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CASE REPORT

IgG4-related Autoimmune Pancreatitis Mimicking Acute Pancreatitis: A Case Report

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IGG4-RELATED autoimmune pancreatitis (AIP) is the most common type of IgG4-related diseases. The observation of a diffused and localized enlargement of the pancreas and strictures in the main pancreatic duct on radiological imaging is essential for diagnosing AIP. The clinical features of AIP which present as a localized pancreatic mass greatly mimic pancreatic cancer. Cases with AIP mimicking pancreatic cancer are reported numerously. However, cases of IgG4-related AIP manifesting acute pancreatitis symptoms and extensive peripancreatic effusion are rarely observed. Meanwhile, acute pancreatitis is a common disease. If misdiagnosed as acute pancreatitis, IgG4-related AIP may progress seriously. Here, we report a rare case of IgG4-related AIP suspected of acute pancreatitis because of epigastric pain and extensive peripancreatic effusion on computed tomography (CT) images.

CASE DESCRIPTION

A 63-year-old woman presented for a medical consul-

tation with epigastric pain that had started one month previously. The pain got worse after feeding, and radiated to back. She also had nausea, anorexia, fatigue, and mild fever in the afternoon. The highest temperature was 37.6°C. She did not feel better after taking rabeprazole. Physical examination showed abdominal distention, and upper abdominal tenderness without rebound tenderness or rigidity. No jaundice was seen. The initial laboratory results showed albumin at 30.8 (normal range 35.0-55.0) g/L, globulin at 39.2 (normal range 15.0-40.0) g/L, total bilirubin at 18.7 (normal range 3.4-25.0) μmol/L, directly bilirubin at 6.8 (normal range 0-7.0) μmol/L; serum amylase at 45 (normal range 0-115) U/L, serum lipase at 45.8 (normal range 0-60.0) U/L; cancer antigen (CA)19-9 at 9.2 (normal range <35) U/ml, CA125 at 101.89 (normal range <35) U/ml, carcinoembryonic antigen (CEA) at 1.43 (normal range <5.0) ng/ml, and alpha fetoprotein (AFP) at 3.05 (normal range <9.0) ng/ml. The abdominal non-enhanced CT showed the diffusely swollen pancreas, with massive pleural effusion and thickening of the left anterior renal fascia (Fig. 1). She was diagnosed as acute pancreatitis and conservative medical treatment including inhibition of pancreatic secretion and anti-infection was applied. Her symptoms relieved 7 days later.

Three months later the patient presented again with epigastric pain and fever. Her body temperature was as high as 39°C. On physical examination, an 8-cm well defined mass was touched under her left rib. Abdomen was soft without tenderness, rebound tenderness, and rigidity. Laboratory exam showed white blood cells at $6.87 \times 10^9/L$ (normal range $3.50 \times 10^9/L$ - $9.50 \times 10^9/L$), neutrophile granulocyte proportion at 88.3% (normal range 40.0%-75.0%), albumin at 34.3 (normal range 35.0-55.0) g/L, globulin at 45.9 (normal range 15.0-40.0) g/L, total bilirubin at 15.0 (normal range 3.4-25.0) $\mu\text{mol/L}$, directly bilirubin at 4.9 (normal range 0-7.0) $\mu\text{mol/L}$; serum amylase at 54 (normal range 0-115) U/L, serum lipase at 33.9 (normal range 0-60.0) U/L; CA19-9 at 9.4 (normal range <35) U/ml, CA125 at 87.22 (normal range <35) U/ml, CEA at 1.26 (normal range <5.0) ng/ml, and AFP at 2.74 (normal range <9.0) ng/ml. Abdominal enhanced CT indicated the swelling of the pancreas and more pleural effusion than ever before, and thickening of the left anterior renal fascia (Fig. 2). A diagnosis of acute pancreatitis was made. She was managed with inhibition of pancreatic secretion and anti-infection treatment. After 7 days, her epigastric pain

did not relieve and the mass did not show reduction in the size.

Considering no remission achieved and globulin level elevating, IgG subclass test was performed again, showing IgG1 at 21 200 (normal range 4900-11 400) mg/L, IgG2 at 6080 (normal range 1500-6400) mg/L, IgG3 at 611 (normal range 200-1100) mg/L, and IgG4 at 4610 (normal range 80-1400) mg/L. To definite diagnosis, we recommended an endoscopic ultrasound guided fine needle pancreas biopsy. However, she refused to undergo the invasive test. Based on the above findings, a possible diagnosis of IgG4-related AIP was made.

She started on methylprednisolone 20 mg/d instead of antibiotic therapy. Her epigastric pain relieved a week later, and physical examination showed the volume of the left upper quadrant abdominal mass was reduced. After 1 month, methylprednisolone dosage was tapered by 5 mg every month until the maintenance dose of 5 mg per day. Non-enhanced CT showed that the peripancreatic effusion volume reduced and the size of the pancreas decreased 6 months later (Fig. 3). And IgG4 returned to the normal level 772 (normal range 80-1400) mg/L.

Figure 1. Non-enhanced CT images on admission demonstrated the swollen in the body and tail of the pancreas (white arrow in A) with massive effusion around the pancreas (black arrow in A) and thickening of the left anterior renal fascia (white arrow in B).

Figure 2. Enhanced CT images 3 months after the initial diagnosis demonstrated the enlargement in the body and tail of the pancreas (white arrows in A and B); the enhancement of the body and tail was lower than that of the head during arterial phase (A), and higher than that of the head during portal phase (B). More effusion around the pancreas (black arrows in A and B) was observed than 3 months before. There still existed thickening of the left anterior renal fascia during arterial phase (white arrow in C), with more effusion around the left kidney (black arrow in C) than 3 months before.

Figure 3. Non-enhanced CT imaging half a year after taking methylprednisolone demonstrated the swelling in the body and tail of the pancreas (white arrow in A), which was smaller than that half a year before. The effusion around the pancreas (black arrow in A) also decreased. It still showed thickening of the left anterior renal fascia (white arrow in B), while the effusion around the left kidney (black arrow in B) had relieved.

DISCUSSION

AIP is a rare and benign disorder of the pancreas that occurred in individuals over 55 years old. Its characteristic is diffuse or localized enlargement of the pancreas and obstructive jaundice. Two subtypes of AIP have been recognized. Type 1 AIP is a manifestation of IgG4-related disease that often affects extra-pancreatic organs and is rich in IgG4-positive cells. Type 2 AIP is a pancreas-specific disorder that is lack of IgG4-positive cells.¹⁻²

IgG4-related AIP is a common type of IgG4-related disease that is characterized by an enlarged organ, elevated serum levels of IgG4 and tissue infiltration of IgG4+ plasma cells accompanied by tissue fibrosis and sclerosis.³ The radiological features of IgG4-related AIP is the diffuse or focal pancreas enlargement, sometimes accompanied by mass, and irregular narrowing of the main pancreatic duct, usually associated with bile duct stricture.⁴ It is often misdiagnosed as pancreatic cancer.

The CT imaging of this patient showed the massive effusion around the pancreas, with renal fascia thickening, making it seem more like acute pancreatitis rather than pancreatic cancer; meanwhile, her initial symptom was abdominal pain with normal CA19-9 level. Therefore, she was initially diagnosed as acute pancreatitis. But the symptoms aggravated during treatment; meanwhile, serum amylase and lipase did not elevate during hospitalization, suggesting that the initial diagnose might be inaccurate. Considering globulin level elevating, a diagnosis of AIP was established. IgG subclass test showed the markedly increased serum level of IgG4. Therefore, the experimental treatment of IgG4-related AIP was started at 20 mg methylprednisolone for safety, instead of the recommended dose 0.6-0.8 mg/kg, which alleviated her symptoms,

reduced effusion around the pancreas and declined IgG4 level. A final diagnosis of IgG4-related AIP was made although histopathological examination was not performed.⁵

Glucocorticoids are preferred to treat IgG4-related AIP. International Consensus Guidance Statement on the management and treatment of IgG4-related disease⁶ recommends that the initial dose of glucocorticoids is prednisone 30-40 mg per day, and the dose was adjusted based on body weight or the severity of illness. The initial dose should be maintained 2-4 weeks, gradually reducing to a minimum amount or withdrawal.⁶ Patients may have the risk of recurrence as dosage reduction or drug withdrawal. It is recommended that IgG4-related AIP patients should maintain to take low dose of glucocorticoids for several years.⁷ The patient's symptoms gradually disappeared, the size of the pancreas decreased on CT images and serum IgG4 level go back to the normal range after glucocorticoid therapy, suggesting that the treatment was effective. Further, the patient need to be treated with small dose of glucocorticoid and regularly reviewed.

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