

Case Report

Chilaiditi's Syndrome in Children: A Case Series of a Rare Cause of Abdominal Pain

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Abstract

Chilaiditi's syndrome is a rare cause of abdominal pain that due to its highly variable presentation it is mostly diagnosed as an incidental finding while performing ancillary studies.

We present 3 cases of Chilaiditi's syndrome in paediatric patients. First case is a 13-year-old patient consulting initially for diffuse chest and abdominal pain of approximately 3 weeks. Chilaiditi's syndrome was diagnosed radiologically with an X-ray, he was managed conservatively without any complications developed further on.

The second case is a 19-year-old patient in follow-up by our Gastroenterology unit for Crohn's disease that during a control MRI an interposition of the colon between the splenic flexure and the diaphragm was discovered (Chilaiditi's sign).

The last patient is a 4-year-old boy who, in study of a fever associated with tachypnoea a chest X-ray was performed showing Chilaiditi's sign.

Keywords: Abdominal pain; Bowel obstruction; Chilaiditi's syndrome; Chilaiditi's sign; Colon interposition; Constipation

Introduction

Chilaiditi's syndrome is a benign condition in which a segment of the intestine (usually right segment of colon) is interposed between the liver and the diaphragm [1]. This can cause gastrointestinal symptoms, which can be confused with other more serious pathologies, resulting in unnecessary procedures [2].

When a patient develops symptoms, it is categorized as Chilaiditi's syndrome, while if it's asymptomatic it is denominated Chilaiditi's sign [1].

Its name comes from Demetrius Chilaiditi, a Greek radiologist who described it for the first time in 1910 [1,3,4]. It has a worldwide incidence of 0.025%-0.28% (rare), being more frequent in men than in women, with a 4:1 ratio [1,3,5]. Since incidence is higher in adults, there are not sufficient studies that can estimate the real incidence in children [4,6]. We present 3 cases of this finding, two of them asymptomatic and the other presenting abdominal pain.

Case Presentation

Case 1

A 13-year-old patient previously followed by the Paediatric Neurology Unit for autism and macrocephaly (with initial normal ancillary studies) consulted for diffuse chest and abdominal pain of approximately 3 weeks.

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Due to this acute symptomatology, an anterior-posterior and lateral chest X-rays were performed, exhibiting an interposition of the colon between the liver and right hemidiaphragm (Figure 1).

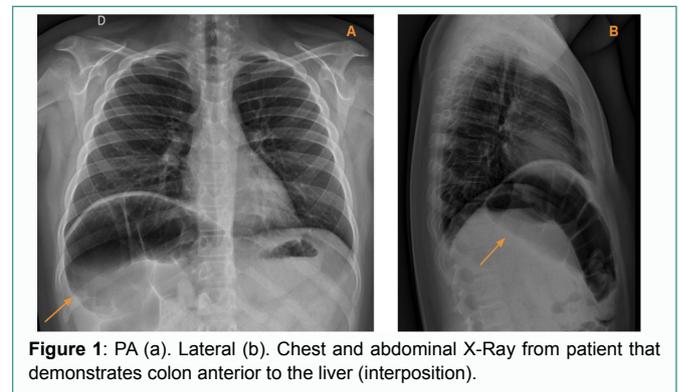


Figure 1: PA (a). Lateral (b). Chest and abdominal X-Ray from patient that demonstrates colon anterior to the liver (interposition).

Clinical examination revealed an obese patient (BMI of 27.4) and a globular abdomen without abdominal pain. Therefore, in view of these findings, Chilaiditi's syndrome was diagnosed and managed conservatively without any complications developed further on.

Case 2

The second case report is a 19-year-old patient with Crohn's disease that during a control MR Enterography an interposition of the colon between the splenic flexure and the diaphragm was discovered (Chilaiditi's sign).

An assessment of previous MR revealed indirect signs that the interposition was present previously (Figure 2). Since he was asymptomatic and no clinical findings on the physical examination were found, we decided on a conservative treatment. No complications developed through follow-up and the interposition resolved in subsequent studies.

Case 3

The third and last case is a 4-year-old boy who consulted our

emergency department for presenting fever for 3 days associated with mild cough, rhinorrhoea and vomiting. The patient presented tachypnoea and mild right hypophonesis. An X-ray was performed showing the Chilaiditi's sign (Figure 3).

The patient was diagnosed with an upper respiratory infection and due to lack of gastrointestinal symptoms we decided a conservative management.



Figure 2: Coronal images of the patient's abdominal CT showing interposition of colon between liver and diaphragm.

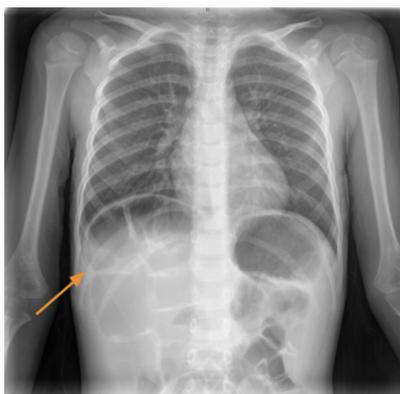


Figure 3: Chest and abdominal X-Ray from a patient that demonstrates colon anterior to liver (interposition).

Discussion

This finding consists in an interposition of a segment of the bowel between the liver and the diaphragm in most cases, although it has been seen between two other organs (diaphragm and spleen, etc.).

Under normal conditions, human beings have suspensory ligaments that prevent the interposition of the colon with the diaphragm and the liver. Anatomical variants such as an increase in ligamentous laxity, elongation of the ligaments of the transverse or falciform colon or their absence, can predispose to this interposition [4,6-9].

Furthermore, other predisposing factors have been studied as congenital malformations, aerophagia (these first two of relevance in children), functional disorders (specifically chronic constipation due to colonic elongation), colonic distension, causes of high intra-abdominal pressure (ascites, multiple pregnancies, ...), cirrhotic liver or hepatectomy (decreasing hepatic size), etc [3,5,6].

Its clinical presentation is highly variable. The most common

clinical presentation is abdominal pain but other symptoms may be nausea, vomiting, constipation, respiratory distress, chest pain, arrhythmias, or patients may even be asymptomatic [3,6,7]. Patients explain nocturnal worsening of symptoms due to supine decubitus. Abdominal distension, sometimes associated with disappearance of hepatic dullness and presumed hepatomegaly (since the colon rejects the liver downwards) can be found [6,10].

Imaging tests (abdominal or chest X-ray or abdominal CT scan) are necessary for the diagnosis of this entity. The 2 characteristic radiological findings are the elevation of the right hemidiaphragm and the bowel interposition [2,6,9].

Since the differential diagnosis is vast, delays in diagnosis and under-diagnosis are frequent. The differential diagnosis include: pneumoperitoneum (a change in the air's location would be seen in an ultrasound when asking the patient to move), subphrenic abscess (ruled out by the display of the intestinal circular folds or colon haustra below the diaphragm), intestinal pneumatosis and others (especially diaphragmatic hernia and acute renal colic) [5,7,9,11].

In most cases, the treatment is usually conservative with symptomatic treatment for constipation [3-5,9]. When conservative treatment fails, surgical treatment could be an option. Colopexy normally prevents the colon from ascending again by attaching it to the abdominal wall [12-14].

Since it is a benign entity, the prognosis of the patients is determined by its possible complications. Bowel obstruction due to intestinal volvulus (these patients have a higher risk than the rest of the population), mesenteric ischemia or bowel perforation are the most important to consider [2,4,7,12,14].

Conclusion

Chilaiditi's syndrome is usually an incidental finding on imaging techniques. Its exact aetiology is unknown, but many factors have been related to it, being the ligamentous laxity, constipation and the congenital malformations the most important in children.

Due to its wide range of clinical presentations, radiology is essential for the diagnosis. Since it is often confused with other pathologies that lead to necessary surgical interventions, a correct differential diagnosis is required. The vast majority of patients will benefit from a conservative treatment, with great prognosis and infrequent complications.

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