Clinicopathologic Differentiation of Atrophy of the Pancreatic Body and Tail Aplasia

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Summary

Conclusion. Congenital aplasia of the body and tail of the pancreas is derived from a defect of the dorsal pancreatic anlage and should not be considered a type of acquired atrophy of these structures.

Background. Congenital aplasia of the body and tail of the pancreas radiologically mimics acquired atrophy of the pancreatic body and tail.

Methods. Two patients with radiologically identified aplasia of the body and tail of the pancreas were studied clinicopathologically.

Results. An 82-yr-old man was diagnosed radiologically as having both carcinoma of the head of the pancreas and aplasia of the body and tail of the pancreas and underwent pancreatoduodenectomy. Pathologically the carcinoma was distributed in the anterosuperior part of the head of the pancreas, and spread into the duct of Santorini and intraductally to a portion of the main pancreatic duct beyond the junction of the ducts of Santorini and Wirsung. Consequently, obstructive pancreatitis of the body and tail of the pancreas developed, resulting in marked atrophy that mimicked aplasia of the body and tail of the pancreas. A 74-yr-old woman was diagnosed radiologically as having two carcinomas, one of the gallbladder and one of the stomach, and aplasia of the body and tail of the pancreas. During surgery, suspected parenchymal disappearance and fatty replacement in the body and tail of the pancreas were noted. Histologic examination of biopsy specimens from the body portion revealed atrophic pancreatic tissue surrounded by fat. Therefore, these patients had atrophy of the pancreatic body and tail.

Key Words: Pancreas body aplasia; pancreas tail aplasia; pancreatic body atrophy; pancreatic tail atrophy; ventral pancreatic polypeptide (PP)-positive cells; dorsal PP-positive cells.

Introduction

Congenital aplasia of the body and tail of the pancreas is an extremely rare anomaly. Heiberg (1), who, in 1911, described its occurrence in a 72-yr-old man, may have been the first to observe aplasia of the dorsal pancreatic anlage. Ghon and Roman (2) reported the case of a 14-yr-old boy in whom the head of the pancreas was flat and disk shaped, but neither the body and tail of the pancreas nor the minor papilla was observed. Case reports in German (3–6) and in Japanese (7) followed, and the report in by Lechner and Read (8) in the English was the first

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Fig. 1. Enhanced CT scan of case 1 showing multifocal cystic lesions in the pancreatic head. No pancreatic parenchyma is evident on the anterior side of the splenic vein.

description of this anomaly in a living patient. According to our previous study (9), after fusion of the ventral and dorsal anlagen, the ventral pancreas is considered to form the posteroinferior part of the head of the pancreas, whereas the dorsal pancreas consists of the remaining anterosuperior part of the head together with the body and tail of the pancreas. Therefore, the term “aplasia of the body and tail of the pancreas” should be reserved for such conditions as those reported by Ghon and Roman (2), derived embryologically from a defect of the dorsal pancreatic anlage.

Recently, however, increasing numbers of patients with such anomalies have been detected incidentally when carrying out diagnostic imaging, including endoscopic pancreatography (10). In such patients, radiology revealed the duct of Santorini with an opening to the minor papilla, because aplasia of the body and tail of the pancreas mimicked radiologically acquired atrophy or fatty replacement of the pancreatic body and tail.

We carried out a histopathologic study to differentiate acquired atrophy of the pancreatic body and tail and congenital aplasia of the body and tail of the pancreas. Pancreatic tissues from two patients with atrophy of the pancreatic body and tail were examined, and the radiologic findings indicated that they had aplasia of the body and tail of the pancreas.

**Patients and Methods**

The two patients in our study are described in detail in the following. The pancreatic tissues were fixed with 10% formaldehyde solution, dehydrated with a graded alcohol series, and embedded in paraffin. Histologic sections were stained with hematoxylin and eosin (H&E) and immunostained with antipancreatic polypeptide ([PP]; UCB, Belgium) and anti-insulin (DAKO, Japan) antibodies. The pancreatic head tissue was mapped for PP-positive cells.

**Case 1**

An 82-yr-old man was admitted to Yamanashi Medical University Hospital on February 16, 1991, with jaundice, anorexia, and right hypochondralgia. He was diagnosed radiologically as having both a car-