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Case Report

Primary Sclerosing Cholangitis Simulating Hilar Plaque Cholangiocarcinoma in a Woman: A Rare Case Report

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Abstract: The inflammatory outbreak and fibrosis of the intra or / and extra hepatic biliary areas demarcated the term of sclerosing cholangitis, this outbreak is blameable for the stenosis of the bile ducts as well as a trouble in the flow of bile and consequently a cholestasis, which can lead to the development of fibrosis when prolonged. We are dealing with cirrhosis (biliary).

Keywords: Primary sclerosing cholangitis, cholestasis, biliary stenosis, female.

INTRODUCTION

Primary sclerosing cholangitis (PSC) is characterized by 3 phenomena: inflammation, fibrosis and progressive destruction of the intra- and extra-hepatic bile ducts or merely one of them, leading to biliary cirrhosis. The narrowing (stenosis) localized that deters the flow of bile defined cholestasis, the latter causes liver fibrosis (biliary cirrhosis). The term "primitive" means that the cause is not recognisable.

The aim of this work is to highlight the particularities of primary sclerosing cholangitis that might mimic cholangiocarcinoma and might lead to inappropriate surgical treatment. Through this work, we propose to report a case of primary sclerosing cholangitis mimicking a cholangiocarcinoma of hilar plaque in a 69-year-old woman.

OBSERVATION

A patient aged 69 years is presented to be observed, its ATCDs were limited to high blood pressure under treatment with amlodipine 5 mg per day, admitted for etiological balance of a cutaneous jaundice generalized, cholestatic-like, permanent mucous membrane with dark urine, discoloured stool, and generalized pruritus with no other digestive manifestations, including no abdominal pain and no exteriorized high or low gastrointestinal haemorrhage and no extra- digestive manifestations, evolving for 3 months in a situation of asthenia and alteration of the general state.

The somatic examination was without peculiarities apart from a free cutaneous icterus and scratching skin lesions.

The biological balance objectivated a biological cholestasis with alkaline phosphatases at 178 IU/L, or 1.8 times the upper limit of the normal value, Gamma-GT at 180 IU/l, or 1.7 times the upper limit of the normal value and cytolysis with ASAT at 100 IU/l, either 3 times the upper limit of the normal value and the ALAT at 188 IU/l or 5.3 times the upper limit of the normal value and total bilirubinaemia elevated to 238 mg/L, predominantly conjugated to 210.5 mg/L, a prothrombin level low to 36% (positive Koller test), tumour markers were negative, including 19-9 carbohydrate antigen (CA

19-9) and the rest of biologic was without anomalies.

An abdominal-pelvic CT was performed in the patient objecting a homogeneous steatosis liver, dented contours, no detectable nodule and no dilatation of the intra-hepatic bile ducts with a main ectasic bile duct measuring 9.6 mm without scannographically visible obstructions.

The imaging was complemented by a biliary magnetic resonance imaging objectified a typical appearance of primary sclerosing cholangitis with the presence of a hepatic hilum stenosis and two main bile duct stenosis and dilatation of the hepatic duct left with rarefaction of the intra-bile ducts distal liver (Figure 1).

These arguments led to the diagnosis of primary sclerosing cholangitis after elimination of other diagnoses. The patient was treated with ursodeoxycholic acid (AUDC) at a dose of 15 mg/d with a good clinical and biological evolution marked by the disappearance of symptoms and normalization of liver balance

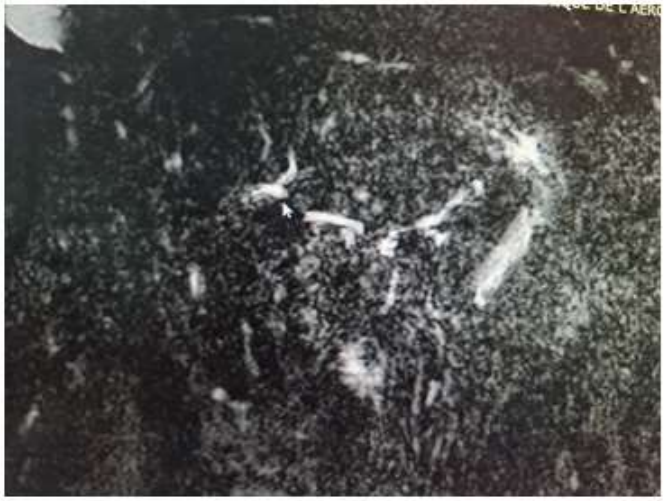

Fig-1: Biliary MRI objectifying two main bile duct stenosis and dilatation of left liver canal

DISCUSSION

Primary sclerosing cholangitis (PSC) is an inflammatory of the biliary tract of etiology not yet determined, whose autoimmune mechanism is suspected. It is characterized by stenosing fibro-inflammatory lesions of the bile ducts [1].

MCP occurs most often in a man (2/1 M/F sex ratio), at a median age of 40 years at diagnosis, often carrying a chronic inflammatory bowel disease (IBD) [2].

The pathogenesis of SPC is not yet determined, but several hypotheses of immunological and non-immunological mechanisms have been suggested. The diagnosis of primary sclerosing cholangitis is based on the combination of 4 types of signs shown in the table (Figure 2). The correlation between its signs is weak [3].

- **Biologics (Liver Test): Cholestasis (sometimes minimal)**
- **Radiological (Bili-MRI): The observed abnormalities are stenosis often long and multiple, typically without clear upstream dilatation; a rosary aspect is very suggestive; wall irregularities, or even diverticular aspects are possible [4].**
- **Histology (Liver biopsy) Peribiliary portal inflammation, discretely atrophic appearance of bile ducts without peri-ductal fibrosis, ductular reaction (proliferation) or ductopenia [5].**
- **Association with IBD (Colonoscopy with systematic biopsies)**

➢ **The diagnosis of SPC is retained in the presence of two (including at least the histological or radiological criterion) of these four criteria, in the absence of any other identifiable etiology.**

Fig-2: Diagnosis of primary sclerosing cholangitis

The clinical presentation of PSC is highly variable. The patient may be clinically asymptomatic and therefore the diagnosis arises only before abnormalities of the liver balance, or the patient may complain of asthenia, pruritus, pain of the right hypochondrium, or the occurrence of repeated angiocholites, or the diagnosis arises before an advanced stage of the disease with decompensated cirrhosis.

Chronic cholestasis with or without jaundice is the main biological manifestation of CSP. Radiologically, PSC manifests itself in lesions typical of the bile ducts on imaging. Bile duct imaging was previously performed by endoscopic retrograde cholangiopancreatography (ERCP) and is now performed by magnetic resonance imaging (cholangio-MRI), a non-invasive technique with very good sensitivity for positive diagnosis of PSC. Thus, cholangio-MRI (bili-MRI) is the recommended first line for the diagnosis of CSP [6]. The identification of multiple biliary stenoses associated with dilatations consists of the radiological diagnosis of SPC [7].

Histologically, fibrous cholangitis that occurs as a concentric lamellar fibrosis around a bile duct that can lead to a ductal obstruction is the typical lesion of sclerosing cholangitis [8].

The combination of the following three criteria: chronic cholestasis, typical lesions to imaging of the bile ducts or liver histology and the absence of arguments for a cause of secondary sclerosing cholangitis allows the positive diagnosis of CSP [9].

The evolution of the SPC is very variable. While some patients will have repeated angioscopies, others will progress to secondary biliary cirrhosis and others remain asymptomatic. The median survival time without liver transplantation is estimated at 21.3 years [10]. Age, bilirubinaemia, histology fibrosis score, and hepatic elastomeric as measured by Fibro Scan are the prognostic factors during SPC [11].

Apart from cirrhosis, patients with CSP are at risk of developing cholangiocarcinoma (risk* 400 times). In 50% of cases, cholangiocarcinoma occurs in the first year following the diagnosis of CSP. After the first year, the risk of cholangiocarcinoma is estimated at 0.5 to 1.5% per year. The occurrence of cholangiocarcinoma is related to several favouring factors: advanced age at diagnosis, male, history of dysplasia or colorectal cancer, and the presence of haemorrhagic recto colitis-type IBD versus Crohn's disease or absence of IBD [12].

Patients with PSC are also at risk of developing gallbladder lesions: calculus or tumor (50% of which will be malignant). Gallbladder carcinoma affects 3% of patients with SCD leading to annual gallbladder imaging [13].

Ursodeoxycholic acid (AUDC) is the only current treatment for PSC. It showed a significant biological improvement in biology but not in their survival. Two studies reported a deleterious effect of AUDC during PSC: an excess liver mortality and an over-risk of colorectal cancer [14, 15]. Its 2 effects led the American Association for the Study of Liver Diseases (AASLD) to ban treatment with AUDC during PSC. Although in both studies, AUDC treatment was given at high doses (28-30 mg/kg/d). For this reason, the European Association for the Study of the Liver (EASL) proposes the treatment of PCB by AUDC at a moderate dose of 15-20 mg/kg/day but does not formally recommend it [6].

Endoscopic treatment is indicated in case of symptomatic tight biliary stenosis, it consists in dilating the biliary stenosis either by a balloon or by the temporary placement of a small prosthesis (stent) biliary.

Not forgetting the treatment of other symptoms such as pruritus and asthenia, Therapeutic management should also include the treatment of possible complications (including osteoporosis or dyslipidemia secondary to chronic cholestasis) [6].

Liver transplantation remains the only treatment in case of decompensated cirrhosis or significant impairment of quality of life (recurrent angiocholites, refractory pruritus. The risk of recurrence of SPC on the liver graft estimated at 20% [1]. However, the efficacy of transplantation is good with a 5-year survival of 75%.

CONCLUSION

SPC remains a difficult diagnosis and treatment disease, often in 40-year-old men, rarely reported in women over 60 years of age, it is important to know the different clinical forms. A better understanding of the mechanisms involved is a key objective. AUDC remains the only current medical treatment at PSC.

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