Abdominal apoplexy, or idiopathic spontaneous intraperitoneal hemorrhage, is a rare and often fatal condition resulting from a variety of disease processes affecting the arterial and venous abdominal vasculature. Preoperative and intraoperative diagnosis and treatment of abdominal apoplexy are challenging. The source of bleeding may remain elusive even after careful autopsy dissection given the absence of intravascular pressure. Despite these challenges, early diagnosis and rapid treatment remain central to a successful outcome, as nonsurgical mortality has approached 100%. Presented here are two fatal cases of abdominal apoplexy, one involving a patient with arterial dissection of the gastroduodenal artery and one involving rupture of the superior mesenteric-portal venous system with perivascular pseudoaneurysm formation.

Abdominal apoplexy, or idiopathic spontaneous intraperitoneal hemorrhage (ISIH), is rare and often fatal. This condition results from a variety of disorders affecting the arterial and venous abdominal vasculature, and nonspecific symptoms usually include abdominal pain, nausea, vomiting, and hemodynamic instability (1). Immediate exploratory laparotomy is the treatment of choice (2), and rapid surgical intervention remains central to a successful outcome. However, accurate preoperative and intraoperative diagnosis remains difficult. Although rare in occurrence, ISIH constitutes a true emergency and should be considered in any patient with atypical abdominal pain and hemodynamic instability (3, 4).

**CASE 1**

A 70-year-old Caucasian woman was transported to Baylor University Medical Center after complaining of nausea and weakness and collapsing at home. She had recently been hospitalized for treatment of a urinary tract infection, acute renal injury, and a flare of gout, and she had been discharged for outpatient therapy. En route to the hospital, she developed cardiac arrest and required cardiopulmonary resuscitation. Upon arrival at the emergency department, a heart beat had been reestablished, but she was severely hypotensive. Her hemoglobin was 7.1 g/dL, representing a decrease of almost 50% compared to her admission less than a week earlier (13.7 g/dL). Her lipase was 519 U/L. A noncontrast computed tomography (CT) scan of the chest, abdomen, and pelvis was interpreted as possible acute hemorrhagic pancreatitis. The superior mesenteric artery and superior mesenteric vein were completely engulfed by “inflammatory stranding.” The integrity of these vessels was in question but could not be adequately evaluated without contrast. There was apparent inflammation involving the distal stomach and duodenum as well (Figures 1 and 2). She was converted to a do-not-resuscitate status, and surgery was not performed. Despite supportive measures, the patient’s status declined, and life support was eventually withdrawn.

At autopsy, there was approximately 700 mL of liquid and clotted blood within the abdominal cavity in addition to extensive retroperitoneal and mesenteric hemorrhage, but an obvious source of hemorrhage was not initially apparent. After fixation, the pancreas, mesentery, regional vasculature, duodenum, and common bile duct structures were again carefully examined and dissected. There was extensive intraparenchymal and peripancreatic hemorrhage; however, inflammatory findings were not present. Surrounding the superior mesenteric artery and smaller...
arterial branches, as well as the superior mesenteric vein and venous branches, there was densely clotted blood. Within areas of diffuse hemorrhage surrounding the gastroduodenal artery and its branches, there was a grossly apparent perivascular space that contained a blood clot circumscribed by surrounding connective tissue planes (Figure 3).

Microscopically, there was extensive hemorrhage surrounding the pancreas; however, there were no changes of pancreatitis involving the parenchyma. Within the media of the gastroduodenal artery, there was a blood-filled, cleft-like disruption, typical of spontaneous arterial dissection (Figure 4). Movat pentachrome staining highlighted the lesion and revealed disruption of the internal elastic lamina with central compression of the vascular lumen by the dissecting blood (Figure 5). There was extensive hemorrhage surrounding the vessel, and there was no histopathologic evidence of a specific vasculitis or vasculopathy. Additional autopsy findings included significant hypertensive cardiovascular disease and severe aortic, coronary artery, and renal artery atherosclerosis.

CASE 2

A 55-year-old Caucasian woman presented to Baylor University Medical Center with acute-onset abdominal pain, diarrhea, nausea, vomiting, and dizziness. Her physical examination revealed diffuse abdominal tenderness, and her hemoglobin was 7.5 g/dL. A CT scan of the abdomen revealed a large hemoperitoneum, predominantly in the lesser sac. There were diffuse inflammatory changes about the head of the pancreas and porta hepatis, along with a circumscribed hematoma near the junction of the superior mesenteric vein and the portal vein (Figures 6 and 7). She was taken emergently for exploratory laparotomy, where a large hemoperitoneum was confirmed. The portal vein, mesenteric vein, and surrounding tissues were extremely friable, and vascular ligation was attempted at sites of possible rupture. Postoperatively, the patient required pressors and numerous units of blood; however, supportive measures were ultimately unsuccessful and she died. An autopsy was requested.

At autopsy, 5100 mL of liquid and clotted blood was found within the abdominal cavity, and there was a 4-cm circumscribed thrombus within the central abdominal region. Again,
a definite source of bleeding was not initially apparent. After fixation and careful dissection of the vasculature, the thrombus was found to be located within connective tissues adjacent to the head of the pancreas and external to the superior mesenteric and portal veins (Figure 8). This thrombus was enclosed within a pseudoaneurysm wall composed of friable connective tissue with recently placed surgical sutures for attempted hemostasis. A true vascular aneurysm wall was not identified at autopsy; however, sutures at the superior mesenteric vein–portal vein junction were consistent with a repaired venous disruption. Microscopic sections showed no evidence of intrinsic vasculitis or specific vasculopathy involving the superior mesenteric or portal veins. There was focal intramural and perivascular hemorrhage with some thinning of the portal vein, but a true aneurysm was not identified. Furthermore, there was no evidence of intrinsic pancreatitis or hepatic cirrhosis, and there were no historical or anatomic findings to support recent abdominal trauma or hypertensive cardiovascular disease. There was no atherosclerotic cardiovascular disease.

These findings were interpreted as consistent with spontaneous intraabdominal hemorrhage associated with idiopathic venous rupture and perivascular pseudoaneurysm formation.

DISCUSSION

In general, intraperitoneal or retroperitoneal hemorrhage may be secondary to blunt trauma, aneurysmal rupture (central or visceral), solid organ malignancy (hepatic or renal), or inflammatory erosive processes (pancreatitis or pseudocyst); however, it may be idiopathic as well (2, 3, 5).

Abdominal apoplexy, or the newer term, ISIH, describes a rare finding of nontraumatic intraabdominal bleeding. ISIH was first reported by Barber in 1909, but the term “abdominal apoplexy” was coined by Green and Powers in 1931 as a comparison to its cerebral counterpart (3–5). Traditionally, abdominal apoplexy refers to spontaneous hemorrhage arising from one of the smaller abdominal arteries or veins, after hemorrhage from a grossly apparent aortic aneurysm or aortic dissection is excluded. Additionally, cases of hemorrhage from visceral malignancy and gynecologic lesions such as ectopic pregnancy, as well as abdominal hemorrhage associated with known traumatic injury, are also excluded (6). Thus defined, abdominal apoplexy is exceedingly rare. There is a male predominance (2–3:1), and the majority of cases present in the fifth and sixth decades of life (3, 6).

Abdominal small vessel rupture often occurs at the site of an aneurysm, but up to 30% of cases have no identifiable source (3, 5). Historically, aneurysms have been mycotic, syphilitic, or traumatic in origin but are now more likely related to essential or portal hypertension (2, 3). Fibromuscular dysplasia has also been associated with aneurysm formation. In the absence of portal hypertension, known trauma, or fibromuscular dysplasia, a specific etiology of venous disruption remains elusive. Predictably, arterial aneurysms often occur at secondary or tertiary branch points from the aorta; 60% involve the splenic artery, 22% renal, and 10% to 20% hepatic, with common celiac and mesenteric arteries less common (3). Most cases with no identifiable source are probably related to common vascular diseases including arteriosclerosis and essential hypertension.
The exact mechanism is unknown but likely represents weakness of the tunica media, predisposing to rupture in the face of abrupt increases in pressure. Pathology specimens regularly exhibit disruption of elastic lamellae (3, 5). An aneurysmic stage does not necessarily precede the spontaneous rupture of a visceral artery (4). Less frequently, spontaneous hemorrhage may be associated with inflammatory and necrotizing processes such as polyarteritis nodosa and rheumatoid arthritis, etc. (3, 4).

Venous rupture, on the other hand, is usually associated with portal hypertension due to hepatic cirrhosis (7).

In addition to the above disorders, rare cases of abdominal apoplexy have been attributed to arterial dissections involving splanchnic vessels such as the gastroduodenal, hepatic, superior mesenteric, gastric, and splenic arteries (1). Various theories regarding risk factors for arterial dissection include common disorders such as essential hypertension and less common connective tissue disorders (1) such as the Marfan and Ehlers Danlos syndromes.

These two cases demonstrate the continued therapeutic and diagnostic challenges associated with the clinical management of patients with abdominal apoplexy. Angiography was not clinically feasible in either case. The limitations of noncontrast CT prevented exclusion of pancreatitis as well as identification of a specific bleeding source. Furthermore, the source of bleeding was not obvious upon initial autopsy examination, and accurate diagnosis required careful preservation of the specimens with subsequent dissection and microscopic sampling before the source of bleeding was localized. These cases also demonstrate the wide spectrum of arterial and venous abnormalities that can result in spontaneous abdominal hemorrhage, and the case involving venous rupture is especially unusual since this patient did not have cirrhosis or portal hypertension. Although autopsy examination demonstrated the source of bleeding in both cases, a specific underlying cause for development was not apparent.

Both cases also demonstrate our currently imperfect understanding of abdominal apoplexy, and further research is certainly required. Although this disorder has been called idiopathic and spontaneous, it is unlikely that such hemorrhage occurs without an underlying vascular lesion (7), which may be apparent only at the molecular level.