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Scleroderma Associated with Chilaiditi Syndrome: Case Report

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Abstract **Case Report**

Chilaïditi syndrome corresponds to the interposition, most often asymptomatic and temporary, of the intestine between the upper surface of the liver and the right diaphragmatic dome. It is characterized by the association of radiological semiology and various digestive manifestations, acute or chronic abdominal pain, intermittent sub-occlusions, nausea, vomiting, transit disorders, and rare intestinal occlusive complications. We report through this observation a case of a patient whose chilaiditi syndrome was the mode of revelation of her scleroderma.

Keywords: Chilaiditi syndrome, scleroderma.

INTRODUCTION

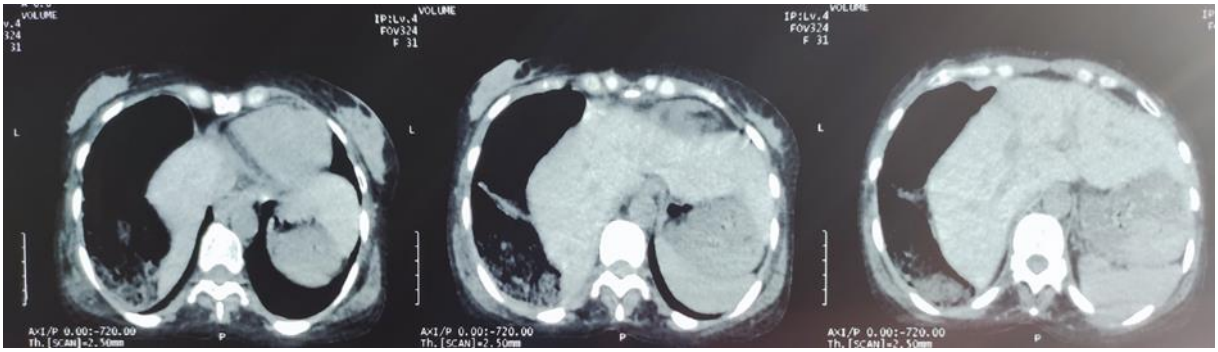
The chilaiditi syndrome is a rare pathology characterized by the interposition of the colon or small intestine in the interhepato-diaphgrammatic space [1]. It is defined as an association of radiological semiology and various digestive manifestations, acute or chronic abdominal pain, intermittent sub-occlusions, nausea, vomiting, transit disorders, and rare intestinal occlusive complications [2]. We report through this observation a case of a patient whose chilaiditi syndrome was the mode of revelation of her scleroderma.

MEDICAL OBSERVATION

We report the case of a 40-year-old patient who had been complaining for 5 months of episodes of constipation and nausea for which she was put on laxative treatment and an antispasmodic without any improvement. Two days before her hospitalization, the patient presented an episode of post-prandial vomiting, and diffuse abdominal pain, associated with constipation without cessation of matter or gas or dysphagia. Faced with this clinical picture, and after having ruled out pancreatitis, an abdominal CT scan is requested showing an interposition of the digestive loops in the interhepato-diaphragmatic area in favor of chilaiditi syndrome without signs of perforation (Figure 1).

Resuming the interrogation; the patient also reported as functional digestive signs only the manifestations already described: gastro-oesophageal reflux, and dyspepsia and as systemic signs: Renauld's phenomenon, inflammatory-like polyarthralgia of the medium and small joints, dry occulobuccal, without dyspnoea or myalgia, no paresthesias or photosensitivity. The clinical examination found disappearance of forehead wrinkles, limitation of mouth opening through 2 fingers, sclerodactyly with a pulp scar at the level of the 2nd finger and cutaneous calcinosis next to the 3rd finger. Faced with these multi-systemic attacks, a biological assessment is requested showing a biological inflammatory syndrome (the sedimentation rate was accelerated to 68 mm, and the CRP increased to 49.5mg/l). The immunological assessment revealed an anti-nuclear antibody rate of 1/1280 with a nuclear appearance, with the positivity of the following autoantibodies: SSA/RO, SSB/LA, SCL70, and centromere. Renal function and troponin levels were normal. On the radiological level, while the radiography of the 2 hands face showed an acro-osteolysis of the 2nd finger of the right hand (Figure 2). The complement by the cervico-thoracic scanner did not objectify any scannographic anomaly as well as the cardiac echo-Doppler. The patient also underwent an ophthalmological examination which revealed a Schirmer test $\leq$ 5 mm/5 min. The diagnosis of scleroderma with a secondary Gougerot-Sjogren syndrome is retained (according to the ACR-EULAR criteria) associated with a Chilaiditi syndrome as digestive involvement. The patient is put on a calcium channel blocker, corticosteroid therapy at a rate of 15mg/day given the joint damage and colchicine for cutaneous calcinosis. However, for the Chilaiditi

syndrome, the gastrologist put the patient on a laxative, an anti-spasmodic and a proton pump inhibitor after clearly explaining the lifestyle and dietary measures.



Figures 1: CT images of colon interposition between the liver and the right diaphragmatic hemicupola

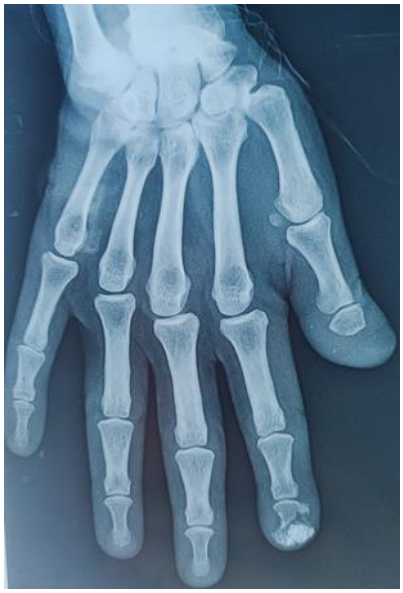


Figure 2: X-ray image of acro-osteolysis of the DIP of the 2nd finger

DISCUSSION

Chilaïditi syndrome was described in 1910, it corresponds to the interposition, most often asymptomatic and temporary, of the intestine between the upper surface of the liver and the right diaphragmatic dome. It is very rare with a frequency estimated between 0.2 and 2.5% of the general population [3]. CT scan appears to be the most reliable diagnostic tool for the detection of Chilaïditi syndrome, it shows interhepato-diaphragmatic digestive incarceration and makes the differential diagnosis with a sub-diaphragmatic abscess and pneumoperitoneum [4]. Risk factors for the syndrome include intestinal causes (abnormal motility, congenital malposition, etc.), hepatic causes (decreased liver size, laxity of hepatic suspensory ligaments), diaphragmatic causes (abnormally high diaphragm), and other causes (ascites, increased abdominal fat) [5]. The association of Chilaïditi syndrome in the literature with scleroderma is described in these 3 case reports [6-8]. In our patient, the predisposing factor for the onset of Chilaïditi syndrome is the decrease in intestinal motility that is seen during scleroderma. While the digestive attack during scleroderma is defined by a syndrome of intestinal pseudo-obstruction and which can be revealed by attacks of diffuse abdominal pain, constipation and meteorism of variable intensity, the Chilaïditi syndrome does not figure among the digestive manifestations of scleroderma given the number of cases reported in the literature.

CONCLUSION

Despite the fact that Chilaiditi's syndrome is rare and less reported in the literature as a sign associated with scleroderma, it deserves to be mentioned in scleroderma patients with digestive manifestations, and good knowledge of this syndrome makes it possible to avoid progression towards intestinal perforation.

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