Case Report

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Chronic Autoimmune Pancreatitis Mimicking A Tumor Of The Head Of The Pancreas

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1. Introduction

Several pathologies of the pancreas can manifest as solid lesions mimicking tumoral masses, notably a tumor of the head of the pancreas. 5 to 10% of pancreatectomies are performed for pseudo-tumoral lesions of the pancreas, considered as tumors preoperatively and diagnosed as benign pathologies on anatomopathological examination. These include sarcoidosis, intrapancreatic accessory spleen, lymphoid hyperplasia, lymphangioma, endometriosis and chronic pancreatitis [1]. The two forms of chronic pancreatitis that can cause a pseudotumoral mass that is difficult to diagnose as carcinoma are autoimmune pancreatitis and paraduodenal pancreatitis [2]. autoimmune pancreatitis may be isolated (type 2) or part of IgG disease (type 1). Type 2 autoimmune pancreatitis is defined by the destruction of the ductal epithelium by neutrophils. Symptoms appear at around 40 years of age. Twenty to 30% of patients have associated chronic inflammatory bowel disease [3]. diagnosis remains difficult, even with pancreatic biopsy.the fear is to perform major surgery for a benign lesion that responds to corticosteroids. We report the case of a 67-year-old patient who presented with jaundice and a diagnosis of unresectable carcinoma of the head of the pancreas due to vascular relationships. After a double biliary digestive bypass, the diagnosis of type 2 autoimmune pancreatitis was made on the basis of a pancreatic biopsy after total disappearance of the mass in the head of the pancreas.

2. Clinical Case

A 67-year-old man, with no particular pathological history, a smoker, a weaned alcoholic, who presented with a 10 kg weight loss over the last six months, with naked icterus evolving in one piece, pruritus, epigastralgia and vomiting. Clinical examination revealed mucocutaneous jaundice, a large palpable vesicle and generalized scratching lesions, with no evidence of a palpable epigastric mass. Biological tests revealed cholestasis with normal pancreatic enzyme levels and high levels of the cancer antigen CA 19-9 at 520 U/ml, although this is not specific for tumour lesions. Other blood tests were normal; abdominal, thoracic and pelvic computed tomography (figure 1) scans showed a retroglandular mass of the head of the pancreas with biductal dilatation, closely related to the portal trunk, superior mesenteric vein and second duodenal portion, and the presence of left latero-aortic adenomegaly; no secondary localization.

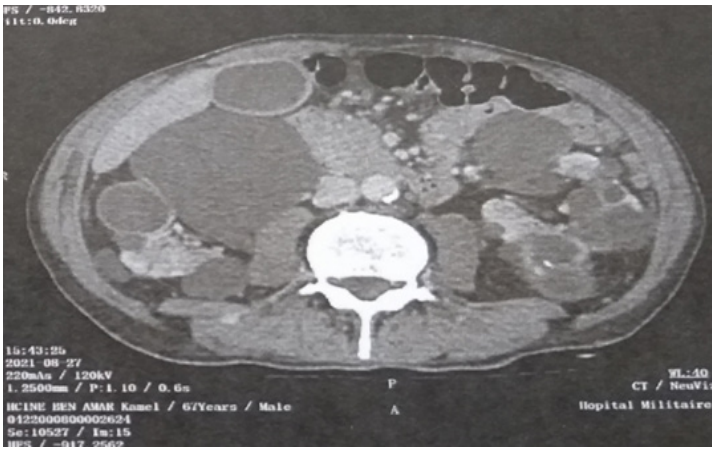
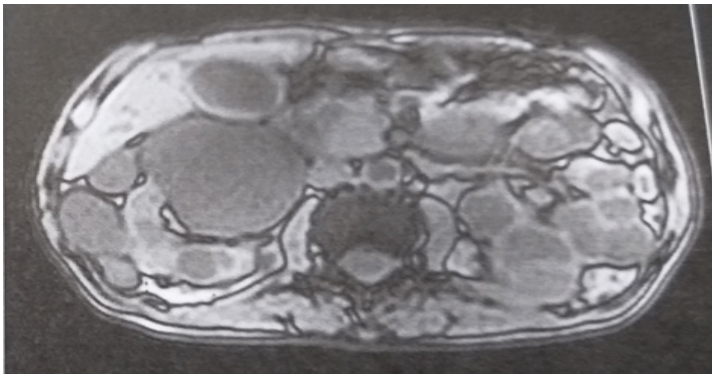


Figure 1: abdominal, thoracic and pelvic computed tomography scans showed a retroglandular mass of the head of the pancreas

Magnetic resonance imaging of the pancreas showed a locally advanced tumour mass of the pancreatic head with close vascular relations to the porto-mesenteric venous network, 50 mm long, associated with pancreatic hypotrophy (figure 2).



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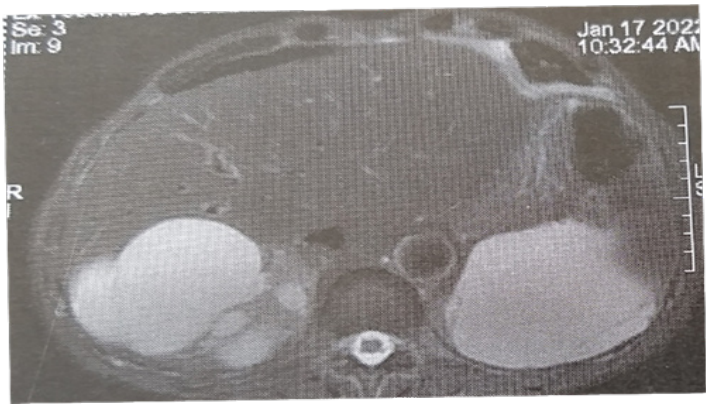

Due to concerns about pancreatic malignancy (pancreatic adenocarcinoma), the patient underwent subcutaneous surgery. Intraoperatively, he had a mass in the head of the pancreas invading the portal trunk, and underwent a biliodigestive bypass with a simple postoperative course. He received two sessions of adjuvant Folfirinox-based chemotherapy. A CT scan after two sessions of chemotherapy, at 1 month post-op, showed that the tumor mass had disappeared, thus ruling out the diagnosis of pancreatic adenocarcinoma. An IgG 4 assay came back normal, ruling out the diagnosis of autoimmune pancreatitis and justifying his being put on corticosteroids with a good evolution.

3. Discussion

Autoimmune pancreatitis was first described in 1950 by Ball [4]. It is a rare pathology, which explains the absence of an international consensus defining diagnostic criteria. autoimmune pancreatitis is a rare form of chronic pancreatitis characterized by an autoimmune inflammatory process leading to pancreatic fibrosis and dysfunction [5]. There are two types of autoimmune pancreatitis: type 1 pancreatitis, defined by the pancreatic involvement of a systemic IgG4-associated disease affecting several organs, with abundant infiltration of IgG4-positive plasma cells; and type 2 pancreatitis, as in our case, defined by the presence of an epithelial granulocyte lesion. The difference between type 1 and type 2 is the absence of IgG4-positive plasma cells in the inflammatory infiltrate and normal serum IgG4 levels in type 2 pancreatitis [6]. There are two forms of autoimmune pancreatitis, segmental and diffuse [3]. The segmental form is localized preferentially in the cephalic region, and may present as a pancreatic cancer, since both may present with abdominal pain, weight loss and cholestasis [7]. Differentiation between the two forms is extremely important, given the difference in management [8].

The symptoms of autoimmune pancreatitis are not specific. The most frequent clinical presentation is cholestatic jaundice caused by a mass in the head of the pancreas compressing the lower bile duct [9], in 50% of cases accompanied by abdominal pain. Symptoms may include asthenia, fever and weight loss. Other, rarer symptoms include diabetes and pancreatic insufficiency [10]. The diagnosis of PAI is based on a number of factors, in addition to the clinical features, including biological and radiological criteria. The study of CA 19-9 levels in discriminating between a carcinoma of the pancreatic head and a pseudotumor mass has been the subject of several studies, with differing results. Morris-Stiff et al proposed a threshold of 70.5 U/mL to distinguish malignant from benign lesions, with a specificity of 85.9% [11]. Elevation of CA 19-9 remains non-specific for distinguishing malignant lesions from pseudotumor masses. There is no effective radiological diagnostic tool for differentiating IAP from cancer. The most typical CT appearance of acute pancreatitis is diffuse enlargement of the gland, with a hypointense peripheral border known as a sausage-like appearance. Other CT findings include parenchymal delobulation, stenosis of the main pancreatic duct without associated dilatation, and stenosis of the intra-pancreatic portion of the main bile duct [12]. In 30% of cases, autoimmune pancreatitis is associated with inflammatory bowel disease [6] .In 2011, the International Pancreatology Association published diagnostic criteria for type II AIP in the journal Pancreas, including pancreatic imaging criteria, histological criteria, association with IBD and response to corticosteroid therapy [13]. Treatment of autoimmune pancreatitis consists of steroids at an initial dose of 0.6 to 0.8 mg/kg for four weeks, with progressive tapering. The total duration of steroid therapy is 3 months. A follow-up abdominal CT scan is recommended after four weeks from the start of treatment.

4. Conclusion

The diagnosis of autoimmune pancreatitis is complex. Histological evidence is rarely obtained after pancreatic puncture and demonstration of a granulocytic epithelial lesion. Diagnosis is based on radiological morphological criteria, the presence of associated IBD and response to corticosteroids. Once the diagnosis has been made, there is no need to be complacent, as precancerous lesions may be discovered or even absent, warranting long-term surveillance.

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