Chilaiditi syndrome: An unusual post-operative complication of congenital diaphragmatic hernia

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ABSTRACT

Chilaiditi syndrome is a condition in which the hepatodiaphragmatic colonic interposition is accompanied by signs and symptoms such as pain abdomen, nausea, vomiting, and respiratory distress. Chilaiditi sign is defined as the asymptomatic radiological diagnosis of segmental interposition of a portion of large intestine or hollow viscera between the undersurface of liver and hemidiaphragm. The syndrome is managed conservatively. Recognition of this syndrome and its differentiation from pneumoperitoneum is important in patients with respiratory or gastrointestinal symptoms with a characteristic radiographic picture of this syndrome to prevent unnecessary surgical intervention. Right-sided congenital diaphragmatic hernias (CDHs) are rare in children and may mimic Chilaiditi syndrome. Here, we report the case of an 18-month-old girl, who was later on diagnosed as a case of Chilaiditi syndrome secondary to a post-operative repair of the right-sided CDH and was managed conservatively with the help of intravenous fluids and bronchodilators.

Keywords: *Chilaiditi syndrome, Congenital diaphragmatic hernia, Pneumoperitoneum*

Chilaiditi syndrome is a condition in which the colonic interposition is accompanied by signs and symptoms such as pain abdomen, nausea, vomiting, respiratory distress, and Chilaiditi sign. Chilaiditi sign is defined as the asymptomatic radiological diagnosis of the segmental interposition of a portion of large intestine, liver, or hollow viscera between the undersurface of liver and hemidiaphragm, which was first reported in 1910 by a Greek radiologist Demetrius Chilaiditi [1]. The sign has an overall incidence of 0.025–0.28% in plain radiographs and 1.1–2.4% in computed tomography (CT) scans of the chest and abdomen [1]. It is more commonly seen in the elderly wherein the incidence is about 1% but can also be seen in children below 5 months of age, with a male-female ratio of 4:1 through all age ranges [2]. We report the case of an 18-month-old girl, who was later on diagnosed as a case of Chilaiditi syndrome secondary to a post-operative repair of the right-sided congenital diaphragmatic hernia (CDH).

CASE REPORT

An 18-month-old girl presented to the emergency department with a 3-day history of fever, cough, and respiratory distress. She had no history of associated vomiting, abdominal pain, distention, or constipation. Her past and family histories were negative for any recurrent respiratory or gastrointestinal complaints. Her birth history revealed that she was born preterm (34 weeks) and had low birth weight and respiratory distress at birth. On day 1 of life, her X-ray chest revealed segmental opacity in the right middle lobe, C-reactive protein was positive with Klebsiella sepsis, and eventually, she was diagnosed as a case of CDH because of prolonged ventilation and relapse of symptoms on weaning from the ventilator. Right-sided repair of CDH was done at 5 months of age.

When the patient reported to our department, her clinical examination revealed a respiratory rate of 68/min, with subcostal, intercostal retractions, and suprasternal recessions. Rest of the vitals was normal, and Sp02 was 67% without oxygen. Rhonchi and crepitation were present all over the chest. There was no cyanosis, heart rate was 130/min, peripheral pulse was well felt, and abdominal, cardiac system, and central nervous system examination were unremarkable.

She was immediately started on oxygen, nebulized with bronchodilators (salbutamol and ipratropium bromide) and given intravenous fluids. Her condition deteriorated over the next 24 h with persistence of severe respiratory distress. Basic laboratory studies revealed: Hemoglobin -7.6g%, total leukocyte count - 15400, polymorphs - 53%, lymphocyte - 44%, eosinophils - 3%, platelets-4.4lakhs,meancorpuscularvolume-55,meancorpuscular hemoglobin concentration - 33, RBC count - 2.5 million/cu mm, sodium - 135 m eq/l, potassium - 3.8 m eq/I, blood urea - 36 mg%. and creatinine - 0.4%.

X-ray chest was suggestive of elevated right hemidiaphragm (Fig. 1) and the presence of bowel loops in between the undersurface of a diaphragm and the right lobe of the liver. Abdominal ultrasonography (USG) was subsequently performed which confirmed the interposition of bowel loops between an undersurface of the right hemidiaphragm.

As her respiratory symptoms continued over the next 4 days and there was no improvement, a possibility of Chilaiditi syndrome was kept, and a CT of the abdomen and chest was planned to consider for the need of decompression or surgical correction. CT scan was suggestive of an elevated right dome of the diaphragm, with colonic interposition between the right hemidiaphragm and the right lobe of the liver with partial atelectasis of the right lower lobe of the lung (Fig. 2).

The child was started on conservative management with rest, fasting, intravenous fluids, nasogastric decompression (to take care of aerophagia), and bronchodilator therapy (salbutamol and ipratropium bromide) every 4 h. Over the next 24 h, the child showed dramatic improvement to the above measures with the subsidence of all the respiratory symptoms in the form of decreased respiratory rate to 38/min and no distress. The marked improvement in the respiratory distress and general condition of the patient after decompression of aerophagia pointed toward a possibility of an acquired Chilaiditi syndrome as sequelae to post-diaphragmatic hernia repair.

The child was found to be stable under follow-up, and a repeat X-ray chest along with magnetic resonance imaging (MRI) was done after 2 weeks which was again suggestive of the persistence of hepatodiaphragmatic colonic interposition, possibly due to fibrosis. A diagnosis of Chilaiditi syndrome secondary to repair of the right-sided CDH was being made, and the patient showed considerable improvement on subsequent follow-up.

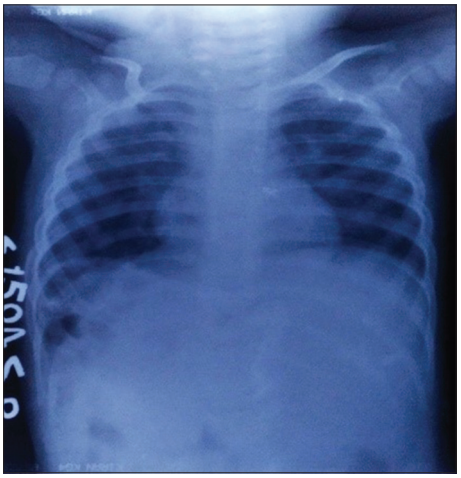

Figure 1: X-ray chest suggestive of elevated right hemidiaphragm and presence of bowel loops in between undersurface of the diaphragm and the right lobe of the liver

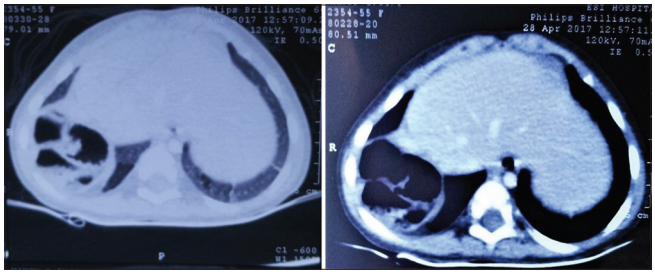

Figure 2: Computed tomography scan suggestive of elevated right dome of the diaphragm with colonic interposition between right hemidiaphragm and the right lobe of the liver with partial atelectasis of the right lower lobe

DISCUSSION

Hepatodiaphragmatic interposition of a colon is rare in children which usually presents as an asymptomatic radiological sign known as Chilaiditi sign or with gastrointestinal or respiratory symptoms called as Chiladiti syndrome [1]. The most common symptoms in patients with Chilaiditi syndrome are gastrointestinal (abdominal pain, nausea, vomiting, and constipation), followed by respiratory (respiratory distress) and rarely multiorgan symptoms.

The interposed segment is hepatic flexure of transverse colon in most of the patients. Normally, the anatomic positions of the diaphragm, colon, and its suspensory ligaments prevent the interposition of the colon. Any anatomical variation, congenital or acquired in the above structures in the form of laxity of suspensory ligaments, gaseous distension of colon, or the conditions causing increased intra-abdominal pressure may contribute to colonic interposition.

The presentation of Chilaiditi syndrome may vary from mild abdominal pain, nausea, vomiting to signs of acute abdomen suggestive of obstruction, perforation, or colonic pseudo-obstruction. Chilaiditi sign mimics "free air" under the diaphragm. The following criteria must be met for the radiological diagnosis of Chilaiditi sign: The right hemidiaphragm must be adequately above the liver by the intestine, the bowel must be distended by air, and superior margin of the liver must be depressed below the level of the left hemidiaphragm [3]. In addition, there should be a presence of haustral folds and the location of the air should not be changed by altering the posture of the patient, unlike in a patient with free air [4,5]. The predisposing factors for Chilaiditi syndrome are elongated redundant mobile colon, absence or laxity of a ligament, suspending transverse colon, aerophagia, chronic constipation, blunt trauma to the abdomen, diaphragmatic eventration, diaphragmatic paralysis, cirrhosis, chronic lung diseases, obesity, and ascites [6].

Chilaiditi sign must be differentiated from pneumoperitoneum by X-ray. Pneumoperitoneum normally shows a crescent-shaped gas shadow under the diaphragm without haustral folds and altering the posture of the patient changes the position of the gas. The common differentials are the subphrenic abscess, pneumoperitoneum, retroperitoneal masses, and posterior liver lesions. It can be confused with CDH, congenital cystic adenomatoid malformation of the lung, internal hernias, bowel obstruction, volvulus, intussusception, ischemic bowel, or inflammatory conditions, such as appendicitis and diverticulitis.

In a study, Huang *et al*. reviewed 13 children with Chilaiditi syndrome in the past 15 years, 6 males and 7 females aged 6–11 years old. He found predisposing factors as aerophagia in 46.2%, diaphragmatic eventration in 23.1%, constipation in 23.1%, and blunt trauma in 7.7%. The most common manifestation was an abdominal pain in 69.2% and respiratory distress in 23.1% of patients. Only four patients with recurrent symptoms had a surgical correction [7].

Chilaiditi sign does not require intervention. Recognizing this sign is important because it can be mistaken for pneumoperitoneum leading to unnecessary surgical intervention [3]. The management

of Chilaiditi syndrome is usually conservative in the form of bed rest, fasting, intravenous fluids, bowel decompression, and enemas. In case of persistent pain or signs of obstruction or acute abdomen, surgical correction may be indicated. In our case, Chilaiditi syndrome was confirmed by USG, CT scan, and later by the MRI, and the patient was managed conservatively with the help of intravenous fluids and bronchodilators.

This case is rare because this was a female child who presented at an earlier age than most reported Chilaiditi syndrome in children and was secondary to a post-operative repair of the right-sided CDH, which itself is a differential of Chilaiditi syndrome.

CONCLUSION

Right-sided CDHs are rare in children and so are Chilaiditi syndrome. A CDH may mimic Chilaiditi syndrome making it difficult for a clinician to differentiate. Chilaiditi syndrome must also be differentiated from other close mimics like pneumoperitoneum to avoid unnecessary surgical intervention. Our case was extremely unusual as it combined both the rare features and was an acquired Chilaiditi syndrome secondary to a post-operative repair of the CDH.

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