Two birds, one surgical stone: The first reported case of superior mesenteric artery syndrome secondary to biliary dyskinesia

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ABSTRACT

This is the first reported case of superior mesenteric artery syndrome secondary to biliary dyskinesia. SMA syndrome involves obstruction of the third portion of the duodenum, causing a gastric outlet obstruction due to narrowing of the space between the abdominal aorta and the superior mesenteric artery (SMA). Rapid weight loss has been shown to be a risk factor for this condition. We report a case found in a 14-year old Hispanic developmentally delayed female and review the literature. Our patient presented with a one-month history of abdominal pain, bilious emesis, and weight loss. She was seen at an outside facility where she had two abdominal operations without symptomatic relief. A HIDA scan performed at our facility was consistent with biliary dyskinesia, and an UGIS confirmed a concomitant SMA syndrome. An open cholecystectomy with a side-to-side duodenojejunostomy was performed. Postoperatively, patient continued with gastroparesis, requiring nutritional support via TPN and insertion of a jejunostomy tube. On post operative day 24, patient was discharged home tolerating full feeds. She has since been seen in follow up, and is doing well.

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1. Purpose

To perform a thorough literature review to ascertain the most appropriate surgical treatment for SMA syndrome having resulted from undiagnosed biliary dyskinesia.

Biliary dyskinesia is a motility disorder affecting the gallbladder and the sphincter of Oddi. Symptoms include nausea RUQ abdominal and epigastric pain, usually after a meal high in fat or cholesterol. It is often diagnosed in children with chronic abdominal pain [1]. Ultrasonography is usually unrevealing for choledolithiasis or other structural causes. The condition can be confirmed by a HiDa scan with CCK showing a poor gallbladder ejection fraction (c<35–40%), [2–5], with Carney et al. showing a PPV of 93% in patients with pain, nausea and an EF <15%. The cause is believed to be defects in the function and distribution of cholecystokinin receptors on the cells of the gallbladder wall leading to abnormal response to the hormone. No medical therapy has been found to be effective. Cholecystectomy has been shown to be curative in 70–90% of select patients [2].

Superior mesenteric artery syndrome, also referred to as Wilkie’s syndrome, is a syndrome characterized by extrinsic compression of the third portion of the duodenum which lies between the abdominal aorta and the superior mesenteric artery. The compression causes an obstruction to outflow of gastric contents. Patients experience most commonly abdominal pain, nausea, and vomiting. Weight loss is not only a symptom, but is also a risk factor for this disease process, as loss of omental fat density may be the cause for the acute change in the aortic-mesenteric artery angle [6]. Common complications of prolonged SMA syndrome include electrolyte abnormalities and their sequelae, nutritional deficiencies, and gastric perforation. Conservative treatments include nasogastric tube decompression of the obstructed stomach, nutritional support via PPN or TPN, and correction of electrolyte imbalances. Several surgical options are known to be available if these methods fail, including duodenojejunostomy, gastrojejunostomy [7] and resection of the Ligament of Treitz [6].

2. Case history

We present the case of a 12-year old female with a 1 month history of intermittent, crampy abdominal pain associated with emesis. She was recently admitted to an outside hospital for similar complaints where she underwent a diagnostic laparoscopy with appendectomy.
and cystectomy for a right paraovarian cyst. She was discharged after 4 days only to return 2 days later with persistent abdominal pain and vomiting. She then had several imaging studies, including an abdominal CT scan, stool studies and a brain MRI. A GI consult performed an upper endoscopy with biopsies and a diagnosis of Helicobacter pylori infection was made by stool studies. She was then started on amitriptyline and Reglan and was treated for H. pylori infection as an outpatient. She showed signs of Reglan induced dystonic reactions including hypertension resulting in discontinuation of Reglan. Review of systems was noted for abdominal pain, nausea, anorexia including 11 kg weight loss in the past two months with a BMI of 15.5. Past medical and surgical history was notable for developmental delay for which she was receiving physical, occupational and speech therapy; as mentioned she had a recent appendectomy and cystectomy. On physical exam, she was tachycardic and her abdomen was soft, non-distended with tenderness in the right upper quadrant. Laboratory data showed no leukocytosis, mildly elevated AST and ALT, normal bilirubin levels as well as CRP and ESR. Abdominal obstructive series revealed a non-obstructive bowel gas pattern and abdominal ultrasound showed gallbladder sludge, no gallbladder wall thickening or pericholecystic fluid. At this point, a diagnosis of biliary dyskinesia was suspected, with a differential diagnoses including gastroparesis, gallbladder pathology, brain tumor, and bulimia. Further work up with a HIDA scan and an upper GI series with small bowel follow through was planned and a GI consult recommended protonix IV.

On hospital day 2 a HIDA scan showed an ejection fraction of 22%; a diagnosis of biliary dyskinesia was then confirmed. An upper GI series with small bowel follow through showed mild dilatation of the second portion of the duodenum with delayed progression of contrast through the third portion of the duodenum as well as mild hyperperistalsis and retrograde peristalsis in the second portion of duodenum (Figs. 1 and 2). Given her history of rapid weight loss, this was felt to be consistent with SMA syndrome.

After failing a course of conservative management including bowel rest and nasogastric decompression, our patient was taken to the OR on hospital day 6 for an open cholecystectomy with side-to-side duodenojejunostomy. The gallbladder was found to be large, dilated, and thickened with multiple omental adhesions which were lysed with electrocautery. A top down dissection of the gallbladder was performed. The Kocher maneuver was performed to expose the duodenum. A loop of jejunum 30 cm from ligament of Treitz was brought up to the anterior sidewall of duodenum through transverse mesocolon. A side-to-side anastomosis was created using a GIA stapler and was reinforced internally with hand-sewn running suture. The mesocolon was sutured circumferentially to the duodenum.

Postoperatively our patient continued to experience nausea and abdominal pain. On post operative day two she began having emesis but did not tolerate a nasogastric tube. She was started on Toradol, Phenergen, Tylenol and Dilaudid PRN.

On post operative day three an obstructive series showed a distended stomach with air-fluid levels and residual barium within pelvis from a prior study. The following day a PICC line was placed for TPN. Her abdominal pain had resolved but she was still having emesis. An upper GI was performed which showed contrast progression with slight narrowing at the duodenojejunal anastomosis site. Our patient continued to have poor PO tolerance and episodes of vomiting, and so TPN was continued. On post operative day 10 an UGIS showed contrast pooling in the stomach with poor progression to the duodenum. We then placed a gastrojejunostomy tube for gastric decompression and jejunal feeds. Our patient slowly began gaining weight and was discharged home on post operative day 24, tolerating PO feeds and in good spirits.

3. Method

A thorough literature review was conducted of articles printed in English in PubMed between January of 2000 and November of 2014. We reviewed the medical and surgical treatments for biliary dyskinesia. We then focused our review towards the surgical management options for SMA syndrome as reported in case reports and case series presented in PubMed.

4. Results

Upon review, cholecystectomy was the primary treatment for biliary dyskinesia [1,2] and duodenojejunostomy was found to be the preferred surgical treatment for SMA syndrome [3,7,8].
Therefore, it was determined that it is not only feasible but preferable to perform both procedures during the same operation.

We combined our results from research on two separate pathologies to determine a unified treatment plan in hopes of resolving both interconnected disease processes. Our solution was one operation to treat both surgical pathologies.

5. Discussion

Biliary dyskinesia is the dysfunction of the autonomic nervous system of the gallbladder leading to uncoordinated contraction and relaxation of the gallbladder and sphincter of Oddi, leading to distension, inflammation and dysfunction [3]. It is diagnosed via a cholecystokinin stimulated hepatobiliary scan, demonstrating a gallbladder ejection fraction less than 35%. Accepted treatment is with laparoscopic cholecystectomy, demonstrating a 67–100% long term success rate [3,9].

Superior mesenteric artery syndrome involves the entrapment of the third portion of the duodenum between the aorta and superior mesenteric artery. This occurs when the aortomesenteric angle reduced from the normal of 10–28 mm to 2–8 mm [8], which generally occurs after rapid weight loss [10,11]. This is diagnosed with barium studies and can be confirmed with CT measuring the angle [11]. Different surgical remedies have been described, including duodenoojejunostomy, gastrojejunostomy [7] and resection of the Ligament of Treitz [6].

This is the first reported case of SMA syndrome as a result of biliary dyskinesia. Biliary dyskinesia is treated most commonly with a laparoscopic cholecystectomy [2–5], and current consensus is to treat refractory cases of SMA syndrome with a duodenoojejunostomy [7,10].

Our research has led us down two paths, one investigating the standard of treatment for biliary dyskinesia and one for SMA syndrome. It is our belief that treating the biliary dyskinesia may have enabled the SMA syndrome to resolve, but by treating both pathologies in one operation, we provided our patient with several advantages, including the following: a faster recovery time to normal gastrointestinal motility and relief of emesis, a shorter length of stay, and elimination of a possible second operation. We believe a two-stage procedure would have been more difficult in this patient due to the likely formation of adhesions, and that due to social issues our patient would have required prolonged hospitalization.

6. Conclusion

It is our conclusion that surgically treating a patient with SMA syndrome secondary to biliary dyskinesia with a cholecystectomy and gastrojejunostomy during one operation is optimal.

References