GAVIN PUBLISHERS

Annals of Case Reports

Chu YK, et al. Ann Case Rep 2016: G114

Case Report

Intrahepatic Cholestasis Complicating Superior Mesenteric Artery Syndrome

Rong-Hsin Yang and Yum-Kung Chu*

Department of Nuclear Medicine, Taipei Veterans General Hospital, Taiwan

***Corresponding author:** Yum-Kung Chu, Department of Nuclear Medicine, Taipei Veterans General Hospital, No. 201, Shipai Road Sec. 2, Beitou District, Taipei, Taiwan 11217, Tel: 886-2-28757301; Fax: 886-2-28715849; E-mail: ykchu@vghtpe.gov.tw

Citation: Chu YK (2016) Intrahepatic Cholestasis Complicating Superior Mesenteric Artery Syndrome. Ann Case Rep 2016: G114.

Received Date: 30 September, 2016; **Accepted Date:** 31 October, 2016; **Published Date:** 07 November, 2016

Abstract

Superior mesenteric artery (SMA) syndrome is a rare but potentially life-threatening entity of proximal intestinal obstruction, and characterized by extrinsic compression loss of fatty tissue of the aortomesenteric aperture as a result of a variety of debilitating conditions. Various gastrointestinal presentations of SMA syndrome have been documented. We herein describe a rare complication of SMA syndrome, in form of intrahepatic cholestasis. SMA syndrome seemingly appears under-diagnosed and the condition is more common in debilitating patients. A high awareness for timely recognition of SMA syndrome is important for proper management.

Keywords: Hepatobiliary scan; Intrahepatic cholestasis; Superior mesenteric artery syndrome; Upper gastrointestinal obstruction

Introduction

Superior mesenteric artery (SMA) syndrome was first described in 1861 by Von Rokitanski [1]. Only 400 cases have been reported in the medical literature [2]. It is caused by pinching of the duodenum between the SMA and the aorta, due to loss of intervening mesenteric fat pad. Classical symptoms are usually chronic, with postprandial epigastralgia, bloating and vomiting, or acute intestinal obstruction [3]. Since symptoms are non-specific diagnosis depends on high index of suspicion. SMA syndrome is far more common in debilitating patients and seemingly goes under-diagnosed [4]. Treatment of SMA syndrome begins with a conservative approach. Severe bowel obstruction is life threatening. Surgical approach is indicated when conservative management fails, or in severe cases.

Case Report

A 64-year-old male presented to the emergency room with severe epigastralgia, postprandial bloating, vomiting, and 8-Kg weight loss over the past 3 months. The patient revealed a history of peptic ulcer for the last 3 years and underwent open cholecystectomy 8 months ago for symptomatic chronic cholecystitis. On examination, it was noted that he appeared emaciated, anemic and febrile at 37.7ºC. There was an obviously distended stomach boundary on percussion. Blood work was significant for WBC count 13,700/mm^3, RBC 3.1 m/mm^3, hemoglobin 6.9 g/dL (normal 12-16), blood urine nitrogen 46 mg/dL (BUN, normal 7-20), creatinine 2.1 mg/dL (normal 0.5-1.5) and amylase 333 U/L (normal 40-140). The liver profile was slightly abnormal with alanine aminotransferase 49 U/L (ALT, normal <35), aspartate aminotransferase 46 U/L (AST, normal <41), and total bilirubin 1.2 mg/dL (normal <0.4). The remainder of his laboratory works was within normal limits. Abdominal ultrasound and computed tomography (CT) revealed marked distention of the stomach and the duodenum, consistent with duodenal outlet obstruction.

Tc-99 m disofenin (Hepatolite®) scintigraphy for an assessment of his hepatobiliary function (Figure 1) demonstrated good hepatocyte function (rapid liver uptake and rapid blood pool clearance), but no bile excretion. Even up to 24 h, persistent intrahepatic cholestasis without biliary-to-bowel transit was noted. Scintigraphic findings were compatible with a classic picture of acute high-grade biliary obstruction. The patient was begun on antiemetics, nasogastric intubation for gastric decompression and total parenteral nutrition. However, conservative approach over 3 weeks failed. At this point, a surgical intervention was commenced. An explore laparotomy showed severe distension of the stomach and the duodenum, and dilatation of the common bile duct. Aortomesenteric compression upon the third portion of the duodenum was noticed, with an acute angle of 15^0 between the SMA and the aorta (Figure 2). T-tube drainage and duodenojejunostomy were done, and the patient did well and symptoms resolved after the surgery. SMA syndrome elucidated his clinical situation.

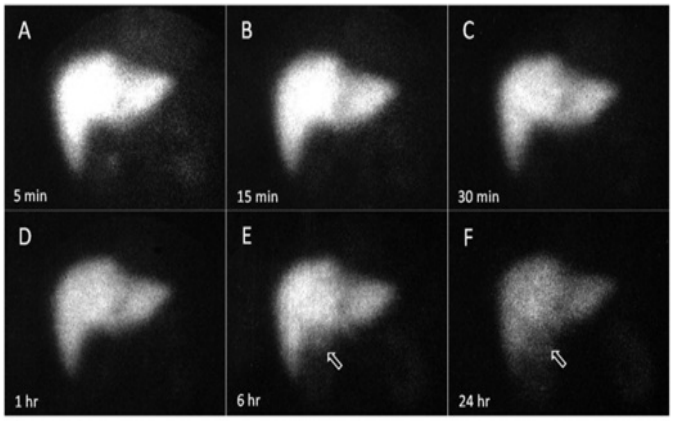

Figure 1: Tc-99m Hepatolite® cholescintigraphy.

Rapid liver uptake and faint cardiac and renal activity were seen at 5 min, suggestive of good hepatocyte function (A). No visualization of the common bile duct and failure of radiotracer transit to the bowel were notable in 5, 15, 30 and 60 min image (B, C & D). Trace amount of activity persisted in the gallbladder fossa (arrow) from 6 to 24 hr, interpreted as stasis of bowel contents in the duodenum (E and F). Substantial radiotracer remaining in the liver at 24 hr but no colonic activity was seen, consistent with intrahepatic cholestasis (F).

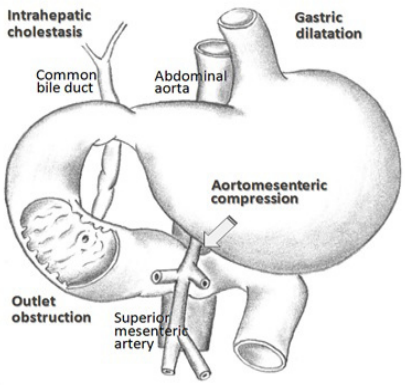

Figure 2: Schematic drawing showing relevant anatomy of the patient with superior mesenteric artery (SMA) syndrome.

Discussion

SMA syndrome arises from a reduction in the angle formed between the origin of SMA and the abdominal aorta, thereby pinching the third portion of the duodenum. Normally the aortomesenteric angle is between 36^0 to 56^0 with an opening of 10 to 20 mm for an aortomesenteric aperture. Lack of periduodenal and retroperitoneal fat pads can lead to a more acute angle resulting in duodenal compression [4]. In the majority of cases, profound weight loss from malabsorption, cancer and anorexia nervosa is responsible for this anatomic phenomenon [3, 5]. Classical symptoms are postprandial epigastric pain, nausea, vomiting, and weight loss, or acute upper gastrointestinal obstruction [3]. The diagnosis of SMA syndrome remains challenging as other disorders can mimic its presentation.

Treatment options should be based on severity of the disease. Fasting, total parenteral nutrition (TPN), correction of fluid and electrolytes, and gastric decompression for short duration constitute a conservative treatment for SMA syndrome with refractory gastroparesis [5]. There are no guidelines regarding the precise duration of the conservative approach [4]. When intragastric pressure from gastric distension gradually exceeds 30 cm H_2O over a period of time, intramural blood flow can be impaired and results in bowel ischemia and perforation [6]. Surgery is indicated when there is a failure of conservative treatment. Various surgical interventions have been associated with SMA syndrome, directly or indirectly, by releasing the extrinsic vascular compression [4], including gastrojejunostomy or duodenojejunostomy with or without duodenal mobilization (known as the Strong's procedure) [4].

Obstructive jaundice as a complication of SMA syndrome has rarely been reported in the literature [3]. The pathogenesis of terminal biliary duct obstruction in SMA syndrome is not clearly understood, but some factors are postulated. In a normal situation, bile excretion into the duodenal lumen is effected primarily by relaxation of the sphincter of Oddi. Manometric studies in humans have shown that the sphincter has a basal pressure of 10 mmHg. As seen in our case, it is reasonable to assume that a prior cholecystectomy can possibly lead to dysfunction of the Oddi sphincter (namely postcholecystectomy syndrome), and that the swollen duodenum can conceivably cause an enormous strain on the common bile duct. These risk factors can obstruct bile flow through the sphincter and therefore cause bile retention and intrahepatic cholestasis.

Hepatobiliary scan with Tc-99m-labeled iminodiacetic acid (IDA) derivate has been used in the diagnosis of various biliary disorders for almost 4 decades [7, 8]. After intravenous injection, the IDA derivates are transported in the blood by binding to serum albumin. They dissociate from albumin in the hepatic perisinusoidal space (space of Disse) and are taken up by hepatocytes via receptor-mediated endocytosis. The agents are subsequently secreted into the biliary system unchanged, without undergoing conjugation [8]. Although hepatobiliary agents are not metabolized, they follow the path of intracellular transit similar to bile salts, free fatty acids, bilirubin, hormones, drugs and toxins [8]. Their transit reflects the whole course of bile formation and excretion (Figure 3).

The diagnostic strength of scintigraphy lies in the fact that the information provided defines pathophysiology rather than anat-

omy of the hepatobiliary system [8]. As illustrated in this presentation, hepatobiliary scintigraphy provided a visual display of the biliary excretion and disclosed the event of subclinical cholestasis. The risk of biliary complications should be considered in the assessment and management of SMA syndrome.

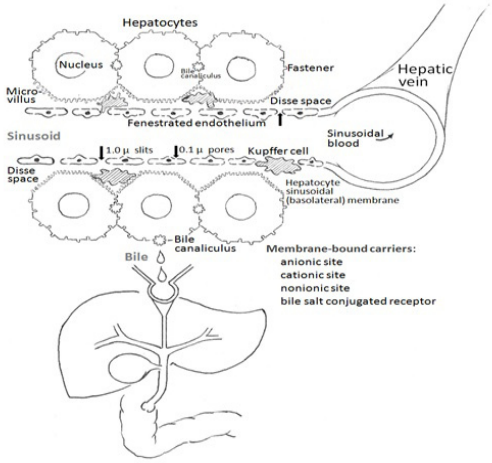

Figure 3: Schematic diagram showing relevant anatomy and physiology of bile excretion.

Conflict of Interests

The authors declare no conflict of interests regarding the publication of this paper.

References

1. Merrett ND, Wilson RB, Cosman P, Biankin AV (2009) Superior mesenteric artery syndrome: diagnosis and treatment strategies. J Gastrointest Surg 13: 287-292.
2. Roy A, Gisel JJ, Roy V, Bouras EP (2005) Superior mesenteric artery (Wilkie's) syndrome as a result of cardiac cachexia. J Gen Intern Med 20: 3-4.
3. Jeune F, d'Assignies G, Sauvanet A, Gaujoux S (2013) A rare cause of obstructive jaundice and gastric outlet obstruction. World J Gastrointest Surg 5: 192-194.
4. Capitano S, Donatelli G, Boccoli G (2012) Superior mesenteric artery syndrome- Believe in it! Report of a Case. Case Reports in Surgery 2012: 3.
5. Mehler PS, Krantz MJ, Sachs KV (2015) Treatments of medical complications of anorexia nervosa and bulimia nervosa. J Eat Disord 3: 15.
6. Kao KH, Tsai SH, Yu CY, Huang GS, Liu CH, et al. (2009) Unusual complication of superior mesenteric artery syndrome: spontaneous upper gastrointestinal bleeding with hypovolemic shock. J Chin Med Assoc 72: 45-47.
7. Loberg MD, Cooper M, Harvey E, Callery P, Faith W (1976) Development of new radiopharmaceuticals based on N-substitution of iminodiacetic acid. J Nucl Med 17: 633-638.
8. Ziessman HA (2014) Hepatobiliary scintigraphy in 2014. J Nucl Med 55: 1-9.