CASE REPORTS

UDC: 616.24-06:616.34
DOI: 10.2298/VSP140603052Z

Chilaiditi's sign and syndrome: theoretical facts and a case report

Chilaiditi-jev znak i sindrom

Biljana Zvezdin*†, Nevena Savić*†, Sanja Hromiš*†, Violeta Kolarov*†, Djordje Taušan‡, Bojana Krnjajić*†

***Institute for Pulmonary Diseases of Vojvodina, Sremska Kamenica, Serbia; †Faculty of Medicine, University of Novi Sad, Novi Sad, Serbia; ‡Pulmology Clinic, Military Medical Academy, Belgrade, Serbia**

Abstract

Introduction. Chilaiditi's syndrome is a rare condition manifested by gastrointestinal symptoms, and radiologically verified by transposition of the large intestine loop. This radiological finding with no manifested symptoms is termed the Chilaiditi's sign. The aim of this case report was to remind the clinicians of the possibility of this rare syndrome, whose symptoms and signs may be misinterpreted and inadequately treated, with consequent diverse complications. **Case report**. We presented the theoretical facts and a patient in whom the diagnosis of Chilaiditi's syndrome was established incidentally, when hospitalized for an exacerbation of his chronic obstructive pulmonary disease. The Chilaiditi's sign was verified as an incidental finding on chest X-ray performed to evaluate the primary disease. **Conclusion**. Chilaiditi's syndrome is a benign condition which rarely requires surgery. Its clinical importance lies in adequate differential diagnostic approach and timely management of potentially serious complications.

Key words:
chilaiditi syndrome; diagnosis; diagnosis, differential; radiography; tomography, x-ray computed.

Apstrakt

Uvod. Chilaiditi-jev sindrom je retko stanje koje se manifestuje gastrointestinalnim simptomima, a radiološki se potvrđuje pozicioniranim vijugama creva u prostoru između jetre i desne hemidijafragme. Postojanje ovakvog radiološkog nalaza i odsustvo simptoma naziva se Chilaiditi-jev znak. Cilj rada bio je podsećanje kliničara na mogućnost postojanja tog retkog sindroma, koje može rezultirati pogrešnim tumačenjem simptoma i znakova bolesti, neadekvatnim lečenjem i različitim komplikacijama. **Prikaz bolesnika**. Prikazane su teorijske činjenice i bolesnik kod koga je dijagnoza Chilaiditi-jev sindrom postavljena prilikom hospitalizacije zbog pogoršanja hronične opstruktivne bolesti pluća. Chilaiditi-jev znak potvrđen je kao slučajni nalaz na radiogramu grudnog koša načinjenom u cilju procene osnovne bolesti. **Zaključak.** Chilaiditi-jev sindrom je benigno stanje koje retko zahteva hiruršku intervenciju. Klinički značaj tog sindroma ogleda se u adekvatnom diferencijalnodijagnostičkom pristupu i pravovremenom zbrinjavanju mogućih ozbiljnih komplikacija.

Ključne reči:
chilaiditi sindrom; dijagnoza; dijagnoza, diferencijalna; radiografija; tomografija, kompjuterizovana, rendgenska.

Introduction

Chilaiditi's syndrome is a very rare condition, characterized by the presence of different symptoms due to either temporary or permanent malposition of the colon loops in between the liver and the right hemidiaphragm [1–9]. This radiological finding accompanied with no symptoms is called Chilaiditi's sign [1, 3–9]. The etiology of this anatomic anomaly is still unknown, and considered multifactorial. The syndrome is manifested by isolated or concurrent symptoms – gastrointestinal, pulmonary, and/or cardiac, of different intensity and frequency [4]. The differential diagnosis includes a variety of diseases of numerous organ systems, primarily the pneumoperitoneum [4, 5]. Treatment is usually conservative, or surgical in case of complications [4–9].

Case report

A old-75-year patient, was admitted to the Institute for Pulmonary Diseases of Vojvodina, Sremska Kamenica, Serbia, due to exacerbation of his chronic obstructive pulmonary disease, outpatiently treated for over twenty years. Having responded poorly to the intensified ambulatory desobstruction treatment, the patient was referred to hospital. He was presented with a variety of comorbidities, including arterial hypertension, atrial fibrillation, valve defect, abdominal her-

Correspondence to: Biljana Zvezdin, Institute for Pulmonary Diseases of Vojvodina, Institutski put 4, 21 204 Sremska Kamenica, Serbia. Phone: +381 21 4805 128. E-mail: biljanazvezdin@yahoo.com

nia, cholelithiasis, prostatic hyperplasia, degenerative spinal disease, depression. The patient complained on the symptoms of cough, dyspnea, fatigue, heart palpitations and arrhythmia, occasional pains below the right rib arch, and dyspeptic problems. On admission, the patient was conscious, oriented, exhausted, moving with difficulties, afebrile, dyspnoic, normocardic, normotensive, with no signs of cardiac decompensation, giving the impression of a moderately severe patient. On auscultation, bronchospasm signs were registered. The abdomen was at the level of the chest, soft on palpation, with a reponible hernia of the anterior abdominal wall, and a mildly painful, sensitive epigastric region, audible peristalsis, and no signs of meteorism or ascites. The chest X-ray finding was presented with bilateral striped paracardial lesions, adhering hemidiaphragms, and the presence of an air collection below the right hemidiaphragm (Figure 1).

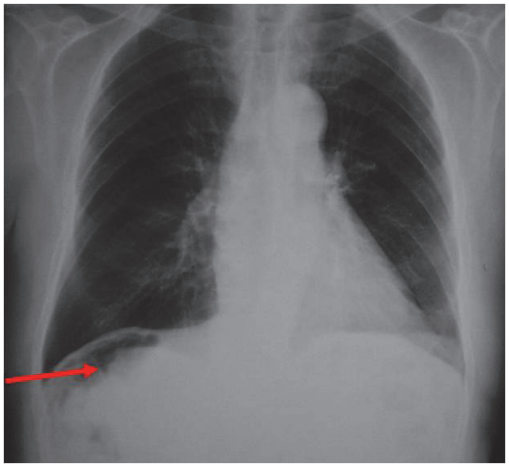

Fig. 1 – The chest x-ray reveals air collection below the right hemidiaphragm.

The pulmonary gas exchange at rest was preserved, and bronchoobstruction was verified on spirometry. Standard laboratory test findings were within normal ranges. The bacteriological sputum finding was normal as well. Computed tomography (CT) of the chest excluded the presence of infiltrative lesions in the pulmonary parenchyma, verified bilateral bronchiectases, bilateral excrescences in the basal pleura, degenerative spinal lesions, and the Chilaiditi's sign (Figure 2).

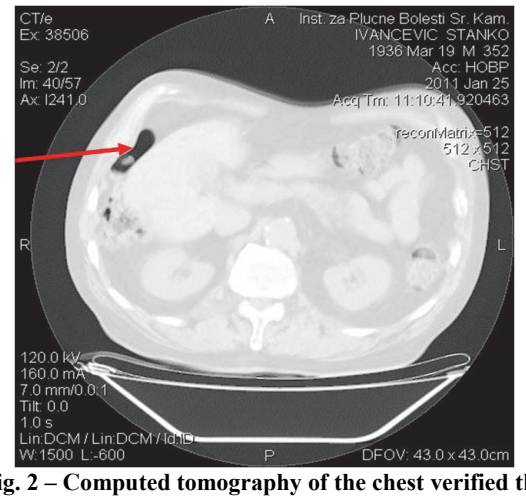

Fig. 2 – Computed tomography of the chest verified the Chilaiditi's sign (arrow).

Echocardiography verified ejection fraction of 50%, paradoxical septal movements, initial concentric hypertrophy of the left ventricle myocardium, sclerosis of aortic valves, mitral regurgitation 3+. The treatment included inhalant and parenteral desobstructive therapy, gastroprotective medication, peroral anticoagulant therapy and other formerly prescribed cardiological therapy. The gastroenterologist was consulted, who recommended conservative treatment measures and advised to consult the abdominal surgeon in case of an acute exacerbation of gastrointestinal symptoms. The applied treatment improved the patient's general and respiratory condition, and he was discharged with recommendations for further ambulatory treatment, and instructed what to do in case of exacerbated respiratory and gastrointestinal symptoms.

Discussion

Chilaiditi's sign is an incidental radiological finding, presented as a crescent lucency below the diaphragm on the right, occurring due to malpositioned loops of the colon and/or small intestine.[5] It was for the first time described by Cantini in 1865 but the first three case reports were published by the Greek radiologist Demetrius Chilaiditi in 1910, after whom this condition has been named [2, 3]. Its incidence ranges from 0.025% to 0.028% [3]. In males and the elderly, it is registered four times as frequently as in other population groups, the incidence amounting to around 1% [3, 4]. Being an asymptomatic condition, its diagnosis is established incidentally, on the occasion of different radiological examinations of the chest or abdomen (CT, standard chest X-ray, or ultrasound) [7].

Predisposing factors include all the conditions resulting in the increased right subphrenic space or intestinal hypermobility. These factors may be classified as congenital or acquired (Table 1) [1, 5, 7–9], diaphragmatic, intestinal, hepatic, and others (Table 2) [4].

Table 1
Congenital and acquired predisposing factors for Chilaiditi's sign and syndrome [1, 5, 7–9]

Predisposing factors	
Congenital	Acquired
Absence of: - suspensory ligament (*ligamentum suspensorium duodeni*) - falciform ligament (*ligamentum falciforme hepatis*) Malposition of abdominal organs Dolichocolon Right hemidiaphragm paralysis *Laxitas* (weak connective tissue)	Chronic obstipation Fecal impaction Cirrhosis with liver atrophy Ascites Aerophagia Obesity Right hemidiaphragm paralysis COPD Multiple pregnancies Colonoscopy Pregnancy

COPD – chronic obstructive pulmonary disease.

The diagnosis is established on the basis of the chest X-ray finding presented with the following three features [5, 7]: elevation of the right hemidiaphragm; a crescent lucency be-

low the diaphragm on the right (the air-distended intestine – "pseudoperitoneum"); shadow of the upper liver line below the right hemidiaphragm.

Table 2
Diaphragmative, hepatic, and intestinal predisposing factors for Chilaiditi's sign and syndrome [4]

Predisposing factors
Diaphragmatic
Elevated right hemidiaphragm due to:
muscular degeneration
injured *nervous phrenicus*
Hepatic (reduced liver size)
cirrhoses
right lobe agenesis
ptoses
absent or weak suspensory ligaments
Intestinal
abnormal motility of the intestines
long intestines with long mesentherium
absence of peritoneal ligaments
malrotation of congenital malposition of the intestines

Most frequently, there is a malposition of the hepatic flexure of the colon, ascending or transversal colon, more rarely of the cecum, independently or in combination with the small intestine [4]. Depending on the position of the intestines in relation to the liver, the anterior and posterior type are differentiated [4], which may be either temporary or permanent [6, 7, 9].

Chilaiditi's sign is easily diagnosed, analyzing the standard chest X-ray finding. CT is the imaging technique of choice here, as it concurrently excludes a rupture of the diaphragm, intestinal perforation, congenital malformations, as well as other conditions and diseases [7–9]. The differential diagnosis includes renal or biliary colics, subphrenic abscess, pneumoperitoneum, or congenital diaphragmatic hernias [1, 5–7]. The most important radiologic indicators excluding these serious complications, particularly pneumoperitoneum, are the presence of the intestinal *haustrum* and the persisting pseudoperitoneum position at changing bodily postures.

The timely diagnosis is important to prevent the complications which may arise while performing various diagnostic procedures, including percutaneous liver biopsy, pleural puncture, and colonoscopy [4, 5].

Chilaiditi's syndrome is a rare condition manifested by diverse gastrointestinal symptoms, and a radiologically verified Chilaiditi's sign. It occurs very rarely, in elderly males four times as frequently as in elderly females, and the cases in children have also been reported [4]. It is manifested by the symptoms differing in intensity and frequency – abdominal pains, flatulence and "pouring" in the bowel, nausea, vomiting, altered discharge habits, more rarely retrosternal pains, heart arrhythmia, dyspnea, and respiratory distress [4, 5]. Different diseases and conditions may be additionally prolonged if accompanied with this syndrome; these most often include chronic obstructive pulmonary disease, sclerodermia, congenital hypothyroidism, paralytic ileus, *melanosis coli*, mental retardation, more rarely lung and colon cancer, bariatric surgery, gastric probe placement, colonoscopy [4].

The treatment of the syndrome is initiated by conservative measures including rest, rehydration, high content of plant fibers in the diet, nasogastric decompression, laxatives and/or antiemetics [7]. If the conservative treatment fails to result in adequate clinical and/or radiologic improvement or obstruction, ischemia or perforation are suspected, surgical treatment should be carried out [4, 7]. Cases requiring urgent surgery were rarely reported in the literature, which included volvulus of the cecum or colon, subphrenic appendicitis, intestinal perforation, intraabdominal herniation [4, 9]. No unique attitude to the most adequate surgical approach has been formulated yet. Invasive surgeries, including colon resection, hepatopexy, colonopexy, right hemicolectomy, sigmoidectomy, and subtotal colectomy have been successfully carried out, as well as less invasive laparoscopic colonopexies [4, 7, 9].

Conclusion

Chilaiditi's syndrome is a benign condition which rarely requires surgery. The clinical relevance of Chilaiditi's syndrome lies in the possibility to recognize and prevent various complications causing acute abdominal symptoms, such as obstruction, perforation or ischemia of the intestines.

REFERENCES

1. *Kamiyoshihara M, Ibe T, Takeyoshi I.* Chilaiditi's sign mimicking a traumatic diaphragmatic hernia. Ann Thorac Surg 2009; 87(3): 959−61.
2. *Chilaiditi D.* Zur frage der hepatoptose und ptose im allgemeinen im anschluss an drei fälle von temporärer, partieller leberverlagerung. Fortschritte auf dem Gebiete der Röntgenstrahlen 1910; 16(1): 173–208. (German)
3. *Glatter RD, April RS, Miskovitz P, Neistadt LD.* Severe recurrent abdominal pain: an anatomical variant of Chilaiditi's syndrome. MedGenMed 2007; 9(2): 67.
4. *Yin AX, Park GH, Garnett GM, Balfour JF.* Chilaiditi syndrome precipitated by colonoscopy: a case report and review of the literature. Hawaii J Med Public Health 2012; 71(6): 158−62.
5. *Moaven O, Hodin RA.* Chilaiditi syndrome: a rare entity with important differential diagnoses. Gastroenterol Hepatol (N Y) 2012; 8(4): 276−8.
6. *Okus A, Ay S, Carpraz M.* Chilaiditi Syndrome. Eur J Gen Med 2013; 10(2): 79−82.
7. *Kang D, Pan AS, Lopez MA, Buicko JL, Lopez-Viego M.* Acute Abdominal Pain Secondary to Chilaiditi Syndrome. Case Rep Surg 2013; 2013: 756590.
8. *Tuncer M, Sahin C, Yazici O, Kafkash A, Sarica K.* A rare cause of renal colic pain: Chilaiditi syndrome. Arch Ital Urol Androl 2014; 86(3): 229−30.
9. *Williams A, Cox R, Palaniappan B, Woodward A.* Chilaiditi's syndrome associated with colonic volvulus and intestinal malrotation: a rare case. Int J Surg Case Rep 2014; 5(6): 335−8.

Received on June 3, 2014.
Revised on March 9, 2015.
Accepted on March 10, 2015.
Online First July, 2015.