

A case of liver cirrhosis and Chilaiditi syndrome with atypical pneumonitis

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ABSTRACT

Respiratory distress is very uncommon as a presenting symptom of Chilaiditi syndrome. Furthermore, pneumonia is not documented with the syndrome, compromising further to the distress. We describe a middle-aged man, chronic alcoholic, recently diagnosed with liver cirrhosis, presented with a 1-year history of slowly progressive breathlessness. Recently, he developed mild-to-moderate hemoptysis and cough with aggravation of breathlessness. He did not have fever, chest pain, or orthopnea. He was cyanosed, requiring high-dose oxygen therapy. Later on, he stabilized with noninvasive ventilation. Chest imagings showed incidental Chilaiditi sign, liver cirrhosis, and atypical pneumonitis. With empirical antibiotics and high-dose steroid, he recovered completely but with baseline breathlessness. Here, we outline Chilaiditi syndrome as a rare association or manifestation of liver cirrhosis, and it can present with a respiratory compromise by both obstructive lung disease and atypical interstitial pneumonia. Early identification, vaccinations against common organisms, and possible early surgery may prevent morbidity and mortality of this type of patients.

Keywords: Chilaiditi sign, chronic liver disease, obstructive lung disease, pneumonia

Introduction

Chilaiditi syndrome is the presence of radiographic Chilaiditi sign (hepatodiaphragmatic interposition of hollow viscera) in a symptomatic patient. The sign is often an incidental finding, more prevalent than the syndrome, more commonly seen in elderly male, and mostly acquired rather than congenital one. The associated acquired conditions are liver cirrhosis (22%), colonic volvulus and malignancies, chronic obstructive pulmonary disease (COPD), and pulmonary malignancies.^[1,2] The syndrome presents often with gastrointestinal symptoms (abdominal pain, nausea, vomiting, and constipation) and rarely with respiratory distress and angina-like chest pain.^[3] Presentation with atypical pneumonitis has not been documented.

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Hereby, we report a rare case of Chilaiditi syndrome and alcoholic liver cirrhosis who presents with severe respiratory distress after atypical pneumonitis.

Case Report

A 55-year-old man from Andaman and Nicobar Islands, laborer by occupation, alcoholic but nonsmoker, presented with a 1-year history of breathlessness, which was gradually progressive; had increased to mMRC Grade-IV for the last 10 days; and was not associated with leg swelling, orthopnea, or paroxysmal nocturnal dyspnea. Fifteen days back, he developed one episode of hemoptysis of 10–20 ml fresh blood and in the last 5 days, it increased up to 4–5 episodes per day with the same amount. Hemoptysis was begun with dry irritating cough, but it had no postural or diurnal variation. He did not have throat discomfort, chest pain, fever, or any blood vomitus, although he had

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melena for 2 days. He was recently diagnosed as chronic liver disease (cirrhosis), mostly due to chronic alcohol intake. Upper gastrointestinal endoscopy was performed 3 months back, which was normal. In addition, he did not have portal hypertension, splenomegaly, or ascites. Other comorbidities such as diabetes, hypertension, asthma/COPD, or tuberculosis were not present.

The patient was conscious, afebrile, but dyspneic (respiratory rate [RR]-24/min), and cyanosed (SpO₂ was 64% on oxygen, 6 L/min with nasal prong). His pulse rate, blood pressure, and jugular venous pulse were within normal limits. Chest examination demonstrated reduced air entry in right infrascapular and infraaxillary region with bilateral fine-coarse crackles (right > left, upper half > lower half). There were amphoric bowel sounds heard over the right lower hemithorax. Cardiac apex was shifted 2 cm laterally from left midclavicular line on the fifth intercostal space. Liver dullness measured 4 cm along the right midclavicular line. Other systemic examinations were unremarkable.

The patient was stabilized with oxygen (6 L/min through nasal prong then 10 L/min through facemask), failing which noninvasive ventilation (BiPAP: IPAP, 12 cmH₂O; EPAP, 8 cmH₂O) was started and monitored with arterial blood gas parameters [Table 1].

Routine investigations showed normal hemogram, liver and kidney function tests, and blood sugar. Total eosinophil count was normal. Urine examinations did not reveal any proteinuria, dysmorphic red blood cell, or casts. Chest X-ray showed grossly elevated right diaphragm with underneath bowel gas suggesting Chilaiditi sign [Figure 1a]. This sign was confirmed with both ultrasonography and computed tomography (CT) of chest and abdomen. Ultrasound revealed shrunken and coarse echotexture liver, suggesting a chronic liver disease (or cirrhosis). Ascites or other signs of portal hypertension were absent. CT revealed interposed colon between right diaphragm and liver, atypical pneumonitis, and cirrhotic liver [Figure 1b-d]. There was no induced sputum production and bronchoalveolar lavage was not performed by the hospital. Hence, etiological diagnosis could not be ascertained much. Viral markers including HIV, hepatitis B, and hepatitis C were negative. Leptospirosis, an endemic disease here and can present isolatedly with pneumonitis, was suspected, but blood serology tests (IgM) came negative. Scrub typhus and *Pneumocystis jirovecii* pneumonia could not be ruled out. Hence, empirical antibiotics (injection ceftriaxone, tablet co-trimoxazole, and doxycycline) covering all these infections were started. Steroid (injection hydrocortisone 100 mg intravenous 8 hourly) was also started suspecting severe *Pneumocystis* pneumonitis (PaO₂ <70 mmHg) requiring BiPAP ventilation and oxygen therapy.

Final diagnosis of alcoholic liver cirrhosis and Chilaiditi syndrome with atypical pneumonitis, possibly *Pneumocystis* infection, was made. With the above treatments, the patient's condition improved gradually, BiPAP and oxygen requirements nullified, and he was discharged on the 10th day of the hospitalization

Table 1: Arterial blood gas analysis reports with time

Parameters	Day 1 (morning)	Day 1 (evening)	Day 3
pH	7.367	7.354	7.451
pCO ₂ (mmHg)	53.4	71.3	40.7
pO ₂ (mmHg)	22.0	58.4	68.4
cHCO ₃ ⁻ (mmol/L)	30.7	39.7	28.4
cSpO ₂ (%)	34.3	87.1	89.1
Lactate (mmol/L)	1.43	0.36	0.42

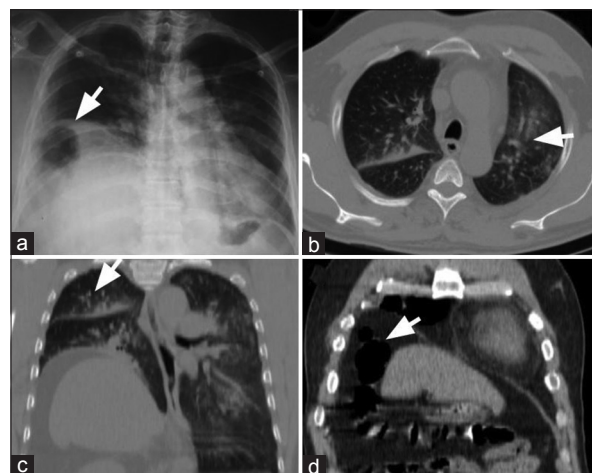


Figure 1: Chest X-ray (a: anteroposterior view) showing gas under right diaphragm (white arrow) suggesting Chilaiditi sign. Noncontrast computed tomography chest axial (b) and coronal sections (c) showing diffuse, bilateral, and perihilar distributed ground glass opacities (white arrow) with interstitial infiltrates suggesting atypical pneumonia. Fissure edema and bronchial wall thickening (white arrow) noted. Coronal section (d) confirming Chilaiditi sign having colon interposed between right diaphragm and shrunken liver (white arrow) with an elevated dome of hemidiaphragm

after receiving the first dose of vaccinations (against influenza, pneumococcal, and *Haemophilus influenzae*). However, his basal RR was in the higher range (RR, 18–22/min), possibly due to mechanical obstruction by elevated diaphragm that was causing respiratory distress to him for the past 1 year.

Discussion

Chilaiditi sign, coined after Demetrius Chilaiditi in 1910, is an incidental finding in chest and abdominal plain X-rays (0.02%–0.28%) and CT-scan (1.1%–2.4%).^[4,5] This sign is diagnosed radiologically based on criteria: (a) the right hemidiaphragm must be adequately displaced superiorly above the liver by the intestine, (b) pseudoperitoneum caused by air in the bowels must be seen, and (c) the superior margin of the liver must be positioned below the level of the left hemidiaphragm. Although many theories have been told regarding the origin of this sign, the basic pathophysiology is recognized to be the change in relative anatomical relations among liver, diaphragm, and colon. Hence, etiological or predisposing factors can be classified as hepatic, diaphragmatic, intestinal, or miscellaneous [Table 2].^[6] Our case was having liver cirrhosis due to chronic alcohol intake which may have contributed to the development of this sign.

Table 2: Etiological factors for Chilaiditi sign

Hepatic causes	Reduced liver volume due to Cirrhosis Right lobe segmental agenesis Liver ptosis due to relaxation/laxity of the suspensory ligaments
Diaphragmatic/pneumonic causes	Abnormally high diaphragm due to Muscular degeneration Phrenic nerve injury Excessive aerophagia Chronic obstructive pulmonary disease Changes in intrathoracic pressure (emphysema) Pulmonary malignancies
Intestinal causes	Abnormal or increased colonic mobility Elongated or redundant colon with long mesentery Absence of peritoneal attachments Malrotation or congenital malpositioning of the bowel Gastrointestinal malignancies
Miscellaneous causes	Enlarged lower thoracic outlet Elevated intra-abdominal pressure Ascites Pregnancy High abdominal fat content/obesity Mental retardation

Hence, all liver cirrhosis should be regularly followed up for early recognition of this sign and further development of the corresponding Chilaiditi syndrome. However, co-occurrence of both diseases cannot be ruled out.

The above sign leading to clinical symptoms, i.e., Chilaiditi syndrome, is even more rare. When present, these symptoms localize to the abdomen and/or chest. More often, the syndrome is a mimicker of acute abdomen or even acute chest condition.^[7] Pulmonary manifestation of the syndrome is rarely described, but physiologically, it is there due to mechanical reduced lung capacity (obstructive lung disease). This factor causes respiratory distress even there is no chest pathology.^[8] The reason our case was suffering from breathlessness for 1 year may be due to this factor.

There are case reports of the syndrome with associated chronic lung conditions such as interstitial lung disease (ILD), COPD, and pulmonary malignancy.^[9,10] These conditions are probably causing enlarged lower thoracic outlet after pushing the liver forward and downward, creating adequate space between the diaphragm and liver that facilitates the development of Chilaiditi sign. Once this sign develops, prevalent respiratory distress increases further due to reduced lung capacity. In another way, isolated case of this sign as a component of obstructive lung disease may increase the frequency of chest infections and associated morbidity similar to COPD and ILD causing the same.^[11] Moreover, the severity of chest infections/pneumonia may also have been increased by the syndrome due to background respiratory distress similar to the present case. Our case is the first documented pneumonia

associated with this syndrome, although cirrhosis itself can increase the risk for the same.

This syndrome is managed conservatively; however, it rarely needs surgery especially in intestinal obstruction. Sometimes, unwarranted surgery is done considering herniated bowel gas as perforation.^[12] Hence, increased awareness of the sign/syndrome and higher degrees of suspicion are required to avoid such unfortunates. Although respiratory compromise is not being reported as indication for surgical intervention, it seems more physiological to halt the progression of obstructive lung disease or the precipitation of severe pneumonia. A recent case study of the success of mini-invasive laparoscopic surgery in this syndrome may also help to decide for early surgery.^[13] Our case is planned for early laparoscopic surgery. Furthermore, to prevent recurrent pneumonia against influenza, pneumococcal, or *H. influenzae*, vaccinations should be considered similar to other obstructive/restrictive lung diseases.

Conclusion

Chilaiditi syndrome is a rare association or manifestation of liver cirrhosis and can present as severe respiratory compromise due to obstructive lung disease and superadded pneumonitis. Early identification including the etiological factors, possible vaccinations against above organisms, and early surgery may prevent morbidity and mortality of the patient.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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