Dysphagia as a Manifestation of Esophageal Pemphigus
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Introduction
Biliary dyskinesia (BD) is a group of functional disorders of the biliary tree, comprised of gallbladder dysfunction (GBD) and sphincter of Oddi dysfunction (SOD). The diagnosis of BD is based on the presence of biliary symptoms in the absence of gallstones with reduced gallbladder ejection fraction (GBEF). Despite diagnostic uncertainty, cholecystectomy for BD due to SOD and not due to actual gallbladder dyskinesia has increased significantly. We report a case of biliary pain and reduced GBEF, which resolved with ERCP and sphincterotomy without cholecystectomy.

Case Presentation
A 63 year-old Cauasian woman with a PMH of GERD presented with RUQ, post-prandial abdominal pain associated with nausea and vomiting over several years. The pain was sharp, waxing and waning, and occasionally radiated to the back. Proton pump inhibitors had not provided relief. Physical exam and extensive workup including laboratory markers, CT Abdomen, capsule endoscopy, and EGD were all unremarkable.

Given persistent biliary symptoms in the absence of any physical exam or laboratory abnormality, a hepatobiliary scintigraphy with cholecystokin (CCK) scan was performed, which showed partial emptying of the gallbladder and notably, a low GBEF (17%). There was reflux of activity back into the gallbladder as well, so SOD was suspected.

The patient underwent ERCP with sphincterotomy. Biliary cannulation revealed a mild stricture in the middle third of the common bile duct.

At 2-month follow-up, the patient’s abdominal pain had resolved. Repeat HIDA scan with CCK showed normal gallbladder contraction and emptying with normalization of the GBEF to 76%. Thereafter, the patient remained symptom free, and no recurrence of symptoms occurred.

Discussion
• In our patient presented with many features consistent with type III BD (symptoms without any objective findings). When extensive workup and conservative management failed to provide a definitive diagnosis or symptomatic relief, she underwent a HIDA and CCK scan.
• In order to evaluate biliary dyskinesia, a HIDA scan is often performed with CCK administration to quantify the excretion of bile from the gallbladder, described as the GBEF. Either GBD or SOD may cause reduced GBEF. As reduced EF is also seen in other clinical conditions, the reliability of EF as a disease-defining feature is unclear.
• Fig. 1C demonstrates the patient’s HIDA scan results, notable for a biphasic emptying pattern of the gallbladder in association with reduced gallbladder ejection fraction, which initially raised suspicion for SOD.
• Given the results of the HIDA scan, patient underwent ERCP and sphincterotomy alone without undergoing cholecystectomy resulting in symptom resolution. As seen in Fig. 2C, she had complete recovery of GBEF after the sphincterotomy and resolution of the biphasic emptying pattern.
• When evaluating patients with biliary pain, special attention should be paid to the CCK-HIDA scan for the biphasic emptying pattern, which may be suggestive of biliary reflux secondary to SOD instead of directly gallbladder dyskinesia.
• The number of elective biliary surgeries increased significantly with the introduction of laparoscopic cholecystectomy. Within the last 10 years, 26-38% of cholecystectomies were performed for an indication of BD. Prior studies showed mixed outcomes with inconsistent relief of symptoms postoperatively. Although the diagnostic and therapeutic approach to BD has not been crystallized, the reliability of EF as a disease-defining feature is unclear.
• When evaluating patients with biliary pain, special attention should be paid to the CCK-HIDA scan for the biphasic emptying pattern, which may be suggestive of biliary reflux secondary to SOD instead of directly gallbladder dyskinesia.

Case Presentation

Figures
Fig. 1A, 1B and 1C: Pre-sphincterotomy HIDA. Pre-CCK images demonstrated a distended gallbladder without bile to bowel excretion (1A). After CCK administration, there was a prominent common bile duct but poor emptying the gallbladder (1B). Quantitative analysis showed markedly decreased gallbladder ejection fraction with biphasic emptying pattern (1C). Findings were suspicious for SOD.

Fig. 2A, 2B and 2C: Post-sphincterotomy CCK HIDA. Although there was delayed radionuclide excretion prior to CCK (1A), emptying of the gallbladder was significantly improved with ejection fraction 76%. There was resolution of previously seen biphasic emptying pattern.

References