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Single Case

Autoimmune Pancreatitis Type 1 Associated with a Pancreatic Pseudocyst

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Keywords

Autoimmune pancreatitis · Corticosteroid · Pancreatic pseudocyst

Abstract

Pancreatic cystic lesions comprise diverse entities with different histopathological characteristics. Differential diagnosis is often challenging. Autoimmune pancreatitis (AIP) is usually not considered an underlying pathology in the differential diagnosis of peri-/pancreatic pseudo-/cystic lesions. We report the case of a 73-year-old male with diffuse pancreatic enlargement and an adjacent cystic lesion (60 × 80 mm) on computed tomography scan. Based on these imaging findings and an elevated serum IgG4 concentration, AIP complicated by a pancreatic pseudocyst was diagnosed, and treatment with glucocorticoids was started. Regular follow-ups showed a good response to treatment with regression of the pancreatic pseudocyst and remittent pancreatic swelling.

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Introduction

Autoimmune pancreatitis (AIP) is a fibroinflammatory disorder of presumed autoimmune etiology that is associated with characteristic clinical, histologic, and morphologic findings [1, 2]. Incidence and prevalence are yet undetermined; however, the prevalence rate of

AIP may account for 9% of the patients with nonalcoholic pancreatitis but is almost never observed in patients with alcoholic pancreatitis. Several publications have indicated that AIP type 1 is part of a systemic IgG4-related disease [3]. Clinical manifestations of AIP are varied and include mild, recurrent pancreatitis, pancreatic duct strictures but mild upstream dilation, diffuse sausage-like enlargement of the pancreas, and a pancreatic mass on imaging. Diagnostic criteria of AIP are summarized in the HISORt criteria, the Japan consensus criteria and the “International Consensus Diagnostic Criteria” from the International Association of Pancreatology and comprise histology, characteristic imaging, elevated serum IgG4 levels on serologic testing, other organ involvement (biliary strictures, parotid/lacrimal gland involvement, mediastinal lymphadenopathy, retroperitoneal fibrosis), and response of pancreatic and extra-pancreatic manifestations to glucocorticoid therapy [4]. First-line treatment is corticosteroids with a response rate of up to 98% [5]. Therapy should be started with prednisone at a dosage of 40 mg daily for 4 weeks followed by a taper of 5 mg/week [6]. While most patients initially respond well to glucocorticoids, a significant proportion of patients relapse once treatment is discontinued. Options are repeat course, maintenance therapy, or considering immunomodulators [7–9]. Complications of AIP comprise endocrine and exocrine failure, whereas pancreatic pseudocysts are very seldom [10]. Here, we report a case of AIP type 1 complicated by a large pancreatic pseudocyst and complete dissolving after steroid therapy.

Case Presentation

A 73-year-old male was referred to our department by Helios Clinic München Perlach with a high-grade suspicion of AIP. First presentation revealed painless jaundice, diarrhea, and a weight loss of 5% within 4 weeks. The initial body weight was 86 kg. There was no evidence of fever or abdominal pain. The patient did not have any history of alcohol abuse or previous liver or pancreatic diseases. Routine laboratory analysis showed increased liver function test results (alkaline phosphatase 781 U/L; aspartate aminotransferase 320 U/L; alanine aminotransferase 698 U/L; gamma glutamyl transferase 1,441 U/L; total bilirubin 6.02 mg/dL) and an elevated serum IgG4 concentration (1,190 mg/dL; normal range 3–200). Computed tomography revealed a diffusely enlarged pancreas (“sausage-like” with loss of lobulation), hypoattenuating, with adjacent edema and a cystic lesion (60 × 80 mm) extending from the pancreatic tail (Fig. 1). Before presenting at our department, the patient underwent biliary stenting due to extrahepatic cholestasis. Based on these findings, the patient was diagnosed with AIP and a pancreatic pseudocyst. A corticosteroid therapy was initiated with 40 mg prednisolone daily and tapered gradually for 30 weeks until drug discontinuation. Magnetic resonance imaging, performed after 6 months of therapy revealed a partial normalization of the pancreas with a remaining prominent duct, and partial regression of the pancreatic pseudocyst (26 × 11 mm) (Fig. 2). Consistently, serum IgG4 concentration also declined to 292 mg/dL after 4 weeks of prednisolone therapy and 159 mg/dL after 16 weeks of therapy (Fig. 3).

Discussion

Differentiating cystic pancreatic lesions remains a clinical challenge. In 1995, Yoshida et al. [11] reported that one of the characteristic features of AIP is the absence of pancreatic cysts. Here, we report a rare case of AIP associated with a pancreatic cyst, highly suggestive of a

pseudocyst based on the initial imaging presentation. Only few reports describing AIP complicated by a pseudocyst are published in the literature [12, 13]. Upon corticosteroid-based therapy, pancreatic swelling and the pseudocyst resolved without the need for drainage. Kawakami et al. [14] suggested that corticosteroid treatment should be started immediately upon appearance of a pseudocyst in AIP, as this may resolve under treatment. Cyst formation in AIP might represent a highly active inflammatory process [13, 15]. We found a high serum IgG4 concentration that closely correlates with the active state of this disease. The exact mechanism of the pathogenesis of the cyst is not completely understood, but stenosis of the pancreatic duct may have caused reduced drainage of pancreatic juice resulting in the pancreatic pseudocysts. Corticosteroids may induce pseudocyst regression through inhibition of inflammation of the pancreatic duct, thus reducing the stenosis and improving the drainage of pancreatic juice. In summary, our report shows the importance of including AIP as an underlying pathology in the early differential diagnosis of cystic pancreatic lesions and suggests that the pancreatic pseudocysts in patients with AIP can be treated successfully without invasive therapy. Corticosteroid therapy should be started immediately, and response to treatment should be controlled by regular imaging.

Statement of Ethics

Written informed consent was obtained from the patient.

Disclosure Statement

All authors have no conflicts of interest to disclose.

Author Contributions

Felix Hesse: Drafting the work and substantial contributions to the acquisition, analysis, and interpretation of data for the work.

Rickmer Braren: Substantial contributions to the conception of the work and analysis, such as the interpretation of data for the work.

Roland M. Schmid: Revising the work critically for important intellectual content, such as final approval of the version to be published.

Veit Phillip: Substantial contributions to the conception and design of the work, such as revising it critically for important intellectual content.

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Fig. 1. Contrast-enhanced computed tomography scan showing sausage-like enlargement with homogeneous attenuation of the pancreas, a peripheral rim of hypoattenuation “halo,” a loss of lobulation (arrows), and a caudally situated cystic lesion (60 × 80 mm; asterisk).

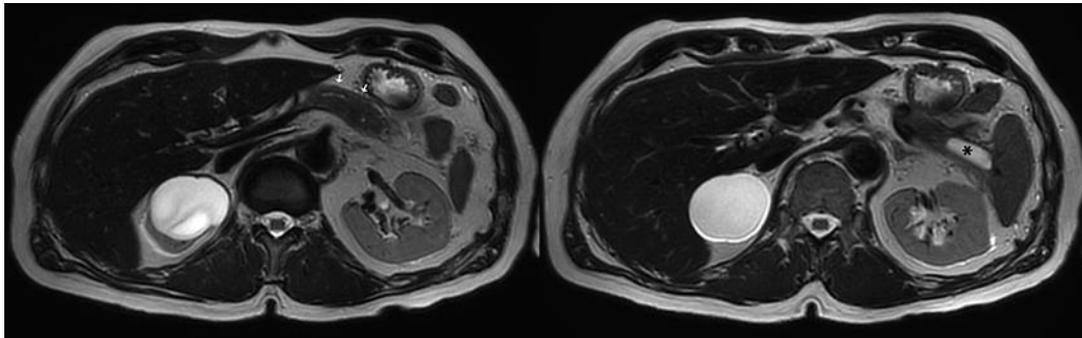


Fig. 2. Magnetic resonance imaging on T2-weighted sequences 6 months after initiation of therapy showing a less swollen pancreas with a prominent pancreatic duct (arrows) and a reduction in size of the caudally situated cystic lesion (26 × 11 mm; asterisk).

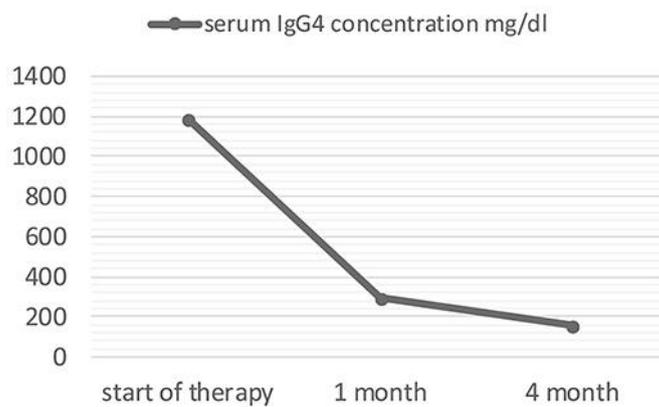


Fig. 3. Graph showing declining IgG4 levels during the therapy.