Chilaiditi Syndrome Complicated by a Closed-Loop Small Bowel Obstruction

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Chilaiditi syndrome is a rare disease in which intestinal obstruction is caused by hepatodiaphragmatic interposition of the colon or small bowel. Demetrius Chilaiditi described the first cases of this disease in 1910.[1] Most patients with this intestinal anomaly are asymptomatic throughout their lives; however, they can manifest with intermittent abdominal pain, distention, vomiting, anorexia, and constipation that on rare occasions require surgical intervention.[2]

Case Report

A 55-year-old African-American woman presented to the emergency department with a history of gradually increasing abdominal pain localized to the epigastrium and right upper quadrant that radiated to the right shoulder. Her pain was associated with nausea, constipation, and obstipation, as well as nonbilious and nonbloody emesis on several occasions. The patient reported having vomiting episodes during her childhood. Her surgical history was significant for a Cesarean section, ovarian cystectomy, and appendectomy. On physical examination, she was hemodynamically stable. Her abdomen was distended with decreased bowel sounds, tenderness in the right upper quadrant, and a positive Murphy sign with rebound tenderness. Results of the patient's complete blood count, liver function tests, and urinalysis were normal. A right upper quadrant ultrasound demonstrated irregular shadowing in the liver, with a morphology similar to that of the small bowel. The patient's gallbladder had normal dimensions and wall thickness, and there was no evidence of cholelithiasis.

Computed tomography (CT) scans of the abdomen and pelvis performed with intravenous and oral contrast demonstrated an abnormal course and configuration of the small bowel and colon, with portions of the transverse colon traveling through the Morrison pouch; multiple small bowel loops were also seen interposed among the liver, abdominal wall, and diaphragm. The CT scans also demonstrated mild small bowel wall thickening with inflammatory changes, air-fluid levels, and an abrupt transition in bowel caliber that was characteristic of small bowel obstruction (Figure 1).

On the basis of the patient's presentation and imaging results, the patient was taken to the operating room for an exploratory laparotomy. A 6–8-inch segment of the terminal ileum that was approximately 3 feet from the ileocecal valve was involved in a closed-loop obstruction above the liver secondary to adhesions between the liver and the abdominal wall. The bowel initially appeared to be mildly ischemic, but it regained viability after the closed loop was untwisted and warm saline was applied; hence, resection was not necessary. The colon did not have any abnormalities.

Discussion

In 1910, the radiologist Demetrius Chilaiditi described 3 patients who had an interposition of the bowel between the liver and right hemidiaphragm. A Chilaiditi sign is thus used to describe the incidental radiologic finding of a colonic or intestinal hepatodiaphragmatic interposition in an asymptomatic patient. This sign is commonly misinterpreted as pneumoperitoneum. The prevalence of

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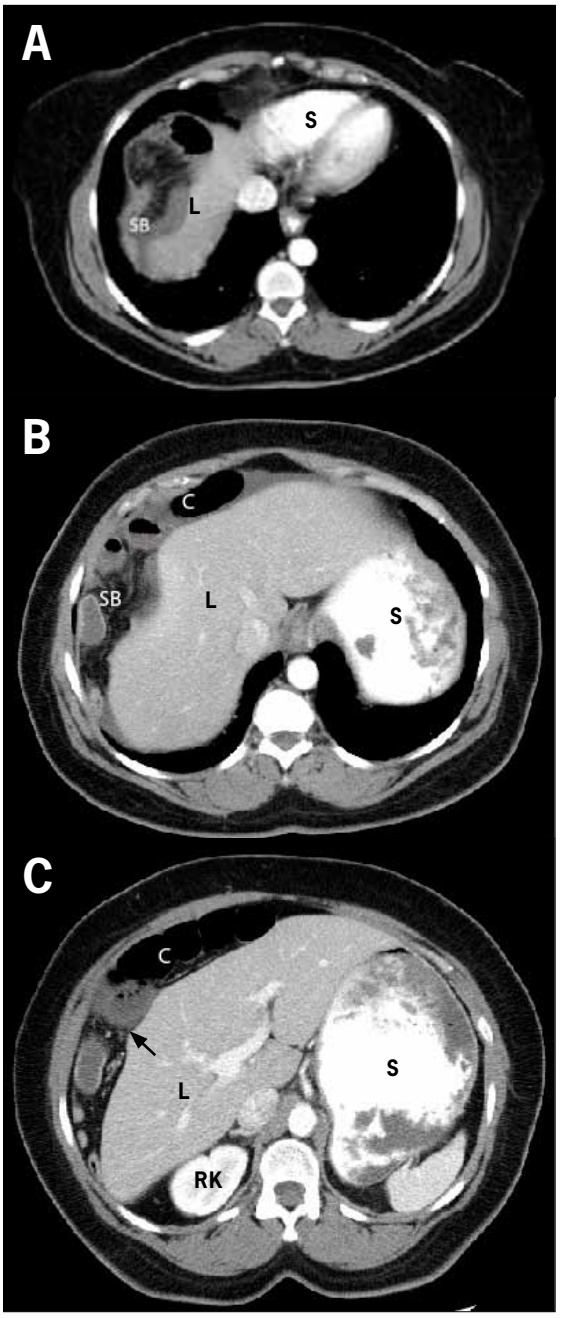

Figure 1. Computed tomography scans with contrast showing loops of small bowel between the dome of the liver and diaphragm, with a subsequent, abrupt change in the caliber of the bowel. Figure **1A** shows a loop of small bowel located between the liver and the abdominal wall. Figure **1B** shows the incarcerated small bowel loop and the hepatic flexure of the colon between the liver and the abdominal wall. Figure **1C** shows the transverse colon interposed between the liver and the abdominal wall. The arrow points to an area of fat stranding in the mesentery.

C=colon; L=liver; RK=right kidney; S=stomach; SB=small bowel.

Chilaiditi sign in the general population is 0.025–0.28%, and the sign is more prevalent in male patients than female patients.[3] The bowel segments most commonly found interposed between the liver and diaphragm or abdominal wall are the colonic hepatic flexure and transverse colon, although interposition of the small bowel has also been reported.[4,5]

Physiologic and conformational patterns of hepatic and colonic embryogenesis and adult anatomy usually prevent the development of colonic interposition. Factors that predispose patients to the development of Chilaiditi sign include reduced liver dimensions, elongation of the ligamentous suspension of the liver, and redundancy of the colon. Congenital diaphragmatic, hepatic, or intestinal anomalies and pathologies associated with the development of this sign include right hepatic lobe segmental agenesis, relaxation or agenesis of the mesentery suspensory ligaments, chronic constipation, redundant and hypermobile transverse mesentery and transverse colon, and significant weight loss. Another important cause of Chilaiditi sign is severe chronic obstructive pulmonary disease and its subsequent elongation of the diameter of the lower thoracic cage, which results in a broader space in which colonic interposition can occur. Elevation of the right hemidiaphragm from congenital hernias and eventration of the diaphragm also predispose patients to development of Chilaiditi sign. Furthermore, the characteristics most frequently associated with Chilaiditi sign—cirrhosis, ascites, and decreased hepatic size—increase the space between the liver and diaphragm; these characteristics occur in up to 5% of patients.[6]

Patients with Chilaiditi syndrome present with symptoms of bowel obstruction, including anorexia, nausea, emesis, abdominal pain, distension, and obstipation.[7] The patient in this case study presented with these symptoms, as well as right shoulder pain from diaphragmatic irritation.

Chilaiditi syndrome has been associated with a volvulus of the transverse colon. Due to mesenteric attachments, normal anatomy prevents the transverse colon from rotating and developing a volvulus. Factors that predispose patients to developing a volvulus include increased colonic mobility and a site of axial colonic rotation; specific factors that predispose patients to developing a transverse colon volvulus include congenital malrotation of the midgut and associated agenesis of the phrenocolic ligament or mesenteric root axial shortening.

Diagnosis of Chilaiditi syndrome is based upon clinical findings and signs observed on plain radiographs and CT scans. CT scans of the abdomen can enable clinicians to differentiate among subphrenic fluid, true pneumoperitoneum, and air within the bowel lumen. This differentiation is of the utmost importance for establishing the diagnosis of hollow viscus perforation, which can also complicate Chilaiditi syn-

drome when the involved bowel segment strangulates and eventually perforates.[8] The radiologic differential diagnosis is established by observing an elevation of the right hemidiaphragm due to caudal displacement of the liver, haustral markings between the liver and diaphragmatic surface, and the absence of image displacement with changes in the patient's position. Pneumoperitoneum and subdiaphragmatic fluid collections are mobile on lateral decubitus radiographs and are accompanied by pulmonary findings such as ipsilateral pleural effusion and basilar atelectasis.

In most cases of Chilaiditi syndrome, management is conservative and consists of bowel decompression, bowel rest, and aggressive fluid rehydration.[9] Patients who fail conservative therapy should undergo an exploratory laparotomy.[10] Failure of nonsurgical treatment for Chilaiditi syndrome has been associated with colonic volvulus and obstruction.[11] To our knowledge, there have not been any previous reports of small bowel obstruction necessitating surgical treatment in patients with Chilaiditi syndrome.

Summary

Chilaiditi syndrome is a rare form of bowel obstruction caused by the interposition of the colon or small bowel into the hepatodiaphragmatic space. This condition usually resolves with conservative management, but in this case report, the patient had a closed-loop small bowel obstruction that required surgical intervention.

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Review

Chilaiditi Syndrome: A Rare Entity with Important Differential Diagnoses

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Over a century ago, the radiologist Demetrius Chilaiditi reported a small case series of 3 patients with the incidental radiologic finding of colonic interposition between the liver and diaphragm.[1] Thereafter called the Chilaiditi sign, this finding is a rare anomaly incidentally seen on chest or abdominal radiographs, with an incidence of 0.025–0.28%.[2] Chilaiditi syndrome refers to the medical condition in which a Chilaiditi sign is accompanied by clinical symptoms.[3]

Intestinal, hepatic, and/or diaphragmatic etiologies contribute to the pathogenesis of Chilaiditi sign and syndrome. Under normal conditions, suspensory ligaments and fixation of the colon impede interposition of the colon between the liver and diaphragm. However, variations in normal anatomy can lead to the pathologic interposition of the colon. These anatomic variations can include the absence, laxity, or elongation of the suspensory ligaments of the transverse colon or the falciform ligament, as well as dolichocolons or congenital malpositions. Anatomic distortions can also result from functional disorders such as chronic constipation (colonic elongation and redundancy), aerophagia (gaseous distension of the colon), cirrhosis (liver atrophy or relative atrophy in the medial segment of the left lobe of the liver), diaphragmatic

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paralysis, chronic lung disease (enlargement of the lower thoracic cavity), obesity, multiple pregnancies, and ascites (increased intra-abdominal pressure). Mental retardation and schizophrenia are also associated with anatomic abnormalities that result in Chilaiditi sign.[2,4-7]

Colonic interposition is usually an asymptomatic radiologic sign. In patients presenting with Chilaiditi syndrome, the most common symptoms are gastrointestinal (eg, abdominal pain, nausea, vomiting, and constipation), followed by respiratory distress and, less frequently, angina-like chest pain.[4] In rare cases, a combination of these multiorgan symptoms are observed.[4] Gastrointestinal symptoms may range from a mild, intermittent complaint to a serious condition (eg, acute abdomen). Complications of Chilaiditi syndrome may include a volvulus of the cecum, splenic flexure, or transverse colon.[3,8-10] Cecal perforation and, rarely, perforated subdiaphragmatic appendicitis can also occur as complications of Chilaiditi syndrome.[11,12]

Chilaiditi syndrome should be considered as a rare cause of intestinal obstruction of either the large or small bowel, as reported by Mateo de Acosta Andino and associates.[13,14] However, colonic pseudo-obstruction (Ogilvie syndrome) has also been observed in patients with Chilaiditi syndrome.[15] Additionally, Chilaiditi syndrome has been associated with a variety of pulmonary or gastrointestinal malignancies (involving the colon, rectum, or stomach).[16-18]

Colonic interposition (Chilaiditi sign) is defined by the presence of air below the right diaphragm on a radiograph. To diagnose Chilaiditi sign based upon radiologic findings, the following criteria must be met: The right hemidiaphragm must be adequately elevated above the liver by the intestine, the bowel must be distended by air to illustrate pseudopneumoperitoneum, and the superior margin of the liver must be depressed below the level of the left hemidiaphragm.[7] Important differential diagnoses of this radiographic sign include pneumoperitoneum and subphrenic abscess. The finding of normal plicae circulares or haustral markings of the colon under the diaphragm can rule out these more serious entities. Moreover, changing the position of a patient with Chilaiditi sign will not change the position of the radiolucency, unlike in a patient with free air. Similarly, when using ultrasound, altering the position of a patient with Chilaiditi sign will not lead to a change in the location of the gas echo, as opposed to a patient with pneumoperitoneum.[19] If a radiograph or ultrasound cannot clearly determine whether the subdiaphragmatic air is free or intraluminal, a computed tomography scan is recommended to establish an accurate diagnosis, assuming that the patient is clinically stable.[20]

The differential diagnoses of Chilaiditi syndrome can also include bowel obstruction, volvulus, intussusception, ischemic bowel, or inflammatory conditions (eg, appendicitis or diverticulitis). However, as mentioned above, these intestinal disorders can also occur within the interposed colon in rare instances. Chilaiditi syndrome can be initially misdiagnosed as a diaphragmatic hernia.[21-23] Interestingly, there have been reports in the literature of bowel interposition accompanied by a right-sided Bochdalek hernia.[21,22] Although Chilaiditi sign is a rare entity overall, this challenging diagnosis should be considered when a patient presents with abdominal and/or respiratory symptoms and has a radiologic finding of air below the right diaphragm.

No intervention is required for an asymptomatic patient with Chilaiditi sign. When evaluating a symptomatic patient with small bowel obstruction, clinicians should first rule out the more serious condition of pneumoperitoneum. On the other hand, a misdiagnosis of bowel perforation might result in unnecessary surgical intervention. It is important to identify Chilaiditi sign in order to prevent complications from occurring during a percutaneous transhepatic procedure or liver biopsy, particularly in cirrhotic patients, who are predisposed to development of Chilaiditi sign. An interposed segment of bowel can also make it very difficult to perform a colonoscopy.[24] Colonoscopy should be performed with great caution due to the risk of progressive air entrapment in an acutely angulated, interposed bowel, which could potentially lead to perforation. Administration of carbon dioxide as the insufflating agent for colonoscopy is appropriate for decreasing this risk.[24]

Initial management of Chilaiditi syndrome should include bed rest, intravenous fluid therapy, bowel decompression, enemas, and laxatives. A repeat radiograph following bowel decompression may show disappearance of the air below the diaphragm. Thus, bowel decompression documented by a follow-up radiograph can confirm both the diagnosis of the condition and the success of the therapy, by showing the disappearance of subdiaphragmatic air and repositioning of distended intestine back to the normal position beneath the liver.

If the patient does not respond to initial conservative management, and either the obstruction fails to resolve or there is evidence of bowel ischemia, then surgical intervention is indicated. In recent years, surgical intervention has been increasingly used in order to manage symptoms of chronic, intermittent abdominal pain.[25] The appropriate surgical approach depends on the nature of the interposed segment of the colon. Cecopexy may be adequate to eliminate the possibility of recurrence in an uncomplicated cecal volvulus, unless gangrene or perforation necessitates surgical resection. However, colonic resection is the best option for a volvulus of the transverse colon, and attempts at colonoscopic reduction are not recommended due to a high frequency of gangrene (16%) in this type of volvulus.[6]

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