Superior Mesenteric Artery Syndrome Masquerading as Recurrent Biliary Pancreatitis

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The superior mesenteric artery syndrome (also known as Wilkie's syndrome) is a rare entity with various manifestations. The syndrome usually affects young adults. It is commonly described as a complication of spinal procedures or after significant weight loss. It is caused by obstruction of the post-papillary duodenum by an acute angle between the SMA and the aorta. It usually presents as crampy upper abdominal pain, relieved by vomiting or crouching. It may mimic pancreatitis due to the location and nature of the pain and an elevation in blood amylase [1-3].

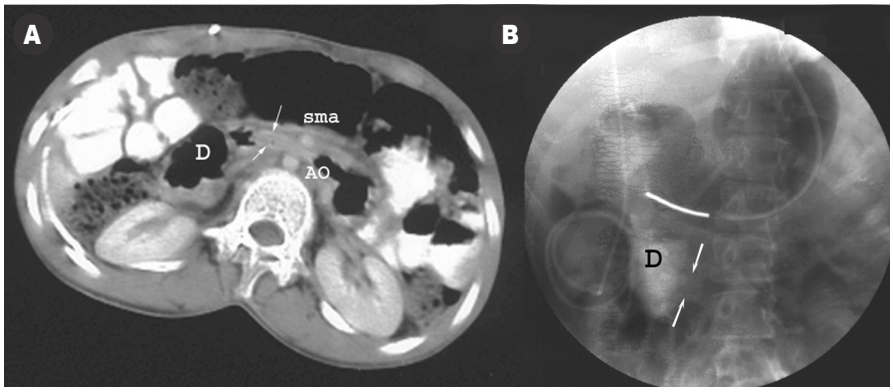
We present a 16 year old boy with cerebral palsy and hydrocephalus who was misdiagnosed as suffering from recurrent bouts of biliary pancreatitis for 2 years. A cholecystectomy, laparotomy and endoscopic retrograde cholangiopancreatography were performed before the correct diagnosis was established, and he was successfully treated conservatively.

Patient Description

The patient described here was born prematurely (800 g) and subsequently suffered from cerebral palsy and hydrocephalus and was treated with a ventriculoperitoneal shunt. At the age of 2 years he had a gastrostomy inserted for feeding purposes and at age 3 underwent a gastric (Nissen) fundoplication due to gastroesophageal reflux. He had no further gastrointestinal complaints until the age of 14 when he was re-admitted to our ward because of severe abdominal pain without vomiting and cholelithiasis on a transabdominal ultrasound. The complaints resolved spontaneously within 24 hours.

His second admission was 1 year later, again due to severe pain with vomiting and obstipation, with blood biochemistry results showing amylase 1600 U and bilirubin 45 mmol/ml with a white blood cell count of 20,600/mm^3. After insertion of a nasogastric tube the patient felt almost immediate relief. An abdominal ultrasound demonstrated a small amount of abdominal fluid without evidence of gallstones. At his third admission 2 weeks later he presented with the same complaints and repeat ultrasound showed sludge in the gallbladder. The patient underwent a laparoscopic cholecystectomy and an intraoperative cholangiogram which was normal. The pathologic report showed signs of chronic cholecystitis without evidence of stones or sludge. Two months later, he was re-admitted for the fourth time due to upper abdominal pain with vomiting and obstipation and amylase 1278 U. Again, the complaints resolved promptly after insertion of a nasogastric tube. He was discharged with a diagnosis of postoperative pancreatitis and was free of complaints for 2 months, when he presented again with the same complaints, which were first ascribed to an obstructed ventriculperitoneal shunt but afterwards were deemed more typical of an intestinal obstruction. A computerized tomography scan showed a normal pancreas but with signs of intestinal obstruction. He was transferred to the operating room for laparotomy, which did not reveal any signs or causes of intestinal obstruction.

Four months later, he was hospitalized for the sixth time, again with the same complaints and objective findings, including a high amylase level. Again, there was a prompt resolution of all symptoms. An



[A] Axial CT scan. The proximal duodenum (D) is distended due to compression on the distal duodenum (arrows) because of a narrow angle between the aorta (AO) and the superior mesenteric artery (sma).

[B] Upper gastrointestinal series. The tip of a nasojejunal tube is present in the antrum of the distended stomach. Contrast material illustrates the distended proximal duodenum (D), with an abrupt cutoff (arrows) due to compression by the superior mesenteric artery. A ventriculoperitoneal shunt is present in the right upper quadrant.

SMA = superior mesenteric artery

ERCP was performed, with normal results. When the patient presented again, for the seventh time within 14 months, again with the same symptoms, meticulous revision of the previous imaging studies [Figure A] raised the suspicion of SMA syndrome. This diagnosis was proven to be correct by means of an upper gastrointestinal series with the patient in the supine and decubitus positions [Figure B]. He was started on a regimen of enteral and parenteral alimentation and after gaining about 4 kg he was discharged. He remained asymptomatic for the following 3 years. After that period he suffered another bout of weight loss and was operated at another hospital where a duodenoduodenostomy was performed.

Comment

The superior mesenteric artery syndrome was first suggested in 1842 by Rokitansky. It was defined pathologically in 1921 by Wilkie, who named it "chronic duodenal ileus." The syndrome was overdiagnosed during the following decades as a reason for vague abdominal pains but was actually found to affect only a very small number of patients. The syndrome is caused by the compression of the third portion of the duodenum as it passes between the SMA and the aorta. Angiographic studies have shown that afflicted patients have a hyperacute angle between these two vessels, therefore enabling the duodenal obstruction. This situation may mimic acute pancreatitis due to upper abdominal pain with vomiting and elevated amylase levels. It is thought that the adipose and lymphatic tissue surrounding the vessels protect against the potential obstruction, and indeed the syndrome occurs almost invariably in thin persons or those who suffer from abrupt weight loss. The classical description is in patients who are confined to spicas or who underwent spinal procedures, because these conditions accentuate lordosis. It has also been described in patients following brain trauma. The key to a correct diagnosis is a high index of suspicion. The definite diagnostic tool is an upper gastrointestinal series using diluted barium with the patient supine and prone or in the decubitus position. Duodenal obstruction while supine, followed by prompt passage of barium while changing position, is considered diagnostic. In recent years, the diagnosis has also been made through CT scans, and abdominal CT-angiography is considered highly specific [4,5]. The treatment varies from conservative means using enteral or parenteral alimentation, to surgery.

The patient we describe here demonstrates how difficult the diagnosis can be if it is not suspected and specifically sought.

ERCP = endoscopic retrograde cholangiopancreatography

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