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## Imaging diagnostic value of autoimmune pancreatitis

Wang F *et al.* Imaging diagnostic value of AIP

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### Abstract

<sup>2</sup> Autoimmune pancreatitis (AIP) is a special type of chronic pancreatitis mediated by autoimmunity factors. It can be divided into two categories: <sup>5</sup> Lymphoplasmacytic sclerosing pancreatitis and idiopathic duct-centric pancreatitis according to pathological characteristics. In the clinical settings, the imaging manifestations of some AIP patients are atypical, so it <sup>1</sup> is difficult to distinguish it from general pancreatitis, pancreatic ductal adenocarcinoma, lymphoma and other malignant disorders. Most importantly, the treatment and prognosis of these diseases are different. Therefore, the timely correct imaging diagnosis of AIP is key for AIP patients. After that, clinicians can take the appropriate treatment measures for those patients, which is helpful for the prognosis of AIP.

**Key Words:** Autoimmune pancreatitis; Diagnosis; Computed tomography; Magnetic resonance imaging; Pancreatic cancer

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<sup>2</sup> **Core Tip:** Autoimmune pancreatitis (AIP) is a special form of chronic pancreatitis mediated by autoimmunity. Because the clinical presentation is not specific, therefore, the progresses of imaging diagnosis and antidiastole of AIP may increase the clinicians' awareness of AIP. In this case, timely intervention is very good for the patient. Clinicians can adopt the effective method in the early course of the disease. We promote

five related research directions. We hope that the clinical work and scientific research can start from these aspects in the future. AIP patients will get benefit through our continuous exploration of imaging differential diagnosis and prognosis assessment of AIP.

### TO THE EDITOR

We read the interesting case report of Zhang *et al* [1]. In that paper, it demonstrated that four patients with autoimmune pancreatitis (AIP) with positive Immunoglobulin 4 (IgG4) manifestation were reported. Although those patients rejected steroid treatment, their AIP symptoms spontaneously became remission during the disease courses.

Firstly, as for pancreatic imaging findings, three main features of the pancreas in those patients are given as follows: (1) The size and density or signal intensity of the pancreas returned to a normal condition; (2) there may be some signs related to progressive fibrosis within the pancreas; and (3) there may be atrophy and calcification signs of the pancreas. Secondly, with respect to clinical manifestations, the case report suggests that pancreatic atrophy and progressive fibrosis of the pancreas in those patients could cause pancreatic endocrine dysfunction, and following result in poor blood glucose control. In addition, it may lead to the left portal hypertension, as well as congestive splenomegaly and variceal bleeding. Based on AIP patients' phenomena, we agree with the authors' opinions. Once a confirmed diagnosis of AIP is made, steroid treatment should be used as early as possible, instead of ignoring it or allowing it for self-remission (SR).

Classically, AIP is divided into two subtypes including type 1 and type 2 [2]. On the one hand, type 1 AIP is called as lymphoplasmacytic cell sclerosing pancreatitis. Typical histological manifestations of type 1 AIP include substantial IgG4-positive lymphoplasmacytic infiltration and stromal mat-like fibrosis in the involved pancreatic tissue. Also, the similar pathological findings may be found in involved extrapancreatic tissues and organs. For type 1 AIP, the most important characteristic is IgG4-related

disease<sup>1</sup> associated with significantly increased serum IgG4 Levels. Those patients are sensitive to steroid hormone therapy. On the other hand, type 2 AIP<sup>1</sup> is termed as idiopathic ductal central pancreatitis. It is histologically characterized by massive granulocyte infiltration around pancreatic duct, and secondary local ductal epithelial injury. Unlike type 1 AIP, the most striking feature of type 2 AIP is the absence of IgG4-positive lymphoplasmatic cell infiltration. Generally, type 2 AIP cases show single pancreatic organ involvement, while the extrapancreatic tissues and organs are rarely involved.<sup>6</sup>

Medical imaging modalities play a vital role in diagnosis and differential diagnosis for AIP patients. In 2011, the 14<sup>th</sup> International Pancreatic Association formulated the international diagnostic criteria for AIP<sup>[3]</sup>. The international standards describe typical diagnostic characteristics of two-type AIP from six aspects, including imaging of pancreatic parenchyma, pancreatic duct imaging, serological findings, extrapancreatic lesions, histology, and hormonal therapy response. Also, evidence-based medicine emphasizes a need of imaging evidence for the AIP diagnosis. In clinical settings, a majority of AIP can be identified by imaging. Firstly, diffuse AIP often shows diffuse pancreas enlargement with reduced density like “sausage” on computed tomography (CT ) images<sup>[4]</sup> (Figure 1A), which is caused by the infiltration of a large number of lymphocytes and the proliferation of fibrous tissue in the lesion sites. The density of pancreatic lesions is uniform, as well as moderate enhancement after contrast-enhanced scan (Figure 1B and C). Interestingly, peripancreatic fat spaces become smaller and make the periphery of the pancreas into a low-density cystic edge, which is called “halo-ring sign” or “a capsule sign”<sup>[5]</sup>. In addition, contrast-enhanced scan can better show the capsule structure than plain scan, and it shows mild enhancement of the capsule during the venous or delayed phase. On the contrary, this sign is rare in ordinary chronic pancreatitis or pancreatic cancer. Furthermore, smooth stenosis at the lower end of the common bile duct can be shown in AIP patients with obstructive jaundice<sup>[6]</sup>. Secondly, the mass-type AIP often shows low density or iso-density mass in the head of pancreas on CT plain scan. And then, the lesion slight enhancement could

be seen in the arterial phase and obvious uniformity and <sup>1</sup>delayed enhancement in the venous phase. Also, the performance of magnetic resonance imaging (MRI) enhanced scan is similar to that of CT (Figure 1D-F). As for pancreatic duct, the main pancreatic duct of AIP patients is usually irregularly narrow and interrupted, and the involved range is consistent with pancreatic lesions. It should be noted that this kind of stenosis often exceeds <sup>1</sup>1/3 of the total length of the main pancreatic duct or appears as “a jumping stenosis”<sup>[7]</sup>, whereas patients with pancreatic ductal adenocarcinoma have limited pancreatic duct stenosis and interruption. In fact, to our best of knowledge, a few AIP patients without aforementioned typical imaging findings are misdiagnosed as common chronic pancreatitis. More importantly, radiological features of some AIP cases are especially similar to pancreatic cancer findings, which is easy to be misdiagnosed as pancreatic cancer for surgical treatment. According to previous data<sup>[8]</sup>, about 27% of AIP patients were misdiagnosed as suspicious pancreatic cancer and underwent Whipple resection. Indeed, this will delay the precious opportunity of effective hormone therapy for AIP patients. Therefore, early and correct imaging diagnosis is very critical for both asymptomatic and symptomatic AIP patients.

Based on this case report, we would like to put forward the following five aspects of consideration. First, it is unknown whether some characteristic changes of AIP images are related to spontaneous remission of AIP patients. Second, key radiological differences between asymptomatic and symptomatic AIP patients should be summarized. Third, it is possible to dynamically evaluate the prognosis of AIP patients through imaging. Fourth, the relationship between changes of serum indicators and the imaging performance of AIP is also interesting. Fifth, if the patients received steroid treatment according to evidence-based guidelines, the correlation between following imaging features and recurrence of AIP is not completely clear. The above points will be the direction of our future research.

To sum up, from the perspective of benefits for AIP patients, most AIP cases are sensitive to steroid hormone therapy. Hence, early diagnosis and timely treatment with steroid drugs should be advocated, regardless of symptomatic or asymptomatic AIP.

Majority of AIP patients have a good prognosis, but they need to be followed up through imaging and laboratory examinations for a long time to evaluate possible late complications. Only in this way, AIP patients will get the best benefit through our continuous exploration of imaging diagnosis and prognosis evaluation of AIP.

**Figure 1 Diffuse autoimmune pancreatitis.** A-C: Diffuse enlargement of the pancreas on pre- and post-contrast enhanced computed tomography scans; D-F: Progressive enhancement of the pancreatic lesion and the pseudocapsule on pre- and post-contrast enhanced magnetic resonance imaging scans.

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