IgG4-related autoimmune pancreatitis complicated by splenic artery pseudoaneurysm

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DESCRIPTION

A 65-year-old man presented with a 1-year history of swelling of the submandibular salivary glands bilaterally. Blood tests revealed C-reactive protein (CRP) level of 0.28 mg/L, leucocyte count of 5.22×10^9/L (neutrophil count 3.07×10^9/L), haemoglobin level of 13.1 g/dL and platelet count of 234×10^9/L. Serum IgG and IgG4 levels were 2054 mg/dL (normal 861–1747) and 540 mg/dL (4.8–105), respectively. Biopsy of the right submandibular gland revealed a dense lymphoplasmacytic infiltrate and storiform fibrosis with increased IgG4-positive plasma cells (IgG4:IgG ratio 67%). Contrast-enhanced CT (CE-CT), which was performed to evaluate other sites of involvement, showed diffuse enlargement of the pancreas with a capsule-like rim (figure 1A). A diagnosis of IgG4-related sialadenitis and autoimmune pancreatitis was made. The patient had no abdominal complaints or jaundice, and he was followed up with careful observation.

One year later, follow-up CE-CT identified a newly formed aneurysm (11 mm in diameter) of the splenic artery (figure 1B). No other vascular abnormalities were identified on CE-CT. Laboratory tests showed serum CRP of 0.21 mg/L and amylase of 38 U/L (normal 44–132). Liver function tests were normal. Serum IgG4 level was unchanged at 472 mg/dL. We considered that the newly formed aneurysm was likely to be a pseudoaneurysm due to autoimmune pancreatitis. Coil embolisation was then performed to prevent rupture of this pseudoaneurysm, and the patient was started on prednisolone 70 mg/day (1 mg/kg) to control the activity of autoimmune pancreatitis. He responded favourably to the corticosteroid treatment, and 1 week later the pancreatic enlargement showed some reduction in size on CE-CT. The patient received 70 mg/day of prednisolone for 1 week, followed by 40 mg/day for 2 weeks, 30 mg/day for 3 weeks, and thereafter the drug was gradually tapered.

Arterial pseudoaneurysm complicating acute or chronic pancreatitis is well recognised, and the splenic artery is the most commonly affected vessel.[1] However, as far as we can establish from a search of the literature, there are no published reports of autoimmune pancreatitis with associated splenic artery pseudoaneurysm. Thus, splenic artery pseudoaneurysm should be recognised as a potentially fatal complication of autoimmune pancreatitis. CE-CT could be the imaging modality of choice in the evaluation of abnormalities of splenic vasculature in this disease.

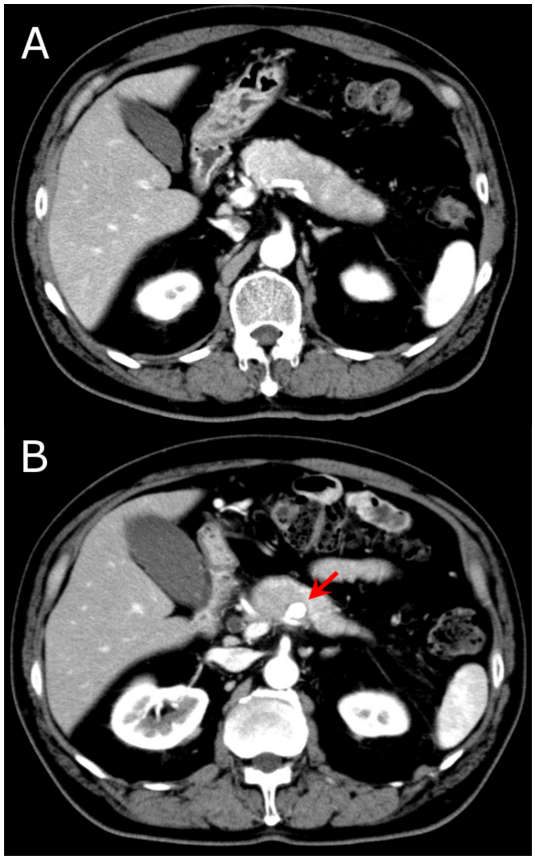

Figure 1 Contrast-enhanced CT on presentation (A) and after 1 year of observation (B) showing diffuse enlargement of the pancreas. A splenic artery pseudoaneurysm (red arrow, 11 mm in diameter) was newly formed at 1-year follow-up (B).

Patient's perspective

"I am relieved that the aneurysm was identified before rupture. I appreciate my doctor's appropriate treatment."

Learning points

- Splenic artery pseudoaneurysm should be recognised as a potentially fatal complication of autoimmune pancreatitis.
- Contrast-enhanced CT could be the imaging modality of choice in the evaluation of abnormalities of splenic vasculature in autoimmune pancreatitis.

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