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Original Article

Digestive involvement in retroperitoneal fibrosis

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ABSTRACT

Retro peritoneal fibrosis is a condition rarely seen in gastroenterology. Through these two observations and data from the literature we will address and focus on the digestive tract in the context of this disease and we will detail the different clinical aspects, radiological, pathological and therapeutic of this entity.

Key words: *Retro-peritoneal fibrosis; Digestive involvement; Ischemia*

INTRODUCTION

Retro-peritoneal fibrosis (RPF) is a rare disease, characterized by a chronic inflammatory process of the peritoneum with development of a fibrous tissue that engages the peritoneal structures including the abdominal aorta especially in its infra-renal portion. Its prevalence is in the order of 1.38 / 100000 with male predominance [1]. This pathology is most often followed by our colleagues nephrologists and urologists, but it can also be seen in gastroenterology since the attack can have a clinical statement simulating a digestive pathology, as is the case of our various observations. Through this work, we report 2 cases of idiopathic retro peritoneal fibrosis with an extension towards the mesenteric vessels, and we will focus on the digestive involvement in the peritoneal fibrosis.

MATERIALS AND METHODS

Observation 1:

Mr. Z.H, 28 years old, with no medical history, admitted to left iliac fossa pains with notion of postprandial vomiting and weight loss , six months before his consultation. The initial clinical examination showed a curvature in the left flank with impaction on palpation. The rest of the exam was normal. The patient had a biological assessment showing normocytic anemia (Hb = 11.2 g / dl), with a CRP of 9 mg / l, the renal function was correct. Abdominal ultrasound showed at the level of the epigastric sections the presence of a heterogeneous hypoechoic mass of 9 cm reaching the contact of the aorta enclosing the superior mesenteric artery and pushing back the body of the pancreas, with a small peritoneal with agglutination and thickeness in the proximal small bowel (Figure 1). The abdominopelvic CT confirmed the presence of an infiltrating process of the mesenteric root measuring 116 x 73 mm, including mesenteric vessels with thrombosis of the superior mesenteric vein and collateral mesenteric-vena cava venous circulation (Figure 2). It has been reported also a defect of raising the small loops (suffering) with a small intraperitoneal effusion. The guided ultrasound biopsy confirmed the diagnosis of RPF. As part of an etiological investigation of a secondary RPF, the thyroid, autoimmune, and infectious status were peculiar. No drug consumption, neither asbestos exposure, or a history of abdominal trauma were reported or observed. The diagnosis of primary RPF was retained and was placed on steroids (60 mg /day). One month after the start of treatment, the patient was readmitted for typical mesenteric angina with rectorragies and deterioration of the general condition. The biological assessment showed a microcytic anemia (Hb at 8.9g / dl). The abdominal CT showed colic distention with thickening of the digestive walls and enhancement of target at portal time (submucosal edema) with presence of tissue coelio-mesenteric and mesenteric root compressing mesenteric vein that appears non-opacified concluding to entero- mesenteric ischemia of venous origin. The colonoscopy showed a very congestive fragile mucosa that may be consistent with ischemic colitis .

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The patient was put under anticoagulation and corticotherapy with a good clinical evolution. An ultrasonographic examination at 4 months of treatment showed the persistence of a mass on both sides of the superior mesenteric artery in contact with the aorta without vascular compression, which measures 2.95 cm in transverse diameter around the superior mesenteric artery. The patient then decided to stop treatment with corticosteroids for adverse effects after 7 months from the beginning. At 14 months of diagnosis, the patient has been always followed regularly in consultation, with a good clinical evolution. The biological does not show any anomaly. The control ultrasound shows a stability of the measurements of the retroperitoneal process.

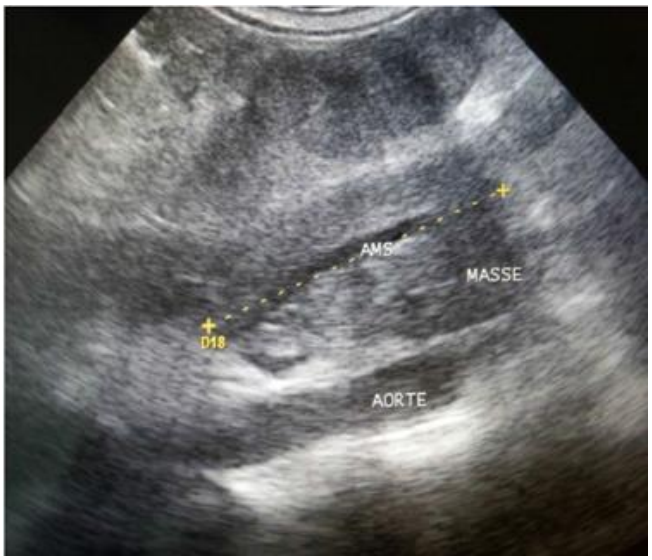

Figure 1: Ultrasound images showing mass tissue encompassing the mesenteric artery

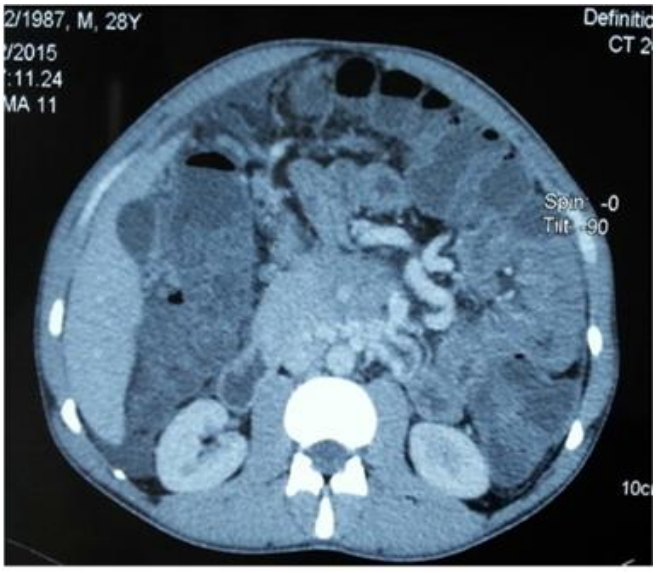

Figure 2: CT image showing the lesion process of the mesenteric root .

Observation: 2

Mrs B.S., 34 years old, with no medical history, was admitted for abdominal pain, vomiting and a dysenteric syndrome with a recent weight loss . The clinical examination was normal. The abdominal ultrasound shows at the epigastric sections a mass of 7 cm surrounding the aorta and the mesenteric artery, repressing the celiac trunk and the splenic artery. This mass is continued until the left renal pedicle responsible for a left pyelocalicielle dilation.

Abdominal CT confirms the presence of 2 hypodense fibrous sheaths low-raised, one encompassing the superior mesenteric artery and vein, which appears to be reduced in size but remains permeable and extending across L1 to L3. The other encompassing the aorta, the iliac branches (Figure 3) and the left ureter with uterine hydronephrosis upstream (anterior diameter - posterior pyelon = 30 mm). It has also been described a raised thickening in halo (submucosal edema) of a few ileal loops as well as the right colon with coeliomesenteric adenhegalies of small axis infracentimetric.

Abdominal CT concluded a suggestive aspect of retroperitoneal fibrosis with ileocolic wall thickening ischemic (chronic entero-mesenteric venous insufficiency). The biological assessment showed normocytic anemia with a sedimentation rate (SR) of 72 mm and a CRP of 58 mg / l, renal function was correct and stool parasitology was negative. The patient had initially benefited from a JJ urinary catheter surge to treat ureterohydratephrosis. Ultrasound biopsy with an anatomopathological study concluded with retro-peritoneal fibrosis. Complementary examinations were carried out in search of RPF cause: the thyroid assessment was correct, the autoimmune and infectious balance including the search for tuberculosis was negative.

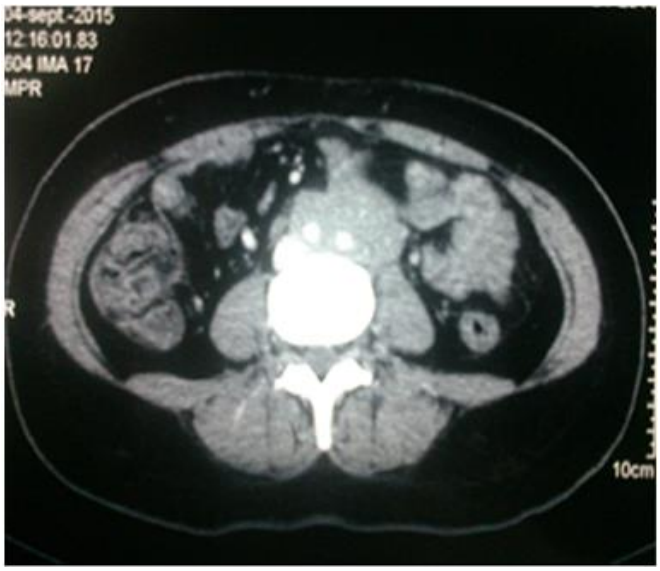

Figure 3 : CT image showing tissue mass including the 2 iliac branches.

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Colonoscopy had returned in favor of ischemic colitis. The patient was put on steroids at a rate of 1 mg / kg for months and then progressive depression, the patient is currently under 10 mg / day. The clinical course was good with standardization of (SR) . The 6-month follow-up ultrasound showed the persistence of a sub-pancreatic mass of hypoechoic echostructure measuring 55x26x21 mm. It is not noted pyelocalicielle dilatation. The patient is currently at 10 months of follow-up, always on corticosteroid 10 mg / day.

DISCUSSION

RPF was first described in 1905 by the French urologist Albaran. In 1948, 2 similar cases were reported by John Ormond; since then, this pathology has been considered as a separate entity and has been called "Ormond Disease". [1,2] Its pathogenesis is not fully understood. There are two types: the so-called "idiopathic" RPF where no cause is found, and which represents 2/3 of cases. The remaining third is usually secondary to a cause such as neoplasia, infections, trauma, radiotherapy, surgery and some drugs. Epidemiological data are rare in the literature. According to a Finnish study, the incidence is 0.1 / 100,000 inhabitants / year with a prevalence of 1.4 / 100,000 inhabitants. Usually the most common and most typical clinical picture is a male patient predominantly between the fifth and sixth decade with obstructive urinary tract syndrome [1,3]. There is no pathognomonic sign of this disease before becoming compressive. Pain is the most common clinical sign early in the illness. It can be of different seats and types. The digestive disorders are varied and are mainly functional to the type of heartburn, nausea, vomiting, constipation and anorexia. They seem to be the noisy translation of a renal failure that is often latent or secondary to the reflex reaction of acute dilation of the excretory cavities. The peritoneum itself is a reflexogenic organ and its isolated involvement can explain the digestive symptomatology.

Digestive syndromes have their origins in the irritation of the autonomic retro-peritoneal nervous system by inflammatory phenomena. The direct attacks of the digestive tract by the RPF remain exceptional. Direct compression of the digestive tract is not common but is described in the literature. This compression may be responsible for an intestinal transit discomfort with repetitive vomiting. Duodenal obstruction has been reported and may be manifested by nausea, vomiting, and weight loss[4,5]. Cholestatic jaundice may be noted in cases of distal canal involvement, which is rare and difficult to distinguish from cholangiocarcinoma or pancreatic cancer [6]. Chronic ischemia of the small intestine can be manifested primarily by abdominal angina punctuated by the meal with diarrhea and weight loss[7]. The intestinal complications are possible in the 3 sites: the mesentery of the small intestine which is the most frequently reported intestinal complication, the retroperitoneal mand recto sigmoid duodenum. Colonic obstruction has been infrequently reported in the literature (13cas). Extensive fibrosis can cause intestinal obstruction. In a multicenter German study involving 204 patients, 4 patients (2.0%) had extensive fibrosis resulting in bowel obstruction requiring colostomy. This rare and severe complication must be recognized[8]. Duodenal obstruction has also been described with fewer than 15 cases reported in the literature.[9] Cases of gastrointestinal bleeding due to portal hypertension secondary to retro-peritoneal fibrosis have also been reported (4 cases) [10]. Clinical examination is often poor and rarely contributive. Severe edema of both legs due to extension or compression of the inferior vena cava can occur at the late stage [11]. Renal artery stenosis may lead to renal vascular hypertension. The urological semiology of RPF is diverse but may be absent. The absence of ureteral involvement is the case of one of our patients, may be attributed to initial site of fibrosis in this case because fibrotic change usually occurs and extends from the pelvis edge upwards.[12] The biological exploration of RPF typically finds an increase in markers of inflammation associated with inflammatory anemia, and an alteration of renal function that can be noted in advanced forms. The value of these data must be recorded and will serve as a reference for monitoring the progress of the disease under treatment Cholestasis has been found in patients with an extension of fibrosis. In the bile ducts or associated pancreatic involvement [6,13]. Imaging plays a key role in this process by making it possible to highlight plaque, assess its extent and guide biopsies for confirmation of diagnosis. It can also be an aid in the search for a secondary cause. Conventional radiography is useless except in cases of complications (bowel dilation secondary to obstruction or pneumatosis related to infarction). Ultrasound is usually the first exam used. This is the case of our two cases.It allowed us to suspect the diagnosis and also the histological confirmation of this diagnosis by performing an ultrasound guided biopsy at the mass. It often shows a repercussion on the upper urinary tract (often bilateral ureteropelocalicic dilatation) and may be useful in the detection of coexistence of biliary abnormalities, portal hypertension by compression of the portal vein, sometimes even pancreatic involvement. The abdominopelvic CT is considered the radiological reference method in establishing the diagnosis. Finally, it makes it possible to differentiate malignant RPF from non-tumoral forms and distinguish between "active" inflammatory fibrosis and "old" mature fibrosis.[14] The therapeutic management of RPF has long been a source of controversy because of the rarity of the condition, the lack of controlled studies and official recommendations. Currently, there is no treatment that can permanently block the evolution of this disease. The treatment can be medical, surgical in case of complications or the combination of both. Medical treatment is essentially based on corticosteroids or other immunosuppressive drugs. Corticosteroid therapy alone can be effective without surgery for duodenal involvement[15]. Recently, some authors have highlighted the benefit of tamoxifen in the treatment of RPF. In all cases, a cause triggering or associated with RPF should be investigated, concomitantly treated, and any drug treatment identified as a possible cause of RPF should be stopped. Surgical treatment is indicated in case of RPF associated with aortic aneurysm. In these situations, the introduction of preoperative corticosteroid therapy is too risky. In some cases, the combination of vascular and urologic surgery (ureterolysis, microsurgery, stent) may be necessary.[16]

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In case of extension to the superior mesenteric artery, the establishment of a stent can be proposed and has been reported with good progress.[17] Treatment response criteria include improvement of symptoms, significant decrease in inflammatory biomarkers, and improvement of renal function. Evolution of the disease in the absence of treatment is unpredictable. The extension of fibrosis to the mediastinum is a bad prognosis. Some spontaneous regressions were observed. This improvement is however inconsistent. The prognosis of idiopathic RPF is renal. Under treatment, between 67% and 80% of patients do not have a renal sequela. No marker can predict relapse. Late relapses are possible. Prolonged monitoring is therefore justified. The prognosis of RPF is generally good with a ten-year survival of more than 70%[16].

CONCLUSION

Retro-peritoneal fibrosis is a rare inflammatory disease. Digestive involvement in retroperitoneal fibrosis is unusual. Extension to superior mesenteric vessels is even rarer. The symptomatology is not very specific. Treatment mainly involves corticosteroids and surgery. The prognosis is generally good, but with the possibility of late recurrences requiring prolonged clinical, biological and radiological monitoring.

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