

Case Report: Pseudo-Valentino's Syndrome – Post ERCP duodenal perforation presenting as acute appendicitis

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Case Report: Pseudo-Valentino's Syndrome – Post ERCP duodenal perforation presenting as acute appendicitis

Significance: Endoscopic Retrograde Cholangiopancreatography, is generally safe but carries risks for pancreatitis, bleeding, infection and perforation. To the date of writing, this is the first reported case of Valentino's syndrome after ERCP – duodenal perforation mimicking acute appendicitis.

Case presentation:

We present a 45 year old male who underwent ERCP for choledocholithiasis identified on MRCP after presenting with right upper quadrant pain. Difficult cannulation was noted during ERCP. Four days after ERCP, he developed tachycardia but no fever, associated with right lower quadrant pain and tenderness, Rovsing's Sign and Iliopsoas sign.

Management:

He was initially managed as a case of acute appendicitis. Abdominal CT scan was done and showed a perforation in the second part of the duodenum, and non inflamed appendix. He was managed as a case of post-ERCP perforation and was referred to surgery.

Immediate return of billous fluid and air was noted upon entry into the peritoneum during surgery. A perforation at the posterior aspect of D2 was noted. He underwent duodenorrhaphy and pyloric exclusion gastrojejunostomy and was sent home after a course of piperacillin-tazobactam.

4 weeks after ERCP, he had recurrence of right hemiabdominal pain. Imaging showed a, right-sided retroperitoneal fluid collection with pelvic extension. He underwent surgical drainage and was sent home after a course of Meropenem.

Recommendation: Duodenal perforation can mimic acute appendicitis. Intestinal contents can leak and pass along the right paracolic gutter to the appendicial fossa. This is an important differential diagnosis which must be considered in patients who underwent ERCP.

Keywords: Case Report, ERCP, complications, Duodenal Perforation, atypical presentation, Appendicitis, Valentino's Syndrome

Introduction

Endoscopic retrograde cholangiopancreatogram is becoming more common. In our institution alone, a total of 155 procedures was done in the year 2016. [1]

Endoscopic Retrograde Cholangiopancreatography is one of the standard therapy for the management of some pancreaticobiliary diseases including bile duct stones, strictures and some masses. This procedure is generally safe but still carries risk for complications such as pancreatitis, bleeding, infection and perforation.

The clinical presentation of duodenal perforation can vary from being asymptomatic, to having mild abdominal pain to generalized peritonitis. [2] However, duodenal perforation can mimic other disease entities. To the date of this writing, this is the first case report of of Valentino's syndrome after ERCP – duodenal perforation mimicking acute appendicitis.

Case Presentation and Management

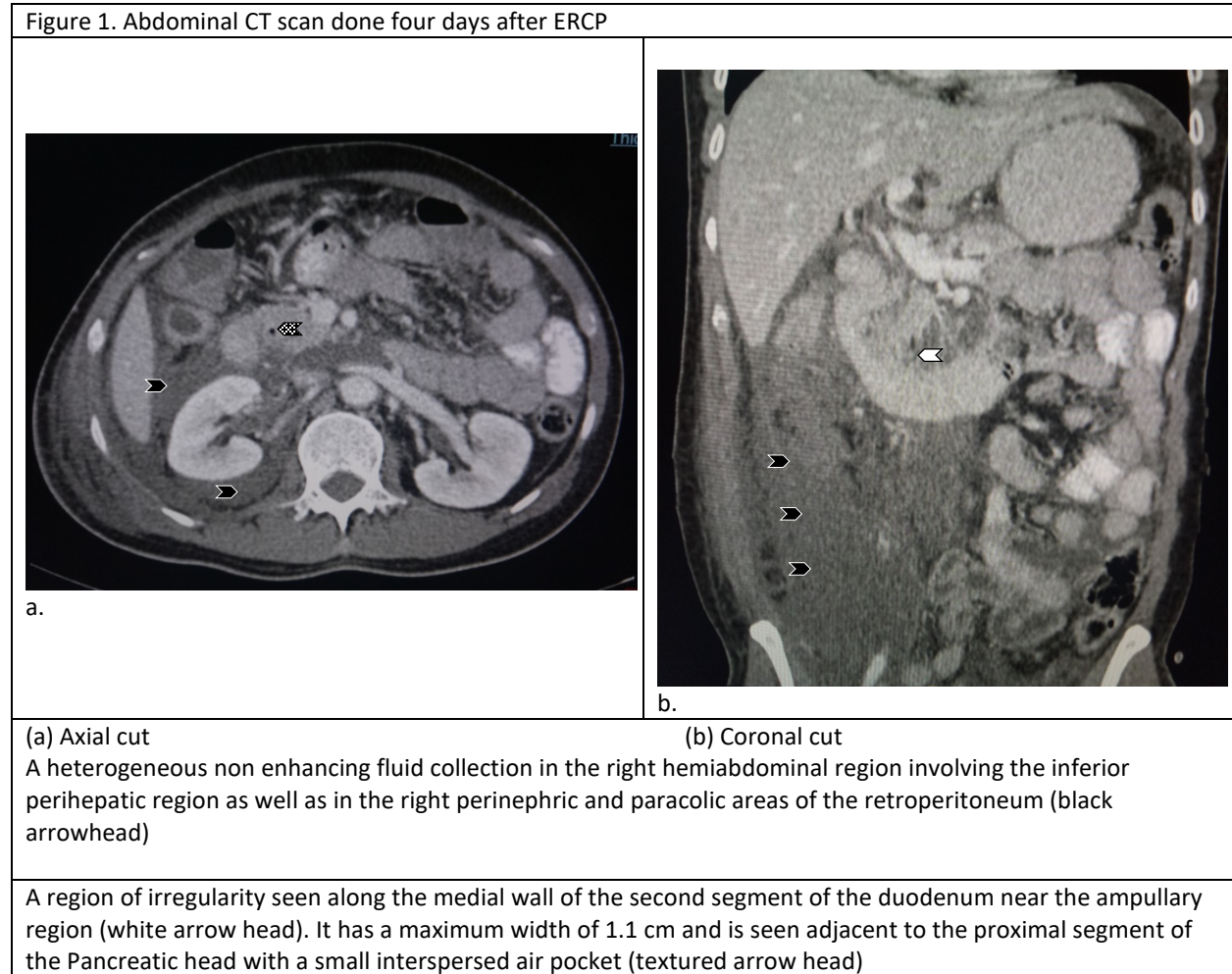
The patient is a 45 year old male who was admitted for Endoscopic Retrograde Cholangiopancreatography. He has no know comorbid conditions and has good baseline functional capacity. He already underwent open cholecystectomy one year prior to present admission. Few months prior to admission, he noted onset of right upper quadrant pain and on consult and further workup, he was noted to have a common bile duct stone on MRCP.

During ERCP, the scope was inserted up to the second part of the duodenum. The ampulla was was normal looking with bile flow. There was note of difficult cannulation. The common bile duct was eventually cannulated using sphincterotome and guidewire technique. Initial cholangiogram showed a dilated common bile duct (0.9cm). A filling defect was noted at the distal common bile duct (0.6cm). Minimal bleeding noted at the sphinctertomy site. Hemostatis achieved using submucosal epinephrine injection. Occlusion cholangiogram showed no filling defects

One day after ERCP, he developed pain in the right upper quadrant and on the epigastrium. Further workup was done showing a serum lipase of 994 U/l and leukocytosis with a complete blood count of Hemoglobin 146, Hematocrit 45, Platelet count 291, Leukocyte count 14.7, Neutrophil 91% and Lymphocyte count of 3% (from a pre-ERCP baseline of Hgb 140, Hct 42, Plateet 282, WBC 6.10, neutrophil 72%, lymphocyte 19%). He was managed as a case of post ERCP pancreatitis and was put on nothing per orem and was hydrated

Four days after ERCP he started to develop right lower quadrant pain. There were no associated bowel movement changes and any form of gastrointestinal bleeding. On examination he was seen normotensive at 110/80mm Hg, tachycardic at 115 beats per minute and febrile at 38.9 C°, the abdomen was normoactive, with mild stiffness of the abdomen. There was tenderness on the right lower quadrant. There was also pain on the right lower quadrant on palpation of the left lower quadrant and there was note of pain on the right flank upon extension of the right hip. At this point, the consideration was acute appendicitis. An abdominal ultrasound was done showing normal liver with smooth borders and homogenous parenchymal echo pattern. The common bile duct measures 0.4cm and intrahepatic ducts are not dilated. There is note of minimal ascites. Real time scan of the right lower quadrant with particular attention to the point of maximal tenderness shows no evidence of a blind ended, tubular, non compressible aperistaltic structure. There was no sonographic evidence of appendicitis.

Abdominal CT scan showed a large, right-sided predominantly retroperitoneal fluid collection with pelvic extension. On radiologic examination of the bowels, a soft tissue irregularity was seen involving the region of the duodenum likely a laceration. Furthermore reactive changes presenting as mucosal thickening of the ascending colon and small bowels were seen. The appendix was not inflamed.



He then underwent an emergent exploratory laparotomy. After the midline cut in the abdominal wall and eventually of the fascia, immediate return of billous fluid and air was noted upon entry into the peritoneum. The duodenum was then identified and mobilized showing a 1cm perforation in the posterior aspect of the second part of the duodenum. After repair of the duodenum, leak test was done and he underwent pyloric exclusion and gastrojejunostomy. He underwent several days of IV antibiotics using piperacillin-tazobactam and was eventually sent home.

Four weeks after discharge from the hospital, he underwent low grade undocumented fever with associated vague right upper quadrant pain. He was again admitted and imaging again showed a right-sided perinephric and right-sided paracolic retroperitoneal fluid collection with pelvic extension. There is no change in the subcutaneous edema and fat stranding present on the adjacent lateral abdominal wall as well as in the superior and mid portions of the right psoas muscle. The medial wall of the second segment of the duodenum remains slightly thickened and irregular. The previously noted interspersed air pocket are likewise no longer appreciated

He eventually underwent repeat exploratory laparotomy where a 10cm x 10cm right paracolic encapsulated abscess with necrotic tissue was noted. The abscess was evacuated and peritoneal lavage was done. He was eventually discharged improved after several days of Meropenem.

Discussion

Common complications associated with ERCP include sedation related, pancreatitis, bleeding, infection and perforation. [3]

In a 2007 systematic survey involving 16,855 patients, ERCP-attributable complications occurred in 6.85% (CI 6.46–7.24%) of patients with an associated mortality of 0.33% (CI 0.24–0.42%). Complications related to Cardiovascular events and/or analgesia-related was 1.33%. While other complications such as pancreatitis occurred in 3.47%, infections in 1.44%, bleeding 1.34%, and perforations in 0.60% of patients. [4] The rate of perforation seemed to decrease from previous reports of 1% of ERCP patients, (with an associated mortality rate of 16% to 18%) [5]

ERCP perforation has variable but frequently mild presentation. Mild abdominal tenderness can be found in 71% of cases while generalized peritonitis developing between 2 to 72 hours (median 6 hours) post ERCP is seen in 29%. In this similar review, 29% had high grade fever, 29% had low grade fever while 43% remained afebrile. [2]

On literature, the sensitivity and specificity of Rovsing's sign is 30.1% and 84.4% respectively and [6] the sensitivity and specificity of iliopsoas sign is 16% and 95% [7]. However, other conditions can still mimic appendicitis despite the specificity of these clinical findings. Duodenal ulcer perforation into the retroperitoneal space can present as acute appendicitis. After perforation, gastroduodenal contents can leak and pass along the right paracolic gutter causing peritonitis and signs and symptoms of acute appendicitis, [8]

This condition, called Valentino's syndrome, is named after Rudolph Valentino an Italian naturalized American actor in the 1920s. After collapsing at his Hotel in Manhattan, he was admitted at New York Polyclinic Hospital where he was diagnosed with appendicitis and gastric ulcers. He eventually died from sepsis from peritonitis. He was the first reported case of perforated ulcers mimicking appendicitis. [9]

In a review of cases of duodenal perforation after ERCP, ten out of fourteen cases had difficult cannulation during the procedure, three out of fourteen had no difficulty with cannulation while one out of fourteen had a perforation even before cannulation. In three out of fourteen cases, the perforations were erroneously identified as a duodenal diverticula. The presumed source of the injury was the sphincterotome in 43% while the remaining 57% of the injury was assumed to be from the endoscope. In 78% of the perforations, the diagnosis was made or suspected during ERCP while the remaining was diagnosed after the procedure. [2]

Post ERCP perforations can be managed medically if there is (1) minimal abdominal tenderness, (2) if sepsis is not present, (3) if the leak is small (small contrast dissipation) and (4) if there are no fluid collections on CT scan. Whereas surgical consult is warranted in patients with large (1) extravasation of contrast on ERCP, (2) intra or retroperitoneal fluid collection on CT scan (3) massive subcutaneous emphysema, (4) perforations associated with retained material such as stones or ERCP accessories or (5) failure of medical management. Failure of conservative management can lead to septic peritonitis from large intraabdominal fluid collections. [2]

Duodenal perforations can present with signs and symptoms similar to that of acute appendicitis. Thus, post ERCP perforation is an important differential diagnosis in patients who underwent ERCP.

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