carcinoma and sarcomas). The present report is the first to describe a duodenocaval fistula in the context of pancreatic malignancy or as a complication of untreated malignancy.

REFERENCES