

Answer: Diffuse enlargement of the pancreas (“sausage pancreas”) is uniquely associated with autoimmune pancreatitis. While A, C, and D are also associated with autoimmune pancreatitis, they can be present in other disease processes. Pancreatic cancer can be associated with elevated IgG4 levels and pancreatic masses, and a variety of biliary diseases are associated with biliary strictures.

Teaching points:

Autoimmune pancreatitis (AIP) is characterized clinically by presentation with obstructive jaundice with or without a pancreatic mass, histologically by a lymphoplasmacytic infiltrate and fibrosis and therapeutically by a dramatic response to steroids. Typical imaging findings for AIP include diffuse pancreatic enlargement (“sausage pancreas”). AIP is divided into Type 1 (IgG4-related) and Type 2, which differ in histology and clinical course. A definite diagnosis can be made using histology, imaging, or steroid response as primary basis for diagnosis and various combinations of collateral evidence.¹ While AIP is an uncommon cause of pancreatitis, it should be considered in patients without another explanation for pancreatitis who have typical imaging findings.

Table 1: ICDC Level 1 diagnostic criteria for Type 1 AIP¹

Criteria	Finding
Parenchymal imaging	Diffuse enlargement with rim-like enhancement
Ductal imaging	Long or multiple strictures without marked dilation
Serology	IgG4 level >2x upper limit of normal
Pancreatic histology	Lymphoplasmacytic infiltrate, fibrosis
Response to steroid therapy	Rapid radiologically demonstrable improvement in pancreatic manifestations

References

1. Shimosegawa T, Chari ST, Frulloni L, et al. International consensus diagnostic criteria for autoimmune pancreatitis: guidelines of the International Association of Pancreatology. *Pancreas*. 2011;40(3):352-358. doi:10.2958/suizo.26.684.