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Congenital partial diaphragmatic eventration presenting with Chilaiditi's sign: a case report

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Abstract

Background Chilaiditi's sign is an incidental radiographic finding, associated with intestinal disposition located between liver and right diaphragm. It is considered as an acquired rather than a congenital condition and the prevalence ranges from 1.18% to 2.4% according to recent adult retrospective studies. The aspects of this rare entity with regards to a 7-month-old male initially misdiagnosed as diaphragmatic hernia is discussed.

Case presentation A 4-month-old Caucasian male was misdiagnosed with a congenital diaphragmatic hernia owing to previous hospitalization with complaints of respiratory tract infection. On admission 3 months later, he was free of any signs and symptoms of intestinal obstruction or respiratory distress. Thorax computed tomography revealed Chilaiditi's sign. A diagnostic laparoscopy was regarded necessary to evaluate the anatomical details. The most prominent finding was the lack of muscle fibers and almost transparent appearance of the medial aspect of the partially eventrated right hemidiaphragm. Owing to delicate anatomical presentation, diaphragmatic plication was considered hazardous. The patient is doing well and under follow-up.

Conclusions It is obvious that Chilaiditi's sign is not always a completely incidental finding of no consequence, and may indicate an underlying congenital diaphragmatic pathology, clearly defined by laparoscopic evaluation in this case.

Keywords Chilaiditi pediatric, Chilaiditi's syndrome pediatric, Laparoscopy

Introduction

Chilaiditi's sign is an incidental radiographic finding observed during routine chest and abdominal radiographs, associated with asymptomatic hepato-diaphragmatic interposition of the intestine. The prevalence has long been reported to range from 1.18% to 2.4% and considered as an acquired rather than a congenital condition [1, 2]. The differential diagnoses include pneumoperitoneum, subphrenic abscesses, and diaphragmatic hernias. We present a young patient and discuss the aspects of this rare entity during infancy.

Case report

A 7-month-old Caucasian male was admitted to our department in Anadolu Medical Center—Istanbul, for a second opinion with complaint of an incidental diagnosis of right diaphragmatic hernia, at the age of 4 months during an episode of cough, cyanosis, and respiratory distress at another referral center (Fig. 1). His symptoms had improved with antibiotic therapy and thrived without recurrence. Upon physical examination, he was a completely healthy infant, free of any symptoms of intestinal obstruction or respiratory distress, whole blood

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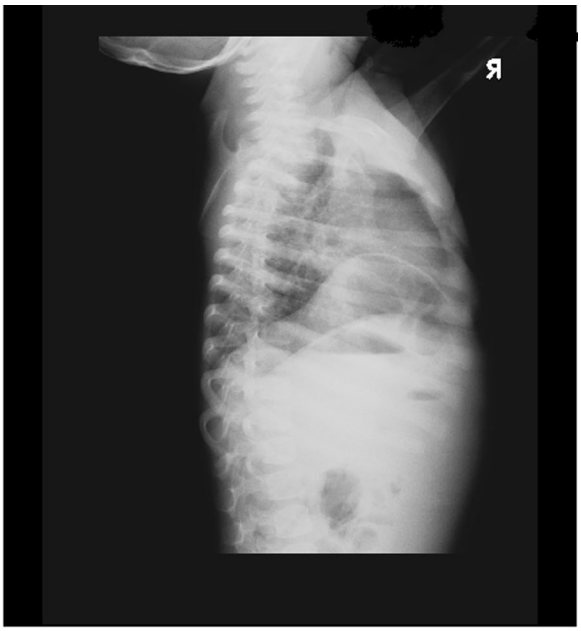

Fig. 1 Hemi diaphragmatic elevation diagnosed as hernia

count and biochemistry measurements were within normal limits. His past medical history revealed a full-term infant, born after uncomplicated pregnancy, labor, and delivery.

To obtain an improved quality three-dimensional (3D) image of the lesion, a 16-detector row computed tomography (CT) was undertaken, which confirmed the Chilaiditi's sign. A dilated bowel loop (hepatic flexure) interposed between diaphragm and liver was observed. There was minimal superolateral displacement of heart. Liver anatomy was normal. The right hemidiaphragm was elevated anteriorly showing no sign of diaphragmatic defect (Fig. 2). Fluoroscopic imaging of diaphragm showed normal diaphragmatic movements with respect to inspiration and expiration.

The parents noticed a great deal of discomfort mainly aroused by the initial diagnosis of diaphragmatic hernia and insisted further evaluation. Parental consent was obtained to perform a diagnostic laparoscopy to evaluate the anatomical details. Under elective laparoscopic examination, one-half the length of the transverse colon was found lying anteriorly in the space between the hemidiaphragm and the right lobe of liver without any anomalous fixation. The most prominent finding was the lack of muscle fibers and almost transparent appearance of the medial aspect of the right hemidiaphragm (Fig. 3). Any surgical correction was regarded as unnecessary and hazardous due to present delicate diaphragmatic anatomy. The patient is followed-up for 11 months and is doing well. His chest x-ray reveals the same findings with no remarkable change.

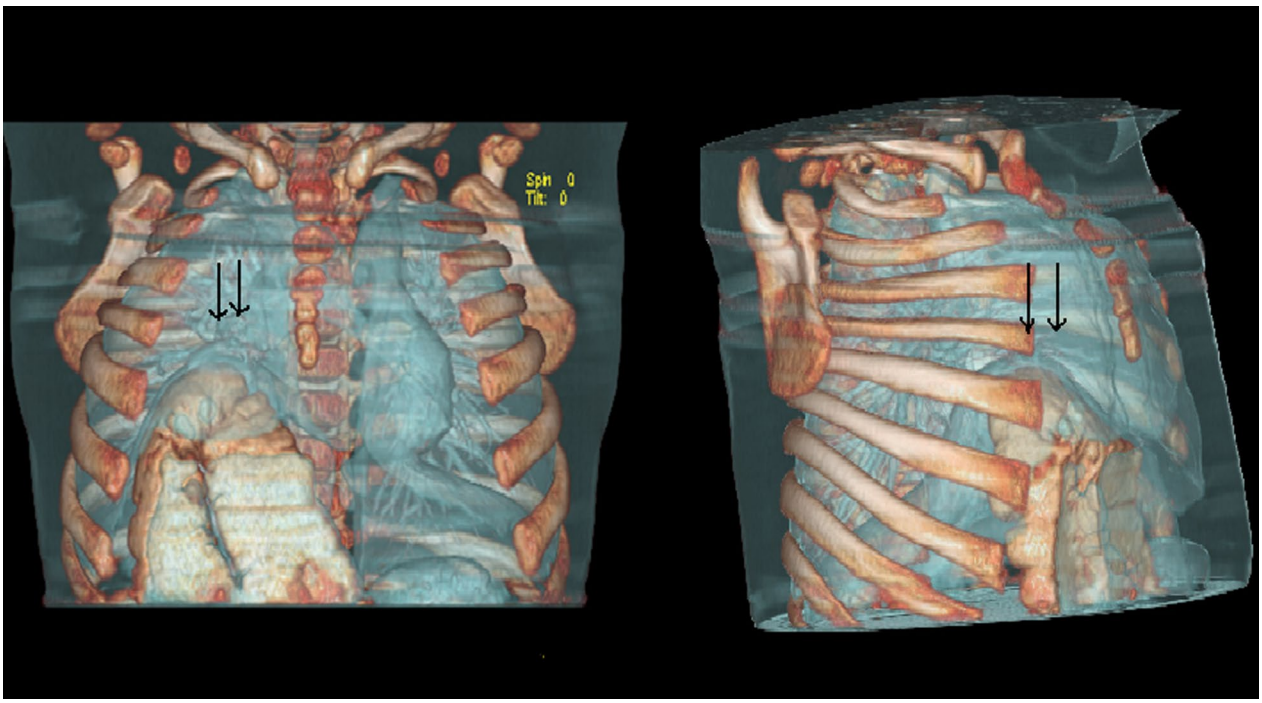

Fig. 2 Coronal three-dimensional reconstruction of contrast-enhanced computed tomography demonstrates colonic loop with its mesenteric fat and vessels ascending along the right anterior perihepatic space (arrows). Right hemidiaphragm is elevated; no sign of defect is present

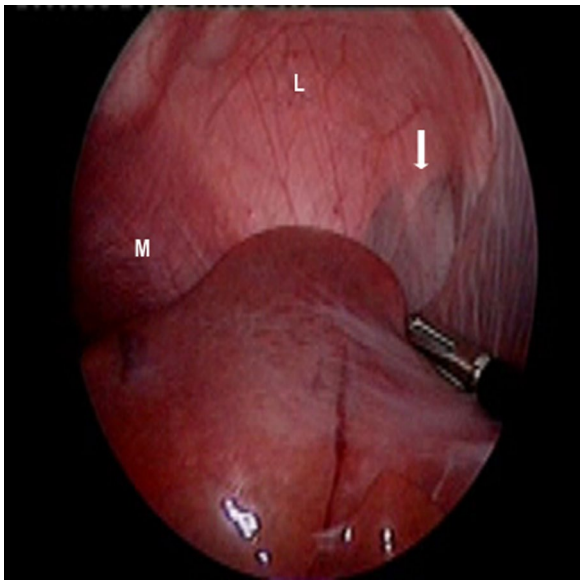

Fig. 3 Laparoscopic view of absence of muscle fibers (clear view of the lung) and almost transparent appearance (large arrow) of the medial aspect of the right hemidiaphragm. M displays appearance of the normal diaphragmatic muscle

Discussion

The incidental radiological finding of hepato-diaphragmatic interposition of bowel or Chilaiditi's sign is first described in 1910 by Demetrius Chilaiditi ("Ky-La-Ditty") [3]. An extensive search of the literature reveals that it is increasingly documented as an important imitator of numerous diaphragmatic disorders and abdominal emergencies [3–6]. Chilaiditi's sign is four times more common in males and much more common in the elderly. The reported incidence has been increasingly published as 1.18%–2.4% with regards to recent retrospective studies [1].

The subdiaphragmatic intraabdominal air may mimic perforated viscus, a subphrenic abscess, posterior hepatic lesions, Morgagni's hernia, and traumatic diaphragmatic hernia [6, 8–10]. Abdominal computed tomography and ultrasound are used to establish a definitive diagnosis [1, 2, 4, 7]. Diagnosis is dependent on radiological findings of the elevated hemidiaphragm and a depressed liver by the distended bowel in either chest or abdominal radiographs. The visible haustration or images of circular plicas of the colon are the key points in differentiating the image from free air. Our case was initially misdiagnosed as congenital diaphragmatic hernia. We managed to delineate the exact anatomy in the infant using diagnostic laparoscopy, avoiding an unnecessary explorative laparotomy.

Chilaiditi's sign is considered an acquired rather than a congenital condition mostly owing to published reports concerning elderly patients [2]. These acquired conditions in the elderly include decreased liver volume owing to cirrhosis, fulminant hepatic failure or resection, ascites, or high abdominal fat content, aerophagia, constipation, traumatic diaphragmatic paralysis, and chronic lung disease [2, 4, 6, 8–12]. The true incidence of this incidental finding as a congenital anomaly is unknown. On the other hand, it is obvious that congenital conditions, such as malrotation or malposition of the colon, elevation, paralysis, or weakness of the hemidiaphragm, as well as laxity of the hepatic suspensory ligaments, and enlargement of the thoracic cage diameter, may result in the clinical picture [4, 9, 10, 13]. The elevation of the diaphragm owing to the weakness of its muscular content may partly bring some explanation in a spectrum of diaphragmatic disorders, with regards to the pathophysiology in our patient. This sign may be a precursor in some cases to diaphragmatic eventration.

Asymptomatic patients presenting with incidental radiologic finding of Chilaiditi's sign must be distinguished from Chilaiditi's syndrome, which produces symptomatology associated with indistinct abdominal pain, distention, vomiting, anorexia, and constipation. Saber and Boros have pointed out a higher incidence of combination of intellectual disabilities with long-standing constipation in their review of literature [4]. Most cases may require only symptomatic treatment, such as bowel rest or decompression, or enemas for constipation. A variety of clinical conditions, including transverse, sigmoid and splenic volvulus, chest trauma, chest tube complication, suprahepatic appendicitis, and intestinal obstruction caused by interposition of small bowel may explain the various aspects of this rare but important entity [1, 4, 6, 8, 11, 13, 14]. Constipation, colonic elongation, and colonic hypermobility are said to provoke the clinical picture in patients with Chilaiditi's syndrome. Patients unresponsive to nonoperative management may require surgery when a complication, such as volvulus or obstruction, develops. Colectomy, colopexy, and hepatopexy have been described as the choice of treatment, as well recent laparoscopic approaches in published reports [8, 13–16].

Pediatric cases may present complaints of cough, cyanosis, and recurrent respiratory distress or cardiac arrhythmias [9, 10]. The presence of the dilated colonic loop related elevation of the diaphragm may be responsible for the respiratory symptoms. The resolution is usually spontaneous in pediatric patients under conservative treatment. Our case had initially been misdiagnosed to suffer from a right diaphragmatic hernia at the age of 4 months. He has not had any recurrent respiratory tract infection during the follow-up period

since he began treatment with stool softeners. The elevation of the right hemidiaphragm, however, has not disappeared.

Conclusion

We believe that Chilaiditi's sign is not always a completely incidental finding of no consequence, but may be an indicator of underlying diaphragmatic pathology, as in this case. Diagnostic laparoscopy may aid in better understanding the clinical picture, including the anatomic variations of the diaphragm in selected cases. It is best to avoid unnecessary surgical correction in a child unless the patient is unresponsive to nonoperative management.

Supplementary Information

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Additional file 1.

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Author contributions
B.H.G and K.R performed the operation; B.H.G. designed and wrote the manuscript; K.R. gave technical support and conceptual advice. All authors read and approved the final manuscript.

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Availability of data and materials
The data that support the findings of this study are available from Dr. B. Haluk GUVENC, but restrictions apply to the availability of these data, which were used under license for the current study, and so are not publicly available. Data is, however, available from the authors upon reasonable request and with permission of Dr. B. Haluk GUVENC.

Declarations

Ethical approval and consent to participate
Our manuscript does not report on or involve the use of any human data or tissue. Written informed consent was obtained from the patient's legal guardian for publication of this case report and any accompanying images.

Consent for publication
Written informed consent was obtained from the patient's legal guardian for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests
B.H.G. and K.R. have no conflicts interest or financial ties to disclose.

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