Autoimmune pancreatitis with multifocal lesions

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Abstract
Two cases of a focal type of autoimmune pancreatitis (AIP) with distinct double mass lesions within the pancreas are described. In both patients, computed tomography (CT) showed localized pancreatic masses with delayed enhancement, and magnetic resonance cholangiopancreatography (MRCP) revealed localized stenoses of the main pancreatic duct (MPD) with mild upstream dilatation. Fluorodeoxyglucose positron emission tomography (FDG-PET) examination, performed in one patient, showed intense uptake concordant with tumors. Both patients received pancreatic resection with a presumptive diagnosis of pancreatic carcinoma. Histologic evaluation of the tumors showed marked lymphoplasmacytic infiltration and fibrosis around the large and medium pancreatic ducts, without any evidence of malignancy. Serum IgG4 concentration, measured postoperatively, was elevated in both patients. The characteristic morphological features of AIP are diffuse swelling of the pancreatic parenchyma and diffuse narrowing of the MPD. Recently, a focal type of AIP, which mimics pancreatic carcinoma, has been recognized. Considering the favorable response of AIP to steroid therapy, it is clinically important to differentiate the focal type of AIP from pancreatic carcinoma and to know that AIP sometimes exhibits multiple lesions within the pancreas.

Key words Autoimmune pancreatitis · Multifocal lesions · IgG4 staining

Introduction
Autoimmune pancreatitis (AIP) is a unique form of chronic pancreatitis associated with an autoimmune inflammatory process. Although diffuse swelling of the pancreatic parenchyma and diffuse irregular narrowing of the pancreatic duct system are morphologically characteristic of AIP, a focal type of this clinical entity has been recently recognized.

The focal type of AIP exhibits a localized mass lesion in the pancreas, similar to pancreatic carcinoma, and it often exhibits obstructive jaundice, which is also characteristic of pancreatic carcinoma, when the lesion involves the head of the pancreas. Consequently, some patients with these features have been subjected to surgical exploration with a presumed diagnosis of pancreatic carcinoma. Considering that AIP shows a favorable response to steroid therapy, the differentiation of these two entities is clinically important.

Although patients with AIP sometimes show multifocal or skipped narrowing of the main pancreatic duct (MPD), there have been only a few cases of AIP with multifocal lesions. In this report, we describe the clinical, radiological, and histopathological features of two patients with AIP who exhibited distinct double masses in the pancreas; the masses were resected on the suspicion of pancreatic carcinoma.

Case reports

Case 1
A 62-year-old male patient with mild epigastralgia was referred for further investigation of pancreatic masses. He had no medical history of autoimmune disease, but he had a history of hypertension. The results of laboratory examinations, including complete blood count, electrolytes, bilirubin, liver function tests, and pancreatic enzymes and tumor markers (carbohydrate antigen [CA] 19-9, and carcinoembryonic antigen [CEA]), were all within normal limits. Computed tomography (CT) showed irregular mass lesions in the head and body of the pancreas (Fig. 1a, b); the lesions were 30mm and 25mm in diameter, respectively. The tumors showed slight attenuation in the delayed phase with contrast
medium. Fluorodeoxyglucose positron emission tomography (FDG-PET) showed intense uptake in both lesions, and their standardized uptake values (SUVs) were 6.6 and 8.3, respectively (Fig. 1c, d). Magnetic resonance cholangiopancreatography (MRCP) revealed skipped stenoses of the MPD, concordant with the tumors, and mild dilatation between the tumors and distally (Fig. 1e).

**Case 2**

A 64-year-old male patient without any symptoms or past medical history was admitted because of pancreatic masses that were picked up on a medical checkup. Laboratory tests showed slight elevation of blood glucose (126 mg/dl; normal, 69–104 mg/dl) and hepatic enzymes (aspartate aminotransferase, 41 IU/l [normal, 13–33 IU/l], alkaline aminotransferase, 75 IU/l [normal, 8–42 IU/l]). Dynamic CT showed two lesions, in the body and tail of the pancreas, 28 mm and 30 mm in diameter, respectively, and exhibited subtle delayed enhancement (Fig. 2a). MRCP revealed obstruction of the MPD in concordance with the tumors, and slight dilatation between the tumors (Fig. 2b).

The proximal lesions in both patients were considered to be pancreatic carcinomas because the finding of localized stenoses with upstream dilatation of the MPD was suggestive of pancreatic carcinoma. The distal lesions were deemed to be either obstructive pancreatitis demonstrating mass lesions because of severe inflammation, or other primary pancreatic carcinoma. In case 1, core needle biopsy of the tumor in the pancreatic body was performed during surgery, and this revealed parenchymal fibrosis and infiltration of inflammatory cells, including plasma cells, without any evidence of malignancy. Therefore, Whipple resection was performed to resect only the head lesion. In case 2, the patient received distal pancreatectomy.

The three resected tumors (case 1, head; case 2, body and tail) resembled each other on both macroscopic and microscopic examinations. On gross appearance, all cut surfaces of the tumors demonstrated swelling of the parenchyma, but the border between the tumor and the surrounding pancreatic tissue was unclear, while the existing lobular structure and narrowed MPD were clearly identified (Fig. 2c). Microscopic examination confirmed marked lymphoplasmacytic infiltration and fibrosis around the large and medium pancreatic ducts.

**Fig. 1a–e.** Case 1. Computed tomography (CT) revealed irregular mass lesions in the head (a) and body (b) of the pancreas (arrowheads); early phase. c, d Fluorodeoxyglucose positron emission tomography (FDG-PET) showed intense uptake in both lesions (arrows), and their standardized uptake values (SUVs) were 6.6 and 8.3, respectively. (c, head; d, body). e Magnetic resonance cholangiopancreatography (MRCP) revealed skipped stenoses in the main pancreatic duct (MPD) concordant with the tumors (head, arrows; body, arrowheads), and mild dilatation between the tumors and distally.