

Dysphagia as a Manifestation of Esophageal Pemphigus

Grace H. Yang, MD¹, Catherine Choi, MD¹, Yiyan Liu, Ph.D, MD², Weizheng W. Wang, MD¹

¹Division of Gastroenterology and Hepatology, Rutgers New Jersey Medical School, Newark, New Jersey ²Department of Radiology, New Jersey Medical School, Newark, New Jersey

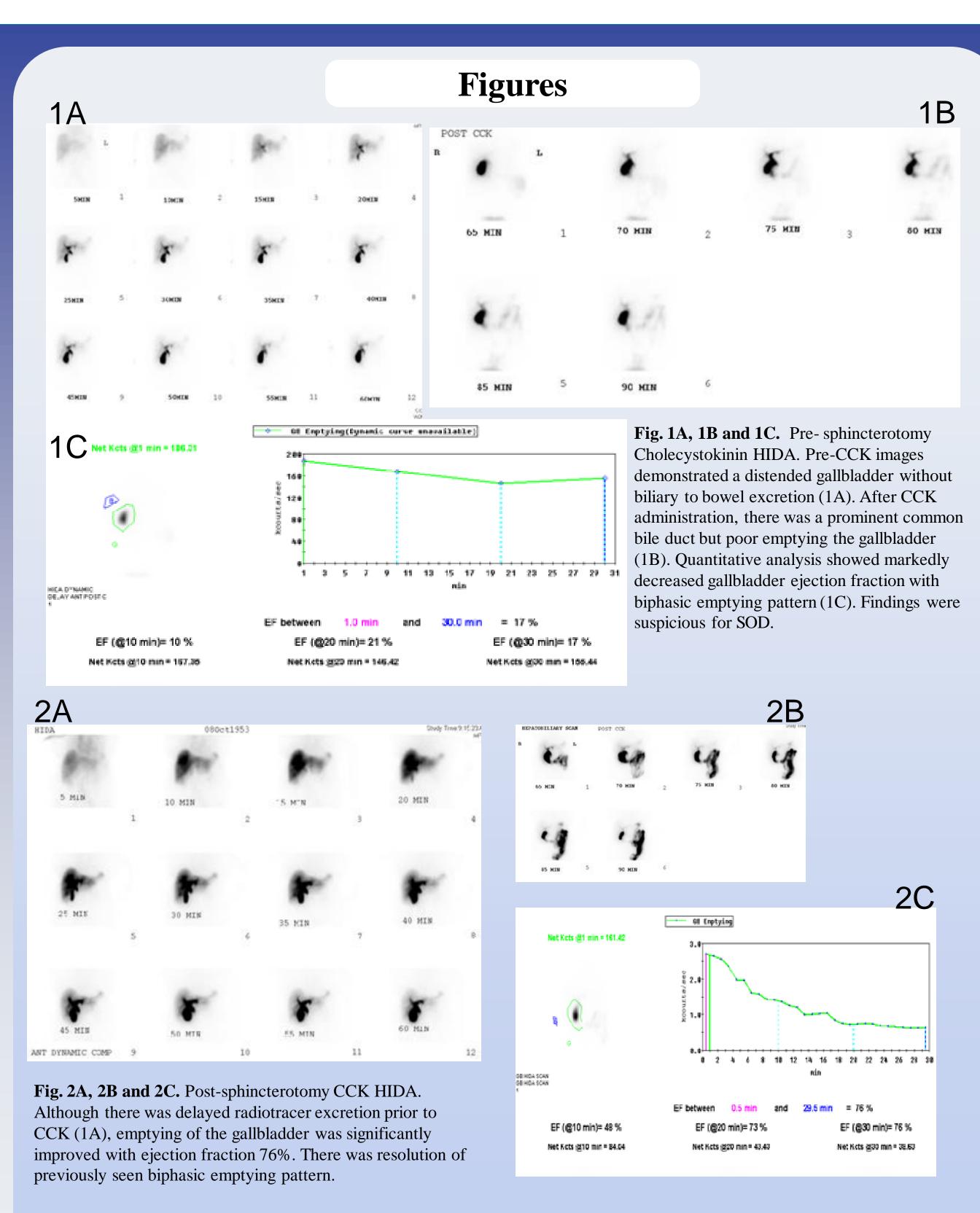


Introduction

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 dysfunction 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 Caucasian 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 iminodiacetic acid (HIDA) scan with cholecystokinin (CCK) 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%. At subsequent 6-month follow-up, the patient remained symptom free, and no recurrence of symptoms occurred.



Discussion

- Our patient presented with many features consistent with type III SOD (symptoms without any objective findings). When extensive workup and conservative management failed to provide a definitive diagnosis or symptomatic relief, she underwent a HIDA scan with CCK.
- 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 strictly 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, cholecystectomy still remains the mainstay of treatment in patients with BD.
- If conservative symptom management fails, ERCP and sphincterotomy should be considered prior to surgery in such patients as it can provide both symptomatic relief and improvement in objective measures such as the GBEF.
- Collaboration with Nuclear Medicine specialists to identify the biphasic emptying pattern characteristic of SOD in addition to low GBEF on the HIDA scan may prove to be invaluable towards the diagnosis and treatment of SOD.
- Further studies should be done to explore the observations from this case study further, which may provide a paradigm shift to the approach in identifying and treating SOD.

References

- 1. Wybourn CA, Kitsis RM, Baker TA, Degner B, Sarker S, Luchette FA. Laparoscopic cholecystectomy for biliary dyskinesia: Which patients have long term benefit? Surgery. 2013;154(4):761-767; discussion 767-768.
- 2. Goussous N, Kowdley GC, Sardana N, Spiegler E, Cunningham SC. Gallbladder dysfunction: how much longer will it be controversial? Digestion. 2014;90(3):147-154.
- 3. Veenstra BR, Deal RA, Redondo RE, et al. Long-term efficacy of laparoscopic cholecystectomy for the treatment of biliary dyskinesia. Am J Surg. 2014;207(3):366-370; discussion 369-370.
- 4. Bielefeldt K, Saligram S, Zickmund SL, Dudekula A, Olyaee M, Yadav D. Cholecystectomy for biliary dyskinesia: how did we get there? Dig Dis Sci. 2014;59(12):2850-2863.
- 5. Cairo SB, Ventro G, Sandoval E, Rothstein DH. Long-term results of cholecystectomy for biliary dyskinesia: outcomes and resource utilization. J Surg Res. 2018;230:40-46.
- 6. Singhal V, Szeto P, Norman H, Walsh N, Cagir B, VanderMeer TJ. Biliary dyskinesia: how effective is cholecystectomy? J Gastrointest Surg. 2012;16(1):135-140; discussion 140-131.
- 7. Yaghoobi M, Romagnuolo J. Sphincter of Oddi Dysfunction: Updates from the Recent Literature. Curr Gastroenterol Rep. 2015;17(8):31.
- 8. Hyun JJ, Kozarek RA. Sphincter of Oddi dysfunction: sphincter of Oddi dysfunction or discordance? What is the state of the art in 2018? Curr Opin Gastroenterol. 2018;34(5):282-287.
- 9. Wilcox CM. Sphincter of Oddi dysfunction Type III: New studies suggest new approaches are needed. World J Gastroenterol. 2015;21(19):5755-5761.