The liver and Chilaiditi’s syndrome: Significance of hepatic surface grooves

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Abstract
Chilaiditi’s syndrome describes a symptomatic patient with radiographic findings of interposed colon between the diaphragm and right lobe of liver. It may mimic a pneumoperitoneum on plain radiographs. We present a case in which Chilaiditi’s syndrome was entertained, delaying a decision for laparotomy. This case reinforces the diagnostic difficulty associated with Chilaiditi’s syndrome, and it increases awareness of an uncommon variation in the liver surface anatomy.

Keywords
Liver, grooves, diaphragm, slips, Chilaiditi, syndrome

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Background
Demetrios Chilaiditi described interposition of the colon between the diaphragm and right lobe of liver in 1910. This finding, when associated with abdominal pain, has become known as Chilaiditi’s syndrome. Chilaiditi’s syndrome may mimic a pneumoperitoneum on plain radiographs. As such, it is important to be aware of this diagnosis to avoid unnecessary investigations and surgery.

We present a case in which Chilaiditi’s syndrome was entertained, delaying a decision for laparotomy. This case reinforces the diagnostic difficulty associated with Chilaiditi’s syndrome, and it increases awareness of an uncommon variation in the liver surface anatomy.

Report of a case
A 59-year-old man presented to the emergency room complaining of vague abdominal pain across the upper abdomen that was present for 72 h. The pain became more intense in the hour prior to his presentation, prompting his visit to the facility. There were no other symptoms present.

On clinical examination, the abdomen was soft and mildly distended. There was mild tenderness on palpation, but guarding and rebound were not appreciated. Bowel sounds were present and normal. Apart from mild upper abdominal tenderness, the clinical examination was normal.

The white cell count was noted to be 10 × 106/dL. Due to institutional limitations, C-reactive protein (CRP) assays were not available for clinical use in this case. The remaining blood results were normal. There was the impression of air above the right liver on plain erect radiographs of the abdomen (Figure 1). However, the pockets of air were limited by haustra, prompting a diagnosis of Chilaiditi’s syndrome. A multi-row detector computed tomography (CT) scan was ordered. Resuscitation was commenced, while awaiting CT scans, with intravenous hydration, nasogastric decompression and serial abdominal examinations.

The same clinician re-examined the abdomen 6 h later, prior to CT scans being completed and noted that the abdomen remained mildly distended, but there was now increased upper abdominal tenderness associated with guarding. Therefore, consent was secured for laparoscopic exploration, and the patient was taken to the operating room.

The abdomen was entered using Hasson’s technique. Immediately, free air and fresh bowel content were evacuated from the peritoneal cavity. Inspection at the upper abdomen revealed the presence of hepatic surface grooves and
associated diaphragmatic slips (Figure 2). Further inspection revealed that the source of contamination was free perforation of a peptic ulcer at the first part of the duodenum (Figure 3). The ulcer was debrided and repaired primarily with sutures. The closure was water-tight when leak-tested with air via a nasogastric tube. A vascularized flap of omentum was used to cover the repair as a modified Graham’s patch. There was an uneventful recovery after operation. Oral intake was recommenced on day 3, and the patient was discharged, without complication, on day 5.

Discussion

After its description in 1919, Chilaiditi’s sign was recognized as a differential for a pneumoperitoneum. When associated with symptomatology, the condition is then known as Chilaiditi’s syndrome. It is an uncommon syndrome, with a reported worldwide incidence of 0.25%–0.28% and a 4:1 male preponderance.

Although the cause remains unknown, several theories have been touted including congenital meso-colonic redundancy, laxity of hepatic suspensory ligaments, liver atrophy associated with cirrhosis, a high diaphragm after phrenic nerve injury and capacious sub-diaphragmatic spaces that may occur after hepatectomy, ascites or excessive weight loss.

No specific surgical management is required for Chilaiditi’s syndrome. It is usually treated conservatively with analgesics, bowel decompression, intravenous fluids and laxatives, if necessary. Since Chilaiditi’s sign closely mimics a pneumoperitoneum on plain radiographs, it is important for clinicians to be able to make the diagnosis in order to avoid unnecessary laparotomy.

A co-existing danger is the under-diagnosis of serious abdominal pathology in favour of Chilaiditi’s syndrome. This occurred in our case and, although the clinical outcomes remained good, this error could have easily led to grave consequences if the clinicians persisted with the diagnosis. This reinforces the principle that clinical assessments should supersede findings on imaging.

Unfortunately, the diagnosis is often challenging because Chilaiditi’s syndrome is associated with non-specific symptoms, such as vague abdominal pain, anorexia, nausea, vomiting, constipation and/or respiratory difficulty. For this reason, many authorities advocate liberal use of diagnostic CT scanning when a diagnosis of Chilaiditi’s syndrome is entertained.
In this case, we were not able to complete CT scanning in a reasonable time frame. However, we agree fully with the liberal use of diagnostic CT scans because there are no features on plain radiographs that can differentiate Chilaiditi’s sign from a true pneumoperitoneum. Luijkx and Jones suggested that the presence of rugal folds within the gas, suggesting that it is within the bowel and not free was diagnostic of Chilaiditi’s syndrome. However, rugal folds were clearly present in our case despite this being a true pneumoperitoneum. In this case, the ‘rugal folds’ demonstrated in Figure 1 were due to hepatic surface grooves. These are abnormal, permanent vertical depressions that appear on the surface of the liver. The reported incidence of hepatic surface grooves varies by location from 5% in Malaysia to 25% in the United States and 40% in Italy. In our population, they are seen in 12% of unselected persons.

The aetiology of hepatic surface grooves remains uncertain, but the prevailing theory proposed by Macchi et al. theorized that there are weak zones on the surface of the liver that offer low resistance to external pressure. Any source of pressure could produce permanent depressions on the liver surface. The presence of fibrous bands between the diaphragmatic muscle fibres, known as diaphragmatic slips, could be a potential source of external pressure. These diaphragmatic slips are often found on the right hemidiaphragm, with their concavity parallel to the falciform ligament, just as seen in this case. The presence of diaphragmatic slips and associated hepatic grooves in this case created the ‘indented’ appearance of free air on the plain radiographs. This very closely mimicked the appearance that Luijkx and Jones suggested which was pathognomonic of Chilaiditi’s sign. This reinforces our assertion that there are no pathognomonic signs on plain radiographs.

This case adds to the world literature since it highlights the fact that a true pneumoperitoneum contained between diaphragmatic slips and corresponding hepatic surface grooves can very closely mimic Chilaiditi’s syndrome. A true pneumoperitoneum, in the presence of diaphragmatic slips and corresponding hepatic surface grooves, can easily be confused with Chilaiditi’s syndrome. This association should now be added to the list of clinically significant effects of surface grooves. In these cases, supporting investigations such as CRP assays and diagnostic CT scanning should be performed liberally to ascertain the diagnosis.

Conclusion

A true pneumoperitoneum, in the presence of diaphragmatic slips and corresponding hepatic surface grooves, can easily be confused with Chilaiditi’s syndrome. This association should now be added to the list of clinically significant effects of surface grooves. In these cases, supporting investigations such as CRP assays and diagnostic CT scanning should be performed liberally to ascertain the diagnosis.

Declaration of conflicting interests

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Ethical approval

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Informed consent

Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

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