

Clinical Images

Is an accessory nipple associated with an intrapancreatic accessory spleen?

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Intrapancreatic accessory spleen (IPAS) is a rare differential diagnosis of a solitary hypervascular lesion. It is not uncommon to misdiagnose IPAS as pancreatic tumors since no specific imaging has been developed for the differential diagnosis. We described a rare case of a male patient with a unilateral accessory nipple and a solitary hypervascular lesion in the pancreas. The patient underwent a distal pancreaticosplenectomy, and IPAS was finally diagnosed.

A previously healthy 41-year-old man presented melena in the absence of nausea, hematemesis, dizziness, diarrhea, abdominal pain, or distention. He was admitted to a local hospital and the melena disappeared after a 2-week treatment (e.g., proton pump inhibitor and hemostatic drugs). For further diagnosis and treatment, he was admitted to our hospital. The physical examination revealed nothing abnormal except for a third nipple (Fig. 1A). The blood routine and biochemistry parameters showed that liver and kidney function and electrolyte and metabolic balance were all normal. The blood tumor biomarkers were negative. Gastroscopy showed a 2-cm gastric ulcer at the healing stage. Enhanced computed tomography (CT) and magnetic resonance imaging (MRI) showed a well blood-supplied solid mass measuring 2.2×1.8 cm in the tail of pancreas (Fig. 1B).

A multidisciplinary team, including a surgeon, a radiologist, an endocrinologist, and a gastroenterologist, suspected pancreatic neuroendocrine neoplasm (PNEN). To confirm the diagnosis, a set of tests, such as serum hormone levels (e.g., gastrin, parathyroid, insulin, and C-peptide), were performed. No positive result was observed from the glucose tolerance test or the serum hormone measurement. Thereafter, a biopsy was performed; the punctured tissue was a fibrinous exudate with a few endocrine-like cells, as confirmed by two independent pathologists. The treatment of non-functioning PNEN remains controversial. Our

colleagues have shown a promising outcome after surgical treatment in patients with non-functioning PNEN [1]. Therefore, the patient decided to resect the pancreatic mass the day after he was informed. First, we did a laparoscopic exploration and found enlargement of the tail of the pancreas. Then, we performed a distal pancreaticosplenectomy, and an accessory spleen was finally diagnosed by a pathologist (Fig. 1C). No complications (e.g., pancreatic leakage or sepsis) occurred after the operation. He recovered well, and was discharged on postoperative day 7. The patient was arranged for routine follow-up in the outpatient clinic (e.g., platelet, ultrasonic imaging for spleen vein thrombosis) for distal pancreaticosplenectomy.

IPAS is a rare differential diagnosis of a solitary hypervascular lesion, such as PNEN. In the majority of cases, the lesions are relatively small without clinical symptoms and are found incidentally. It is not uncommon to misdiagnose IPAS as PNEN [2–4] because even diffusion-weighted MRI does not well differentiate IPAS from a small hypervascular PNEN [4]. No specific imaging is available for the differential diagnosis. Therefore, multiple techniques, including ⁶⁸Ga-DOTA-TATE PET/CT, ^{99m}Tc technetium heat-damaged red blood cell/colloid scintigraphy, and endoscopic ultrasonography-fine needle aspiration, have been recommended [5,6]. Nevertheless, IPAS is still frequently misdiagnosed as PNEN, leading to unnecessary surgery [5].

This is a rare case of a male patient with a unilateral accessory nipple and IPAS. An accessory nipple and IPAS are both congenital anomalies, possibly with specific genomic profiles. An accessory nipple is much easier to diagnose than IPAS [7]. It is noteworthy that an accessory nipple can be found in a number of syndromes, and it is associated with other diseases such as mastitis, cysts, cardiac arrhythmias, pyloric stenosis, epilepsy, and renal malformations [7,8]. Although an accessory nipple is a cosmetic blemish

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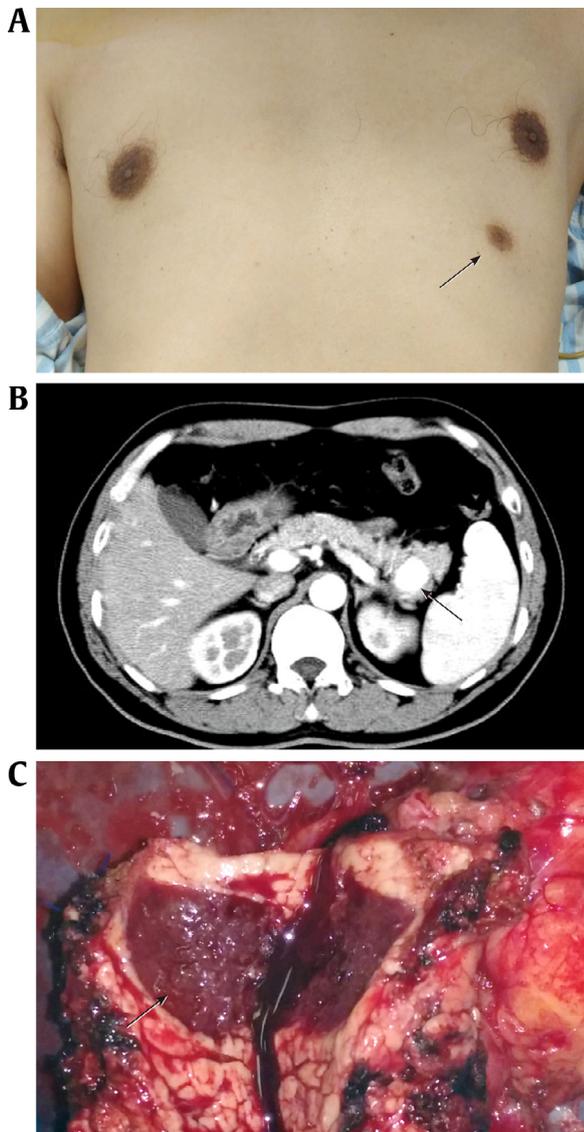


Fig. 1. The physical examination revealed nothing abnormal except for a third nipple (A, black arrow). CT/MRI showed a well blood-supplied solid mass measuring 2.2×1.8 cm in the tail of pancreas (B, black arrow), which was finally diagnosed as an accessory spleen by the surgical dissection and pathology (C, black arrow).

with limited medical significance, it may indicate a congenital anomaly in the body, and it could be associated with other inherited diseases. Therefore, the case presented here indicates that IPAS should be considered in an individual with accessory organs if he or she is diagnosed with a solitary hypervascular lesion in the pancreas. Genetics may further explain the finding from the current case and provide more evidence.

Contributors

LQ proposed the study. HHT and LQ performed the research and wrote the first draft. Both authors contributed to the design and interpretation of the study and to further drafts. LQ is the guarantor.

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Ethical approval

This study was approved by the Ethics Committee of the First Affiliated Hospital of Zhejiang University School of Medicine (No. 2013-0022). The patient agreed on the publication of his medical history and signed an informed consent.

Competing interest

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

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