

Rapid deterioration of diabetes mellitus with morphological changes to the pancreas

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DESCRIPTION

An 82-year-old man was referred to the hospital for further examination of a pancreatic tumour diagnosed on abdominal ultrasonography. He underwent regular comprehensive medical check-up, and a pancreatic tumour was detected by chance. Laboratory findings revealed the following: amylase level, 125 IU/L; blood sugar level, 114 mg/dL; HbA1c level, 5.9%; and IgG4 level, 377 mg/dL. CT showed multiple low-density masses on his pancreas (**figure 1A**). Five months after the first CT, he presented with thirst and polyuria and exhibited a weight loss of 10%. Laboratory findings revealed the following: lipase level, 23 IU/L; amylase level, 66 IU/L; blood sugar level, 439 mg/dL; HbA1c level, 10.0%; and IgG4 level, 541 mg/dL. Endogenous insulin secretion was suppressed, as the serum fasting C-peptide level was 0.60 ng/mL and the 24-hour urinary C-peptide level was 9 µg. A second CT showed diffuse enlargement of the pancreas and a capsule-like rim (**figure 1B**). Endoscopic ultrasound (EUS)-guided fine-needle aspiration (FNA) showed slight lymphoplasmacytic infiltration, but did not show storiform fibrosis, obliterative phlebitis or nuclear atypia. IgG and IgG4 staining results were too intense to assess. He was diagnosed with IgG4-related autoimmune pancreatitis (AIP) because of high IgG4 level and diffuse enlargement of the pancreas. He received prednisolone (PSL) 30 mg (0.5 mg/kg) and insulin therapy. Six months later, IgG4 was decreased to 210 mg/dL, and pancreatic size was reduced (**figure 2**). PSL was tapered to 5.0 mg and HbA1c decreased to 6.6%. Insulin therapy was switched to an oral hypoglycaemic agent.

IgG4-related AIP is one of the clinical manifestations of IgG4-related disease. The incidence of IgG4-related AIP is 0.28–1.08 per 10 000 in Japan.¹ The male:female ratio is 1:0.77.² Chronic exposure to occupational antigens could play an important role in IgG4-related diseases. In a Dutch study of 25 patients with IgG4-associated cholangitis and/



Figure 2 Reduction of pancreatic size.

Learning points

- ▶ Morphological evaluation plays a key role in diagnosing IgG4-related autoimmune pancreatitis (AIP).
- ▶ Focal-type AIP could develop into diffuse-type AIP with clinical manifestations.
- ▶ Careful morphological follow-up of AIP is needed.

or IgG4-related disease, 88% had a history of blue-collar work of at least 1 year.³ The differential diagnosis is pancreatic cancer. In the case of pancreatic tumour, ruling out the diagnosis of AIP is important because AIP is highly responsive to PSL therapy; thus, surgery could be avoided. Only 43% of cases were diagnosed with AIP based on histologic examination of samples obtained during EUS-FNA.⁴ Morphological evaluation plays a key role in the diagnosis. One study showed that among 87 AIP patients, 63% of them displayed focal type, while 37% showed diffuse type.⁵ A morphological change from focal to diffuse type is rare. However, the focal type could develop into diffuse type with clinical manifestations.⁶ Therefore, careful morphological follow-up is needed.

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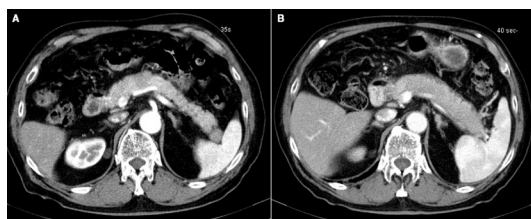


Figure 1 (A) Multiple low-density masses on his pancreas. (B) Diffuse enlargement of the pancreas and a capsule-like rim.

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