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IgG4-Related Disease Presenting with Jaundice: A Case Report from Senegal

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Abstract

Introduction: IgG4-related disease is a recently recognized fibroinflammatory condition that can involve multiple organs. We report a case with pancreatic, biliary, and renal involvement. **Observation**: This was a 38-year-old patient presenting with jaundice associated with generalized pruritus and weight loss. He had a history of allergic rhinitis. Physical examination revealed jaundice and excoriations. Laboratory tests showed cholestasis and cytolysis. The serum IgG4 level was 3.63 g/L. Abdominal computed tomography (CT) and MR cholangiopancreatography demonstrated features consistent with autoimmune disease with pancreatic, biliary, and renal involvement. The clinical course was favorable under long-term corticosteroid therapy. **Conclusion**: IgG4-related disease is rare but probably underdiagnosed in Africa. It often has a favorable course with appropriately administered corticosteroid therapy.

Keywords

IgG4-Related Disease, Autoimmune Pancreatitis, Sclerosing Cholangitis, Nephritis

1. Introduction

The concept of IgG4-related disease was introduced in Japan in 2003 by Kamisawa *et al.* [1] on the basis of cases of type 1 autoimmune pancreatitis. This concept made it possible to group, under a single nosological entity, several long-recognized inflammatory disorders characterized by organ swelling, infiltration by IgG4-positive plasma cells on immunohistochemistry, storiform fibrosis, and a frequent but inconsistent elevation of serum IgG4 levels [2]. Publications on

IgG4-related disease are numerous in Asia, Europe, and the Americas, whereas in Africa, data remain limited and are mainly based on isolated reports [3] [4]. Here, we report a case from Senegal with pancreatic involvement associated with sclerosing cholangitis and nephritis.

2. Observation

This was a 38-year-old patient admitted for jaundice associated with generalized pruritus for 2 weeks, in a context of epigastric pain and a 10-kg weight loss over 4 months. He had previously undergone an abdominal ultrasound that was normal and an esophagogastroduodenoscopy (EGD) showing erythematous gastritis (not biopsied). His medical history was unremarkable, and he was a chronic alcohol user (30 g/week). Physical examination was normal except for jaundice and excoriations on the abdomen and upper limbs. The complete blood count and prothrombin time (PT) were normal. Total bilirubin was 221.2 mg/L, including 146 mg/L conjugated bilirubin; alkaline phosphatase (ALP) was 2.9× the upper limit of normal, gamma-glutamyl transferase (GGT) 1.8×, and aspartate aminotransferase (AST) and alanine aminotransferase (ALT) were each 4×. HBs antigen, anti-HCV antibodies, and anti-HIV antibodies were negative. Polyclonal hypergammaglobulinemia was present at 16.5 g/L. Serum IgG4 level was 3.63 g/L. Serum creatinine was 12.18 mg/L, corresponding to an estimated glomerular filtration rate of approximately 80 mL/min/1.73 m^2. Urinalysis was unremarkable. Abdominal computed tomography (CT) showed diffuse pancreatic enlargement with a peripheral hypodense rim without abnormalities of the peripancreatic fat (**Figure 1**), associated with dilatation of the common bile duct and intrahepatic bile ducts; peripheral hypodense foci were also seen at the lower pole of the left kidney. Pancreatobiliary MRI revealed diffuse pancreatic enlargement with loss of lobulations, T1 hypointensity and T2 isointensity, heterogeneous delayed enhancement, a subtle peripheral hypointense halo, and mild dilatation of the common bile duct with wall thickening and enhancement associated with distal stenosis (**Figure 2**); the gallbladder was empty with enhanced, thickened walls, and there were also T2-hypointense areas in the renal cortex that did not enhance after contrast administration, consistent with renal involvement. The diagnosis of IgG4-related disease was established based on the clinical and biological findings and the medical imaging features.

The patient was started on prednisolone at an initial dose of 0.6 mg/kg/day for 4 weeks, followed by gradual tapering, with an initial clinical and biological improvement. After 3 months of treatment, a partial decrease in the serum IgG4 level (1.497 g/L) was noted, along with persistence of mild dilatation of the common bile duct and a small area of hypodensity in the left kidney. Corticosteroid therapy was continued. At the 5-month follow-up, the outcome was favorable, with normalization of the serum IgG4 level to 0.933 g/L. Imaging showed complete resolution of the pancreatic and biliary abnormalities, as well as near-complete regression of the renal lesions, with persistence of a small residual posterior cortical hy-

podense area in the left kidney. The patient was maintained on prednisolone at a dose of 5 mg/day.

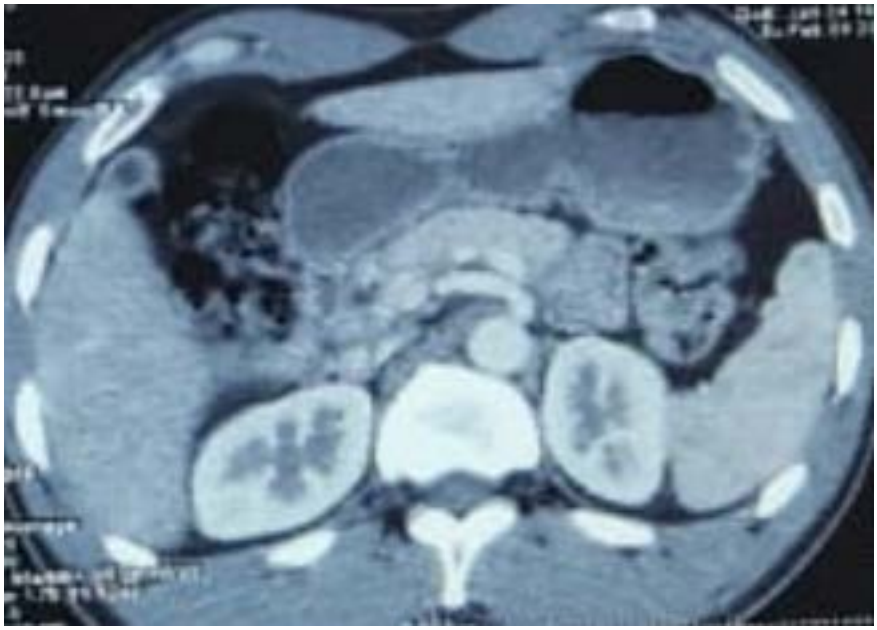

Figure 1. Abdominal CT scan showing diffuse pancreatic hypertrophy with a peripheral hypodense rim.

Figure 2. Biliary MRI showing mild dilatation of the common bile duct with associated wall thickening, together with a distal stricture, consistent with cholangitis.

3. Discussion

IgG4-related disease is a systemic fibroinflammatory disorder with an etiology that remains poorly understood. Its exact incidence and prevalence are not well established. In Japan, its incidence is estimated to range from 0.28 to 1.08 per 100,000 inhabitants [5]. It predominantly affects men around 60 years of age. In-

volvement may affect one or several organs, either synchronously or metachronously [6] [7]. The organs most frequently affected are the pancreas, bile ducts, salivary glands, and lacrimal glands [8]. In Africa, data on IgG4-related disease remain limited and are mainly based on isolated case reports or small case series, suggesting probable underdiagnosis in this setting [3] [4]. Our case illustrates a multiorgan form (pancreatic, biliary, and renal), notable for its early onset at 38 years of age.

The Comprehensive Diagnostic Criteria for IgG4-RD (CDC) are the most widely used criteria for diagnosing IgG4-related disease [8]. They apply regardless of the organ involved and are based on clinical, laboratory, imaging, and histological findings.

In our patient, jaundice and generalized pruritus indicated pancreaticobiliary involvement.

From a biological standpoint, aside from organ-specific signs of involvement, polyclonal hypergammaglobulinemia and elevated serum IgG4 levels are frequently observed. Our patient had an IgG4 level of 3.63 g/L, well above the diagnostic threshold of 1.35 g/L.

Imaging (CT and MRI) revealed organ involvement suggestive of the condition.

Histological examination remains the gold standard for diagnosis and is based on three main abnormalities: a dense lymphoplasmacytic infiltrate with IgG4 predominance and an IgG4+/IgG+ ratio > 40%, storiform fibrosis, and the presence of obliterative phlebitis [9].

The absence of histological confirmation is a limitation of our case. Pancreatic or biliary tissue sampling procedures, such as endoscopic ultrasound or ERCP, were not performed because of their limited availability in our setting, and no easily accessible biopsy site was identified. According to the CDC criteria, our patient had characteristic organ involvement and a marked elevation in serum IgG4 levels, but no histological confirmation. The diagnosis was therefore classified as possible IgG4-related disease. However, the application of organ-specific criteria further strengthened the diagnostic likelihood.

The pancreatic involvement observed in our patient met the criteria for type 1 autoimmune pancreatitis as defined by the ICDC (International consensus diagnostic criteria) [10], which are characterized on imaging (CT/MRI) by diffuse enlargement of the pancreas with loss of lobulation, producing the appearance of a "sausage-shaped enlarged pancreas." Contrast enhancement is typically delayed, and a peripheral subcapsular hypodense or hypointense rim that is almost pathognomonic may be observed [6]. Typical ductal abnormalities include narrowing, or even disappearance, of the main pancreatic duct over a length greater than 5 cm, without upstream dilatation or with moderate dilatation < 5 mm, or multiple strictures without upstream dilatation [6].

Unique or multiple extrapancreatic lesions, whether synchronous or metachronous, may also be associated.

IgG4-related sclerosing cholangitis (IgG4-SC) is observed in 65% - 85% of cases

of autoimmune pancreatitis and typically involves the extrahepatic bile ducts. It is characterized by circumferential, symmetric thickening of the bile duct wall and the development of strictures. In our patient, the diagnosis was established on the basis of the clinical diagnostic criteria for IgG4-SC published in 2020 [11]. MRI showed mild dilatation of the common bile duct with mural thickening, associated with a distal stricture.

The renal involvement observed is a classic manifestation of the disease, most often presenting as tubulointerstitial nephritis. It may be asymptomatic and detected on imaging, with bilateral pseudotumoral hypodense lesions on CT and hypointense lesions on MRI [7].

The main differential diagnoses were pancreatic adenocarcinoma, cholangiocarcinoma, and primary sclerosing cholangitis. The absence of a focal mass, the diffuse nature of the abnormalities, and the presence of a peripheral rim made pancreatic adenocarcinoma unlikely. The absence of a tumoral biliary lesion and the multiorgan pattern involving the pancreas, bile ducts, and kidneys argued against cholangiocarcinoma. Finally, elevated IgG4 levels, the association with autoimmune pancreatitis, and the favorable response to corticosteroid therapy supported IgG4-related sclerosing cholangitis rather than primary sclerosing cholangitis.

Corticosteroid therapy is the standard treatment for remission induction. The Japanese protocol used in our patient consists of an initial dose of prednisolone 0.6 to 1 mg/kg/day for 2 to 4 weeks, followed by gradual tapering until discontinuation at 3 months [6]. Despite an initial effectiveness of 98.5%, the relapse rate remains high, ranging from 26% to 70% [12]. Several experts therefore recommend low-dose maintenance corticosteroid therapy (2.5 to 5 mg/day) for 3 years. After 3 months of treatment, our patient achieved complete clinical remission, associated with a partial decrease in IgG4 levels. Imaging showed mild dilatation of the common bile duct as well as a small area of hypodensity in the left kidney, justifying continuation of corticosteroid therapy. In the event of relapse or steroid dependence, immunosuppressive therapy (azathioprine, mycophenolate mofetil) or rituximab may be considered.

4. Conclusion

IgG4-related disease has been the subject of numerous publications, initially in Asia and subsequently in Europe and the United States. In Africa, very few cases have been reported. It is a rare condition but is likely underdiagnosed in our countries. Our case report illustrates a rare and severe form with multiorgan involvement in a young adult. In the presence of certain clinical presentations, clinicians should consider and actively investigate this diagnosis, as the disease is highly responsive to corticosteroids and often has a favorable course when appropriately treated.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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