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IgG4 Disease Revealed by a Type 1 Autoimmune Pancreatitis: A Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. Author BA designed the study. Author LALM reviewed the manuscript. Authors KLRM and KYY wrote the first draft of the manuscript. Authors RVD and KYL managed the literature searches. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Aim: To describe clinical, paraclinical and evolutive features of an IgG4 disease.
Presentation of Case: The patient was a male, 51 years old, seen in consultation in the gastro-enterology and hepatology department of University teaching hospital "Campus" of Lome for epigastric pain of sudden onset six days before admission, with transfixing irradiation, evolving intermittently in a febrile context. On admission, the patient has a subicterus. The evolution during hospitalization was marked by the sudden onset three days later of bilateral inflammatory

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polyarthritis of the knees and ankles. Biological examination revealed diabetes mellitus; lipasemia was elevated to 122U/l. There was a disturbance of the renal balance; and a significant increase of plasma IgG4 at 2.380 g/l. The abdominal ultrasound showed a globally hypertrophic pancreatic gland, of heterogeneous structure, without focal lesions or necrosis; without biliary lithiasis. An abdominal CT scan showed, after injection of contrast medium, a global hypertrophy of the pancreas with a corporal-caudal predominance, giving a classic "sausage" appearance; associated with a lack of enhancement of the pancreatic parenchyma.
Conclusion: Type 1 autoimmune pancreatitis is the pancreatic manifestation of IgG4 disease. It is a rare pathology of recent discovery, with a predominance in elderly and male subjects. The diagnosis must be evoked in front of a pancreatic pain associated with articular manifestations, or in case of absence of obvious etiology of a pancreatitis. This diagnosis is defined by recommandations of International Association of Pancreatology. The treatment is based on the use of corticosteroids unlike autoimmune pancreatitis type 2 which is self-limiting with a low risk of relapse and complications.

Keywords: Pancreas; pancreatitis; autoimmune; IgG4.

1. INTRODUCTION

IgG4 disease is a systemic disorder characterized by an increase in serum immunoglobulin G subclass 4, associated with a lymphoplasmatic infiltrate of the various affected organs [1]. The organs frequently affected are: the pancreas, bile ducts, salivary glands, kidneys, aorta and lymph nodes [1-3]. Pancreatic involvement (type 1 autoimmune pancreatitis) is characterized by jaundice, altered general condition, abdominal pain, pancreatic mass and diabetes due to pancreatic insufficiency [4]. Because of its clinical and paraclinical symptomatology, the major challenge in the diagnosis of autoimmune pancreatitis lies in the differential with pancreatic or biliary cancer [5]. The prevalence and incidence of autoimmune pancreatitis in the general population are not known [6]. This is related to the fact that it is a rare disease of recent description. The majority of cases in the West are diagnosed on resection specimens for suspected pancreatic adenocarcinoma [7]. The revised criteria for clinical diagnosis of autoimmune pancreatitis (according to the Japan Pancreatic Society) include three arguments: morphological, biological and histological. The morphological argument is the demonstration of pancreatic hypertrophy associated with narrowing of the main pancreatic duct. The biological argument is the demonstration of autoantibodies or the increase of the serum level of immunoglobulin G4. The histological argument is the presence of fibrosis with infiltration of lymphoplasmacytic cells. The diagnosis of autoimmune pancreatitis is made when there is a morphological argument (major criterion) associated with a biological or histological argument (minor criteria) [8].

2. PRESENTATION OF CASE

The patient was a male, 51 years old, seen in consultation in the Gastro-enterology and hepatology department of University teaching hospital “Campus” of Lome, for epigastric pain of sudden onset six days before admission, with transfixing irradiation, evolving intermittently in a febrile context. The patient was neither an alcoholic nor a smoker and had no known personal (diabetes, lithiasis) or family history of digestive cancer, biliary lithiasis or crohn’s disease/hemorrhagic rectocolitis. On admission, the patient was found to have an altered general condition, a subicterus, and a normal-sized abdomen with diffuse tenderness, without any palpated mass or peritoneal irritation syndrome. The evolution during hospitalization was marked by the sudden onset three days later of bilateral inflammatory polyarthritis of the knees and ankles, associated with absolute functional impotence.

Biological examination revealed diabetes mellitus: fasting venous glycaemia at 6.40 g/l (N: 0.74g/l – 1.10g/l), qualitative glucosuria at four crosses, qualitative acetonuria at one cross, with glycated haemoglobin at 7.9%. Lipasemia was elevated to 122U/l (N<60U/l). There was a disturbance of the renal balance with urea=1.38 g/l (N:0.15g/l – 0.45g/l) and creatinine=35 mg/l (N: 7mg/l – 14mg/l); and a significant increase of plasma IgG4 at 2.380 g/l (N: 0.040g/l – 0.087g/l).

The abdominal ultrasound performed on admission showed a globally hypertrophic pancreatic gland, of heterogeneous structure, without focal lesions or necrosis; without biliary lithiasis. An abdominal CT scan showed, after injection of contrast medium, a global

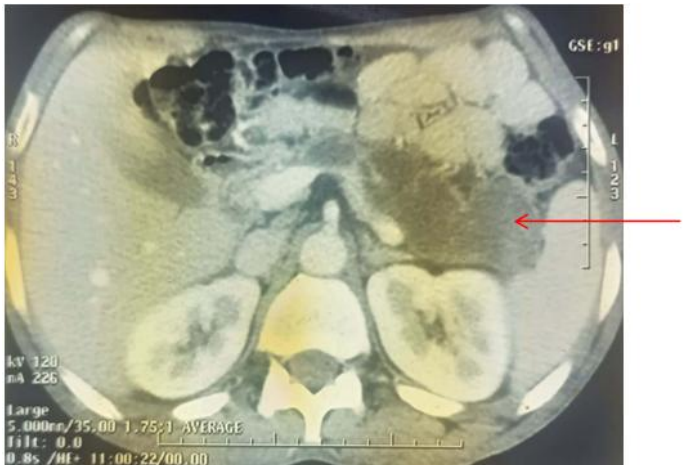

Fig. 1. Abdominal CT scan showing hypertrophy of the pancreas with lack of enhancement after injection of contrast medium (red arrow)

hypertrophy of the pancreas with a corporal-caudal predominance, giving a classic "sausage" appearance; associated with a lack of enhancement of the pancreatic parenchyma and an absence of dilatation of the Wirsung canal (Fig. 1).

The radiographic work-up showed: global joint pinching, hypertrophy of the tibial spines and calcification of the quadricipital tendon insertions with fluid effusion in both knees. In both ankles: pinching of the tibio-talar joint with calcaneal tendon calcifications.

The patient was treated with level 2 analgesics, regular insulin and corticosteroid therapy with 40 mg of methylprednisolone/day. The clinical evolution was made fifteen days after the beginning of the treatment towards a complete improvement of the symptomatology (icterus, fever, abdominal pains, arthralgias). The evolution of the biological assessment showed a negativation of glucosuria and ketonuria; normalization of venous glycemia, lipasemia and liver function tests. The control abdominal ultrasound showed a partial regression of the pancreatic hypertrophy. At the end of the 30 days of hospitalization, the patient was discharged with an insulin protocol and a diabetic diet. The corticosteroid dose was gradually reduced in 10mg increments every 14 days.

The diagnosis of type 1 autoimmune pancreatitis was therefore retained, subject to histological examination, in view of the imaging appearance correlated with the serology, the joint involvement and the favourable evolution under corticosteroids, in accordance with the recommendations of the International Pancreatology Association.

3. DISCUSSION

Autoimmune pancreatitis is a rare pathology of recent description [7]. We report in this study the first case described in the department of Gastroenterology and Hepatology of University teaching hospital “Campus” of Lome. Nevertheless, we believe that it is a pathology of underestimated diagnosis.

The case we report is that of a 51 year old male patient with no previous pathological history. Chari et al in the United States also reported a predominance of male patients (83%) and patients over 50 years of age (mean: 63+/-18 years) [9,10].

The symptoms of autoimmune pancreatitis are not specific. The most common clinical presentation is painless jaundice caused by a mass in the head of the pancreas. This symptomatology is similar to that of our patient. The extrahepatic manifestations were renal involvement and inflammatory polyarthritis. Rheumatoid factor assay was not performed; however, elevation of this factor has been reported in 25% to 29% of patients with autoimmune pancreatitis [4,11]. Our patient did not present any lymph node or lymphatic involvement as described in some series [12].

The biological work-up showed diabetes mellitus with proteinuria and acetonuria. This finding

confirms the data in the literature according to which autoimmune pancreatitis is often associated with pancreatic insufficiency [4] leading to the development of diabetes mellitus [13] in more than 80% of cases.

Serum IgG4 levels measured by nephelometry showed a specific increase. IgG4 is the best marker of autoimmune pancreatitis [14]. Its sensitivity is between 52% and 95%, and its specificity between 81% and 89% [15]. We made the diagnosis of autoimmune pancreatitis on the basis of a significant increase in serum IgG4 associated with global hypertrophy of the pancreatic gland; in accordance with international recommendations [8,16]. No histological analysis was performed.

The diagnosis of type 1 had been made, subject to histological examination in view of the epidemiological (age > 65 years; male sex), clinical (jaundice, absence of signs suggestive of a chronic inflammatory bowel disease), biological (increase in IgG4, diabetes) and evolutionary (remission after corticosteroid therapy) arguments.

4. CONCLUSION

Type 1 autoimmune pancreatitis is the pancreatic manifestation of IgG4 disease. It is a rare pathology of recent discovery, with a predominance in elderly and male subjects. The diagnosis must be evoked in front of a pancreatic pain associated with articular manifestations, or in case of absence of obvious etiology of a pancreatitis. This diagnosis is defined by recommandations of International Association of Pancreatology. The treatment is based on the use of corticosteroids unlike autoimmune pancreatitis type 2 which is self-limiting with a low risk of relapse and complications.

CONSENT

All authors declare that 'written informed consent was obtained from the patient for publication of this case report and accompanying images.

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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