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Original contribution

Heterotopic pancreas: a clinicopathological study of 184 cases from a single high-volume medical center in China☆,☆☆

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Summary Heterotopic pancreas (HP) is often an incidental finding during operative or endoscopic procedures and described in case reports and small series in the literature. Large clinicopathological studies with a systematic analysis remain lacking. Herein, we identified 184 (0.18%; 184/99 544) consecutive histology-proven HP cases of 89 770 surgical resections and 9774 upper endoscopic biopsies carried out at a single medical center in China. Each case was diagnosed by unequivocal identification of pancreatic acini at a location outside the pancreas. The patients' median age was 49 years (range, 14-82) with a slight female predominance (male/female ratio, 0.94). Clinical presentation at diagnosis was nonspecific. Preoperatively, most (54.9%; 101/184) HP lesions were misdiagnosed. Only 26 HP lesions (14.1%) were correctly diagnosed, all in the stomach except for 1 in the duodenum; 57 (31%) were detected during operative procedures for other conditions. The most frequent location of HP was, in descending order, the stomach (97; 52.7%), small intestine (48; 26%), lesser and greater curvature omentum (18; 10%), spleen and hilar region (5; 2.7%), porta hepatis (2; 1%), gallbladder (1; 0.5%), peridistal esophageal tissue (4; 2.2%), and mesentery (7; 3.8%). The size of HP varied from smaller than 0.5 cm (35.3%), 0.6 to 1 cm (34.8%), to larger than 1.1 cm (29.9%). Because of difficulty in preoperative diagnosis, careful workup for upper gastrointestinal diseases with HP as a differential diagnosis may increase the chance of accurate diagnosis and appropriate patient management.

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1. Introduction

Heterotopic pancreas (*HP*), also known as aberrant or accessory pancreas, is defined as normal pancreatic tissue located in abnormal anatomical sites without any anatomical or vascular connection with the normal pancreas [1]. HP is usually found in the upper gastrointestinal tract, more in male than in female individuals [2], and described in the literature primarily as case reports or in small series [3–5]. Large clinicopathological studies remain lacking. Although the detailed clinical and epidemiological understanding of this abnormality is unknown because of the lack of systemic studies, the incidence of HP is believed to be low, ranging from 0.55% to 13.7% at autopsy [6], and found in approximately 0.2% (1/500) of upper abdominal operations [1]. Preoperative diagnosis of HP is difficult because of its small size. In most cases, it is reported as an incidental histology finding. According to published studies, most HP abnormalities are found in the duodenum (30%); stomach (25%); jejunum (15%); Meckel diverticulum (6%); and, rarely, ileum (3%) [6]. Its location outside the gastrointestinal tract, such as lymph node, mesenteric tissue, omentum, spleen, and others, has not been previously reported.

Because of its rarity and nonspecific clinical presentation, accurate preoperative diagnosis of HP can be challenging. Herein, we retrospectively studied clinicopathological characteristics of 184 consecutive patients with histologically confirmed HP over the past 12 years at a single high-volume medical center in Nanjing, China, to provide more detailed information on HP of this rare congenital anomaly.

2. Materials and methods

We searched electronic pathology reports stored in the Department of Pathology of the Nanjing Drum Tower Hospital in Nanjing, China, for the final pathologic diagnosis of HP. The cases without archival tissue blocks available for recuts were excluded. A total of 89 770 surgical resections over the period from January 2004 to February 2012 and 9774 upper endoscopic biopsy cases for the period from January 2009 to January 2015 were reviewed, among which 184 (126 surgical resections and 58 endoscopic biopsies) HP cases were identified and included for the analysis. The type of resections included partial or total gastrectomy (n = 85), partial hepatectomy with nodal dissection (n = 8), the Whipple procedure (n = 15), partial resection of small (n = 8) or large (n = 5) intestine, splenectomy (n = 3), and distal esophagectomy (n = 2). Each pathologic report along with hematoxylin and eosin–stained slides was reviewed to confirm the diagnosis of HP.

By histology, HP consisted of 3 components: (1) characteristic pancreatic acini with a single row of pyramidal exocrine epithelial cells featuring dense dark-purplish appearances in the basal cytoplasm in contrast to the bright orange-red granular apical cytoplasm; (2) a ductal component, lined by flat columnar epithelial cells, frequently showing dilated, microcystic changes; and (3) an endocrine element of an islet of Langerhans. Depending upon the availability of target tissue, not all 3 components were identified in a single case, but the presence of pancreatic exocrine acini was required for the HP diagnosis in all cases. Any cases with only pancreatic acinar metaplasia in gastric mucosa were excluded.

The corresponding patient medical records were retrieved and reviewed for demographic information, clinical presentation, diagnostic procedure, preoperative diagnosis, and therapeutic intervention. The study protocol was approved by the Medical Ethics Committee of the Nanjing Drum Tower Hospital in China.

Average and standard deviation (SD) were calculated with Microsoft Excel.

3. Results

Among 99 544 cases reviewed over the study period, HP was identified in 184 patients (0.18%). Most patients were middle aged with an average age of 49 years (range, 14-82). HP lesions were near equally distributed in both sexes with a slight female predominance (Fig. 1 and Table). No multifocal HP lesions were observed in the same case.

Although primarily identified in the stomach (52.7%; Table), HP was surprisingly widely distributed in the upper abdomen with approximately 14% in the perigastric fat as small lymph node–like nodules; 25% in the duodenum and proximal jejunum surrounding the pancreatic head and body; and occasional cases in the ileum, paradistal esophagus, perigallbladder tissue, splenic hilar region, and spleen (Fig. 2). Three instances were incidentally found in Meckel diverticula of the ileum. Most HP lesions were small in size, varying from 0.1 to 3 cm. The average size was 0.84 cm (Table). The percentages

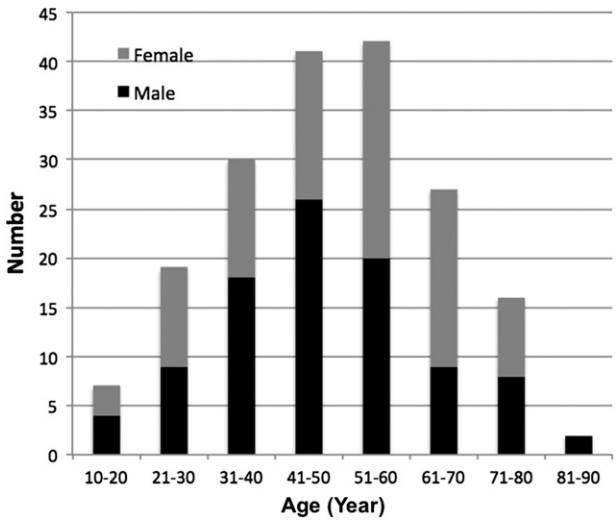

Fig. 1 Age and sex distribution of patients with HP.