CASE REPORT

Chilaiditi Syndrome in Two Cases Presented with Respiratory Distress Symptoms

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Abstract

Chilaiditi syndrome is a rare syndrome known hepatodiaphragmatic interposition of as colon or small intestine. As this pathology is usually asymptomatic, it is incidentally identified in adults and rarely reported in children. Herein, two children cases are presented with respiratory distress, who were diagnosed as Chilaiditi syndrome by chest radiography.

KEYWORDS: Chilaiditi syndrome, child, respiratory distress**Received:** 21.11.2013**Accepted:** 23.01.2014**Available Online Date:** 11.07.2014

INTRODUCTION

Chilaiditi syndrome is defined as hepatodiaphragmatic interposition of colon or small intestine [1]. Whilst the term “Chilaiditi sign” is recommended to be used to define asymptomatic cases, the term “Chilaiditi syndrome” is recommended to be used for symptomatic cases [1-3]. Its incidence increases with age. It is more prevalent in males, the majority of which are elderly and mentally retarded patients [3]. Male/female ratio is 4/1 [1]. Whilst it may be clinically asymptomatic, it also presents itself with gastrointestinal system symptoms such as abdominal pain, vomiting, constipation, loss of appetite, and abdominal distension and/or respiratory system symptoms such as respiratory distress and chest pain, and less frequently with cardiovascular system symptoms [1-6]. Herein, two cases with Chilaiditi syndrome presented with respiratory distress were reported.

CASE PRESENTATIONS

Written informed consent of the patients was obtained from the parents of the children.

Case 1

A 10-year-old boy presented with wheezing and shortness of breath. His medical history revealed that he had frequently admitted to the health care centers for respiratory distress since 8 years of age and received nebulized salbutamol treatment due to recurrent lower respiratory tract infection. His recent complaints appeared 3-4 days ago. On his physical examination, body weight was in the 10th-25th percentile and height was in the 10th percentile. Respiratory system examination revealed bilateral rhonchi with bilaterally equal breath sounds. There were no pathological findings on the examination of other systems. Laboratory examination revealed normal results for complete blood count and blood biochemistry. On chest X-ray, presence of gas below the right diaphragm attracted attention (Figure 1). On the abdominal ultrasonography of the patient, colonic loop was observed in the right anterior lobe of the liver. Magnetic resonance imaging (MRI) of the abdomen was performed for the differential diagnosis between Chilaiditi syndrome and diaphragmatic hernia and the diagnosis of Chilaiditi syndrome was confirmed (Figure 2a, b). His anamnesis revealed no gastrointestinal system complaint. Reversibility was not detected on respiratory function test, which was performed because of history of asthma together with Chilaiditi syndrome. The case, respiratory distress of whom regressed with inhaled steroid, oxygen, and laxative treatments, was discharged. The patient has been following-up for respiratory system and potential gastrointestinal system complications.

Case 2

A 7-year-old boy, who was followed-up for chronic pulmonary disease secondary to gastroesophageal reflux, presented with coughing, wheezing, and shortness of breath. His anamnesis revealed that he had undergone surgery for tracheoesophageal fistula, was on follow-up for achondroplasia in our Endocrinology Clinic, and had had recurrent hospitalization for pulmonary infections. The patient had no additional gastrointestinal symptom except for abdominal distension existed for the last one year. On his physical examination, body weight and height were <3rd percentile. Respiratory system examination revealed crepitating rales and sibilant rhonchi in both lungs. Pathological findings in the other systems included brachycephaly, short extremities, short and wide hand and foot fingers, and abdominal distension. On

This case had been presented as a poster at 20th National Allergy and Clinical Immunology Congress in Antalya between 2 and 6 November 2013.

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Figure 1. Presence of gas below the right diaphragm on the chest X-ray of Case 1

laboratory examination of the patient, complete blood count revealed leukocytosis and a left shift; however, the results of routine biochemistry analysis were unremarkable. Erythrocyte sedimentation rate and C-reactive protein were within normal ranges. On his chest X-ray, bilateral widespread pneumonic infiltration areas, atelectatic changes, and presence of gas below the right diaphragm attracted attention (Figure 3). Abdominal MRI was performed for differential diagnosis and the diagnosis of Chilaiditi syndrome was confirmed (Figure 4). The patient was followed-up under antibiotic and antireflux therapy. Fiber diet was recommended for abdominal distention. The patient, symptoms of whom were regressed, was discharged and follow-up visits were planned for probable complications.

DISCUSSION

Chilaiditi syndrome was first defined in 1910 by Demetrius Chilaiditi in three cases without complaint. Its frequency changes between 0.25% and 0.28% and increases with age [1-3]. Most of the cases are asymptomatic and detected incidentally, whereas some may have respiratory system symptoms such as respiratory distress and chest pain, some have gastrointestinal system symptoms such as vomiting, abdominal pain, constipation, abdominal distension, and loss of appetite, and some have cardiovascular system symptoms such as tachycardia, arrhythmia, and angina-like chest pain [1-7]. As a matter of fact, whilst there were only respiratory system symptoms in the first case, both gastrointestinal and respiratory system symptoms were observed in the second case. There might be rare cases with multiorgan symptoms [1-7].

Gastrointestinal symptoms may be mild, recurrent, chronic, and severe. Our second case was suffering from abdominal distension for the last one year. It may sometimes cause clinical pictures such as volvulus (in cecum, splenic flexure, transverse colon), incarceration, and perforation, which might require emergent intervention [1,4-7]. Pseudo-obstruction may be encountered in colon (Ogilvie's Syndrome). In addition, coexistence with pulmonary and

Figure 2. a, b. Abdominal MRI of Case 1; hepatic flexure is seen interposed between diaphragm and liver in the anterior aspect

MRI: magnetic resonance imaging

gastrointestinal malignancies (colon, rectum, gastric) has been reported [1,7]. In the literature, Chilaiditi sign was reported in a 15-year-old girl diagnosed with multiple endocrine neoplasia type 2B [7]. Chilaiditi syndrome was reported in an 18-year-old case presented with direct hyperbilirubinemia and elevated transaminases, in whom jaundice gradually decreased with supportive treatment [5].

Chilaiditi syndrome may either be congenital or acquired. Hepatic factors such as liver ptosis, small liver (due to cirrhosis, hepatectomy, etc.) and relaxation of suspensor ligament; diaphragmatic factors such as weakening and degeneration in diaphragmatic muscles, and phrenic nerve paralysis and those lead to changes in intrathoracic pressure due to tuberculosis or emphysema; and megacolon, abnormal

Figure 3. Bilateral widespread pneumonic infiltration areas, atelectatic changes, and presence of gas below the right diaphragm on the chest X-ray of Case 2

Figure 4. Abdominal MRI of Case 2; hepatocolonic interposition and dilatation in the intestinal segments

MRI: magnetic resonance imaging

mobilization of colon due to congenital reasons, intestinal hypermotility with loose mesentery, insufficient fixation of cecum, increased intraabdominal pressure gradient (pregnancy, obesity, ascites), trauma and related adhesions, and distension due to excessive aerophagia may be responsible for the development of this syndrome [1-3,6]. Huang et al. [8] reported three cases between 1990 and 2005 and additionally evaluated 10 pediatric cases reported in the literature

between the same years. They mentioned the predisposing factors as aerophagia by 46.2%, diaphragmatic eventration by 23.1%, constipation by 23.1%, and abdominal trauma by 7.7%. Whilst medical and family history of our first case revealed no predisposing factor that might lead to such a condition, achondroplasia and tracheoesophageal fistula surgery in our second case appeared as predisposing factors.

Diagnosis is usually established incidentally by chest X-ray or standing direct abdominal radiography. Subdiaphragmatic abscess, retroperitoneal masses, and liver abscess should be considered in the differential diagnosis [1,2]. These pathologies that were considered in the differential diagnosis of our cases were eliminated via MRI. In addition to these, volvulus, invagination, intestinal obstruction, and ischemic and inflammatory bowel diseases (appendicitis, diverticulitis, etc.) should also be considered in the differential diagnosis. It may sometimes be misdiagnosed as diaphragm hernia; in the literature, an intestinal interposition that accompanied Bochdalek hernia has been reported [1].

Treatment is not necessary in cases with Chilaiditi sign. Clinician should eliminate emergent conditions in symptomatic cases. Detection of Chilaiditi sign in cirrhosis patients is important to prevent complications that are likely to occur during liver biopsy since proclivity is higher in such patients. Performing colonoscopy is also difficult in patients with Chilaiditi sign and risk of colon perforation during procedure is increased [1,2]. Initial treatment includes bed rest, parenteral fluid therapy, decompression, and laxatives [1-3,7]. Intestines may sometime return back to the normal position after decompression [1]. Surgery is the treatment of choice in case obstruction symptoms are not relieved and there are signs of intestinal ischemia [1-5]. Surgical treatment is recommended also in the presence of recurrent symptoms [8]. In such cases, surgical treatment includes subtotal colectomy, fixation of colon to peritoneum at umbilical level, or hepatoxy to narrow subphrenic area. Colonoscopic reduction is not recommended in the presence of transverse colon volvulus because gangrene may be encountered by 16% [1,2]. Huang et al. [8] reported 13 cases aged 6 months-11 years between 1990 and 2005, of which 23.1% had respiratory symptoms and the majority improved with conservative treatment. The youngest case reported in the literature was a one-day-old newborn presented with respiratory distress and abdominal distension; the symptoms regressed also with conservative treatment [9]. Keles et al. [10] reported a 5-month-old baby presented with respiratory distress, the symptoms of whom relieved with conservative treatment. In the present cases, laxative therapy was adequate without need for decompression.

Chilaiditi syndrome is usually detected incidentally but may sometimes be symptomatic. Patients may present with gastrointestinal, respiratory or cardiovascular system symptoms and sometimes with multiorgan symptoms. The diagnoses of the present two cases, which could be easily overlooked due to accompanying respiratory tract diseases, suggested the need to explore extrapulmonary regions when evaluating chest X-ray.

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