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 234×10⁹/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

B

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).

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. 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.

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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