Case report

Management of Dual Diagnosis: Chilaiditi Syndrome and Biliary Atresia Complicated by Cirrhosis

Abstract

Chilaiditi sign is an incidental radiographic finding of a usually asymptomatic condition, where part of the intestine is situated between the liver and the diaphragm. However, the term "Chilaiditi syndrome" is used for symptomatic hepatodiaphragmatic interposition. We report the case of a 15-year-old patient presenting with chronic hepatic colic due to this syndrome, which was diagnosed by abdominal CT scan. Most patients with Chilaiditi syndrome can be managed conservatively. However, surgery is indicated for those who do not respond to conservative management or in cases of suspected serious complications such as ischemia or intestinal perforation. Biliary Atresia (BA) is a rare congenital malformation characterized by an inflammatory and destructive process that obstructs the intra- and extrahepatic bile ducts, leading to rapid progression to complete cholestasis and irreversible biliary cirrhosis, ultimately resulting in the child's death within the first few years of life. Our case is the rare in the literature indicating the association of Chilaiditi syndrome and biliary atresia.

Keywords : Chilaiditi syndrome - Biliary atresia - 15-year-old girl – treatment

Introduction

Chilaiditi syndrome is a rare condition defined by the presence of gastrointestinal symptoms associated with the radiological finding of segmental interposition of the intestine between the liver and diaphragm. Although rarely identified as a source of abdominal pain, Chilaiditi syndrome has clinical significance as it can lead to a number of serious complications including bowel obstruction, perforation and ischemia [8-11].Most patients with this intestinal anomaly are asymptomatic throughout their lives; however, they can manifest with intermittent abdominal pain, distention, vomiting, anorexia, and constipation that on rare occasions require surgical intervention [12].

Case report

A 15-year-old girl has a post medical history of neonatal jaundice revealing a type I biliary atresia complicated with liver cirrhosis. She underwent a Kasai procedure at the age of 1 month with smooth post-operative course and unremarkable follow-up. The patient presented with chronic hepatic colic without any other associated signs including no fever, jaundice, vomiting or constipation. Abdominal examination revealed slight tenderness in the right hypochondrium, and both biological and Abdominal US results were unremarkable.Symptomatic treatment was initiated, but due to persistent symptoms, an abdominal CT scan was performed, revealing colonic interposition between the liver and the right hemidiaphragm (Figure1).The diagnosis of Chilaiditi syndrome was established. Following a multidisciplinary medical team discussion and considering the patient's complex medical history, analgesic treatment was recommended.

Discussion :

The Chilaiditi sign was initially identified by Antoine Béclère in 1899. However, in 1910, Demetrious Chilaiditi, a Greek radiologist first described three cases of right hemi-diaphragmatic interposition of the colon [1]. In the general population,

the prevalence of the Chilaiditi sign is estimated to be between 0.025% and 0.28%, but it could reach 1% in the elderly. The male-to-female sex ratio is 4:1[2].

The Biliary Atresia (BA) is a rare congenital malformation characterized by an inflammatory, destructive process that obstructs the intra- and extrahepatic bile ducts, leading to rapid progression to complete cholestasis and irreversible biliary cirrhosis, ultimately resulting in the death of the child within the first few years of life [3,4].

The Chilaiditi sign is generally asymptomatic and is often discovered incidentally. However, if the patient is symptomatic, it is referred to as Chilaiditi syndrome, which may manifest as abdominal pain, constipation, nausea, vomiting, and occasionally dyspnea, respiratory distress and cardiac arrhythmias [2,5]. It can lead to complications such as obstruction, volvulus, or digestive perforation. Diagnosis is confirmed through a chest X-ray and abdominal CT scan. To diagnose Chilaiditi's sign from radiographic images, following criteria must be met in erect position: An elevation of the right hemidiaphragm compared to the liver, distension of the colon due to the presence of air, and the upper margin of the liver situated below the level of the left hemidiaphragm [2]. The Various contributing factors have been identified, including cirrhosis, absence or laxity of suspensory ligaments or the falciform ligament, aerophagia, increased abdominal pressure (due to conditions such as ascites, obesity, or pregnancy), elevation of the right hemidiaphragm (due to causes like paralysis or herniation), dolichocolon, chronic constipation, prolonged bed rest, intestinal malrotation, chronic bronchitis, and pulmonary emphysema. Other factors such as mental retardation or schizophrenia have also been identified [6,7].

In most cases, conservative medical treatment with analgesics or the use of a nasogastric is sufficient. However, severe complications may require surgical intervention.The primary treatment of biliary atresia is a hepatoportoenterostomy (Kasai procedure), which creates a bile-digestive shunt between the liver hilum and the jejunum. For our patient, after a multidisciplinary discussion, she received long-term conservative treatment due to her severe condition, which contraindicates surgical treatment, along with regular monitoring for her cirrhosis.

To our knowledge this is the first report in a paediatric patient of an association of biliary atresia associated with chilaiditi syndrome.

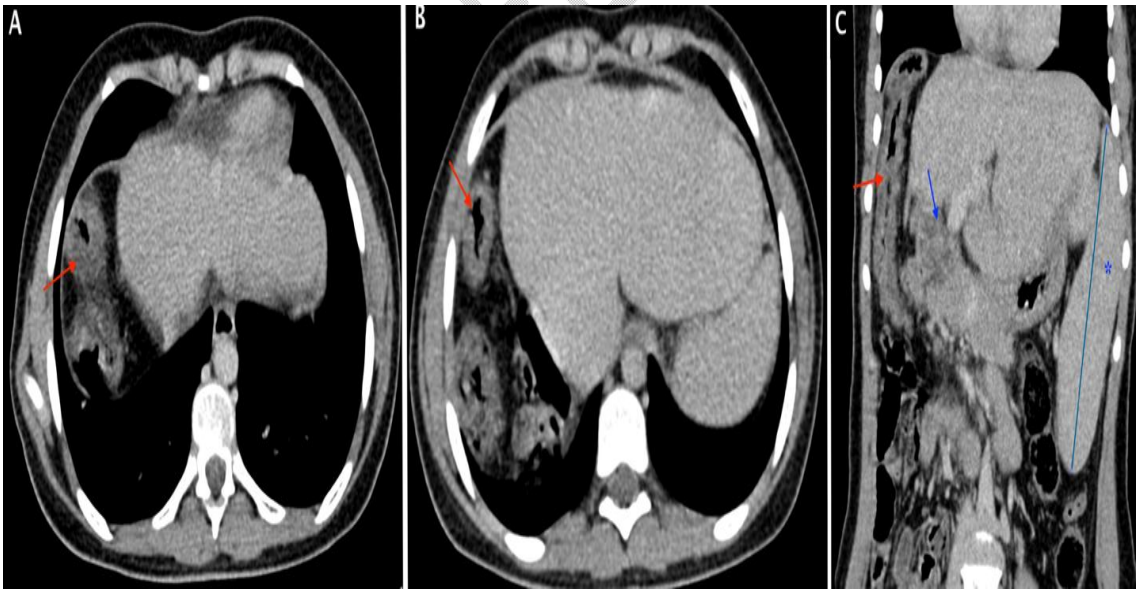

Figure 1:(A and B)Axial sectionwith (C)sagittal reconstructionof abdomen CR SCAN demonstrating the presence of interposed colonic loops between the right hemidiaphragm and liver (red arrows). Note the splenomegaly (blue asterisk) and stigmata of hepato-portoenterostomy, also known as Kasai procedure (blue arrow).

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