Clinical Case Report

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A newborn patient with both annular pancreas and Meckel's diverticulum

A case report of an unusual association

Peng Cai, MD, Zhenwei Zhu, MD, Jianlei Chen, MD, Lulu Chen, MD, Jiang Pan, MD, Wenxian Zhi, MD, Jie Zhu, MD, Bin Wu, MBBS, Zhicheng Gu, MD, PhD, Shungen Huang, MD, PhD, Jian Wang, MD, PhD*

Abstract

Rationale: Annular pancreas (AP) is recognized as a cause of duodenal obstruction in children, while children with Meckel's diverticulum (MD) are usually asymptomatic. Here we present a rare case with both AP and MD, which was identified by abdominal exploration during diamond-shaped duodenoduodenostomy.

Patient concerns: A "double-bubble" sign was found by ultrasound at 35 week of pregnancy. After 39 weeks of pregnancy, the male patient was transferred to the Department of General Surgery, Children's Hospital of Soochow University because of a suspected duodenal stenosis.

Diagnoses: Preoperative abdominal X-ray examination indicated "double-bubble" sign. AP was confirmed by exploratory surgery, with an MD located 30 cm above the ileocecal valve.

Interventions: Diamond-shaped duodenoduodenostomy and a wedge resection of the intestine with end-to-end anastomosis were performed

Outcomes: The patient recovered and his appetite was good without vomiting.

Lessons: Our experience demonstrates that abdominal exploration is essential for children with gastrointestinal malformations.

Abbreviations: AP = annular pancreas, MD = Meckel's diverticulum.

Keywords: abdominal exploration, annular pancreas, congenital anomalies, Meckel's diverticulum

1. Introduction

Annular pancreas (AP) is a rare congenital anomaly with duodenal obstruction, which was first described by Tiedemann in 1818 and was termed "annular pancreas" by Ecker in 1862.[1] Patients most commonly present in infancy or early childhood because the second part of the duodenum is surrounded by pancreas parenchyma.[2] It is usually suspected during routine prenatal ultrasonography by the double bubble sign in the fetal abdomen and children are diagnosed and treated after birth. In adults, AP has been detected to be associated with peptic ulceration, duodenal obstruction and pancreatitis.[3] The presence of AP was reported in 3 of 20,000,[4] but with sophisticated diagnostic modalities, some studies showed an incidence of approximately 1 in 1000.[3] In addition to AP, there are other types of congenital gastrointestinal malformations, including Meckel's diverticulum (MD).

MD is one of the most common congenital anomalies of the gastrointestinal tract, with an incidence of 1% to 3%,[5] but in infants under 2 years old, it is occurring in 4% to 6% of the population.[6] Friedrich Meckel established its embryological origin in 1809,[7] which is caused by the incomplete regression of the omphalomesenteric duct during gestational weeks 5 to 7.[8] MD is generally asymptomatic and the complications include inflammation, gastrointestinal bleeding, and intestinal obstruction. Once the symptoms appear, MD usually occurs in children with hemorrhage and intussusception. Meckel's diverticulum can cause intestinal obstruction and inflammation in adults.[9,10] Although AP and MD are congenital intestinal malformations, infants with both AP and MD are rare. Herein we present an infant with both AP and MD.

2. Case report

Both the father and the 25-year-old mother were healthy; they had no known family history and no adverse genetic history of congenital disease or gastrointestinal anomalies. No significant abnormality was noted at the prenatal examination before 30 weeks.

An ultrasound at 35 weeks showed a "double-bubble" sign. After 39 weeks of pregnancy, the male patient weighed 3350 g at birth. Apgar scores were satisfactory. Radiographs at 3 hours of age confirmed air fluid levels in gastral cavity and the first portion

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PC, ZZ, JC, LC contributed equally; ZG, SH and JW contributed equally.

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Department of Pediatric Surgery, Children's Hospital of Soochow University, Suzhou, China.

* Correspondence: Jian Wang, Department of Pediatric Surgery, Children's Hospital of Soochow University, Suzhou, China (e-mail: wangjiansuzhou@163.com)

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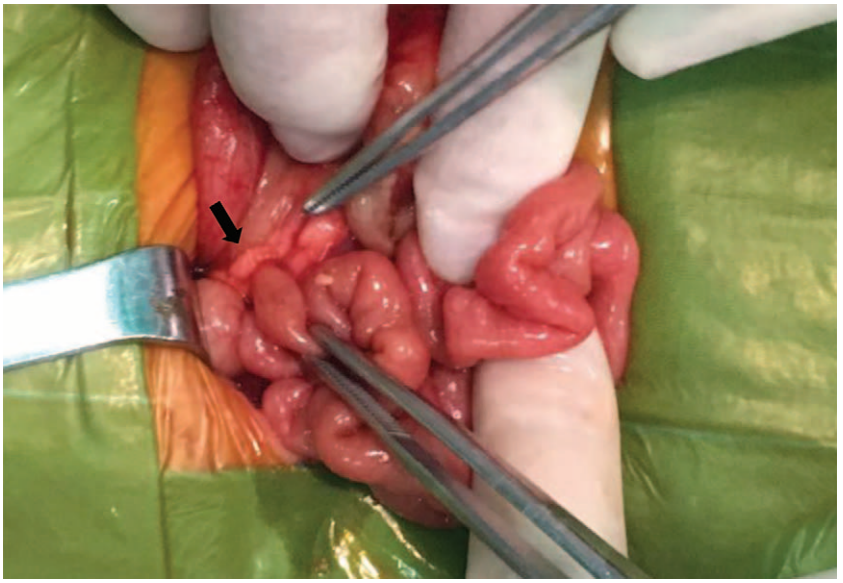

Figure 1. Annular pancreas is marked.

of the duodenum, an absence of air in the distal jejunum. The patient was transferred to the Department of General Surgery, Children's Hospital of Soochow University, as duodenal stenosis was suspected. Six days later, a diagnostic transverse right laparotomy was conducted, which showed AP (Fig. 1). A diamond-shaped duodenoduodenostomy was performed (Fig. 2). Upon intraoperative abdominal exploration, an MD was found located 30 cm above the ileocecal valve in the antimesenteric side (Fig. 3). A wedge resection of the intestine with end-to-end anastomosis was performed (Fig. 4). The pathologist confirmed the clinical diagnosis, revealing no ectopic gastric mucosa in the diverticulum but segmental small bowel (Fig. 5). The child recovered uneventfully, and was discharged 1 week later. The ethical committee approval was not necessary and informed consent was obtained from the patient's parents.

3. Discussion

Stenosis of the duodenum is one of the frequent causes of high neonatal congenital obstruction in infants. Obstruction can be

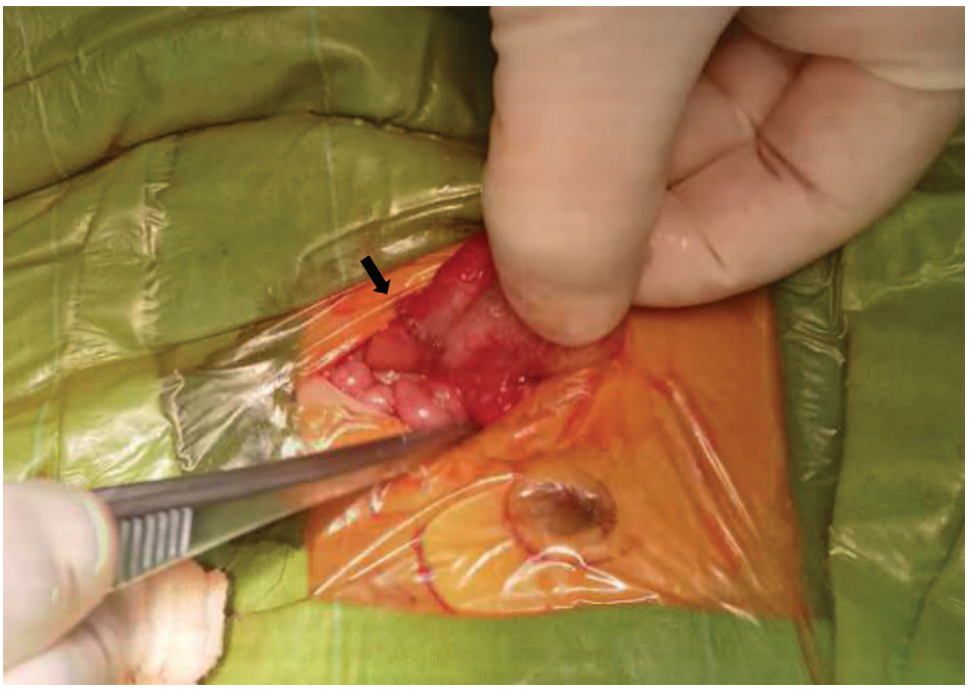

Figure 2. Diamond-shaped duodenoduodenostomy (the point of surgical anastomosis is marked).

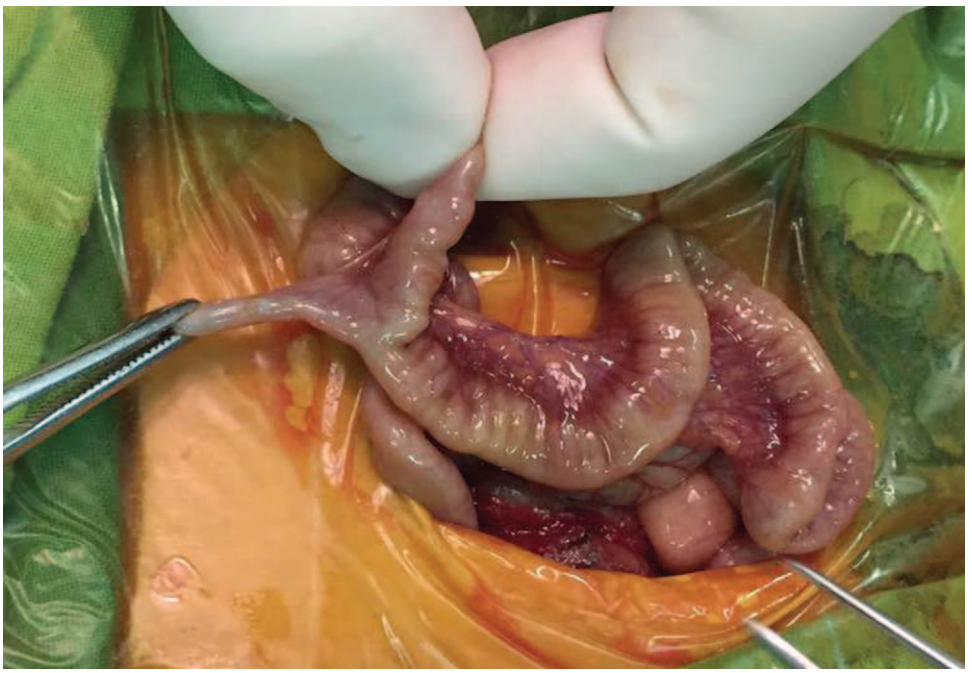

Figure 3. Meckel's diverticulum was found during abdominal exploration.

caused by intraluminal diaphragm. Nearly 40% of the intrinsic duodenal obstruction cases are accompanied by other abnormalities.[11] During weeks 4 to 8 of embryonic development, the dorsal and ventral pancreatic buds merge to form the pancreas during gut rotation. In this process, duodenal stenosis may be associated with pancreatic malformations, as the tip of the ventral pancreas pulled around the right side of the duodenum forming an annulus, defined as annular pancreas. Some studies demonstrated that genes contribute to pancreatic development, for instance, the mutations of FOXF1 and RFX6 might be associated with AP.[12–14] Embryonic development is associated with many factors, not only genes. Factors that extraordinarily modulate the proliferation, apoptosis, cell cycle, and autophagy may lead to abnormal development of the digestive system. Some small molecules, like microRNAs (miRNAs), act as repressors in post-transcriptional control. MiRNA-involved regulatory net-

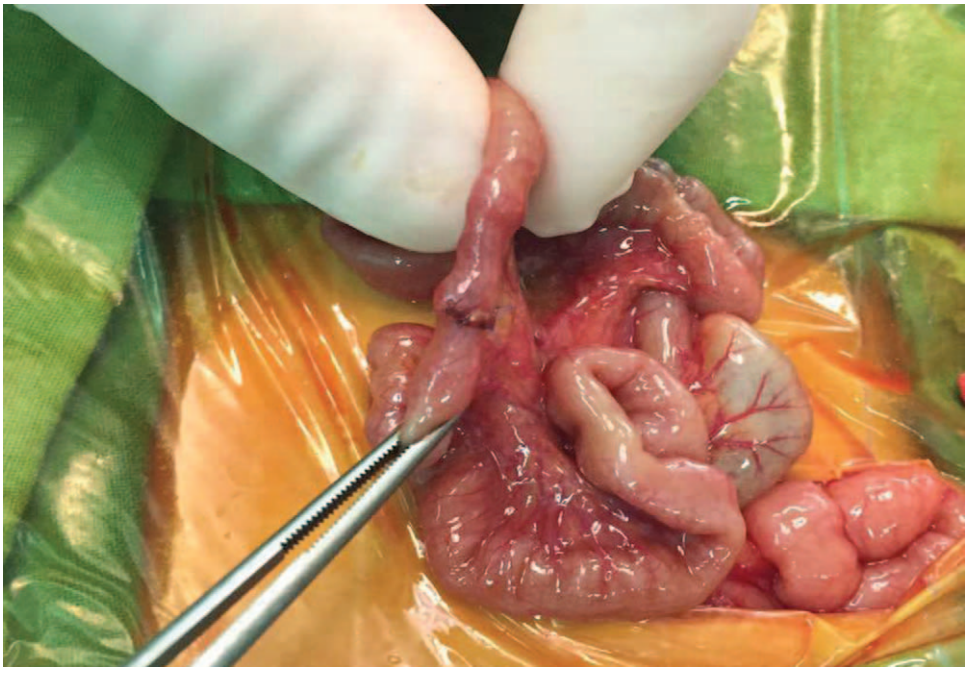

Figure 4. A wedge resection of the intestine with end-to-end anastomosis was performed.

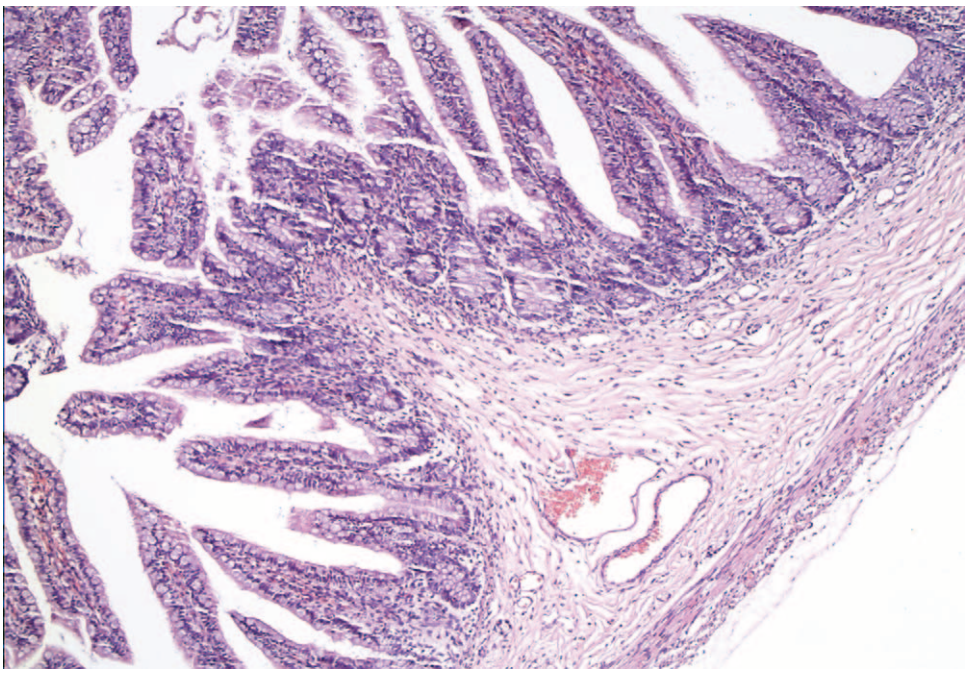

Figure 5. Pathological investigation showed no ectopic gastric mucosa in the diverticulum.

works are involved in various cellular processes. Our previous studies confirmed that miR-24-1*, let-7a*, miR-146a were involved in the pathogenesis of Hirschsprung's disease.[15,16] No investigations clarified the role of miRNAs in the pathogenesis of AP, which may be a focus of future research.

Some other congenital anomalies associated with AP in children are well recognized, with Down's syndrome representing the most common congenital chromosomal anomaly with an incidence of 10% to 30%.[17] Besides, cardiac defects, intestinal atresia, and biliary anomalies were also reported to be associated with AP.[18] Considering the infant mentioned above, although MD is understood to be one of the commonest congenital malformations, little is reported about the potential relation between AP and MD, as well as infants suffering from both AP and MD.

MD is defined as an embryological remnant of the vitelline duct or omphalomesenteric duct during the 5th to 7th weeks of fetal life.[19] Most MD are asymptomatic in children and adults, and symptomatic Meckel diverticula in children are more common in males than females (3:1ratio). It is usually located in the 2 ft proximal to the ileo-cecal valve, and commonly contains heterotopic mucosa, include gastric, pancreatic or jejunal tissue. Epigenetic studies on MD are rare. MiRNA, circRNA (circle RNA) is also involved in embryonic development.[20,21]

AP may result in obstruction, which can be suggested by plain abdominal radiographs. Some children were suspected to have AP by prenatal ultrasonography. Although AP can be diagnosed with CT and MRI,[22] this abnormality is usually identified during laparotomy procedures or laparoscopic exploration. Diamond-shaped duodeno-duodenal anastomosis is the appropriate treatment for AP. It is described that ultrasonography and CT usually cannot make the diagnosis as MD resembles a normal bowel loop,[23] but emission computed tomography may assist in diagnosis. Sometimes, irreducible pediatric intussusception is caused by MD acting as leading point of intussusception. The symptomatic MD should be resected and the surgical decision regarding segmental bowel resection or wedge-shaped excision is dependent on the height-dimension ratio.[24] Compared with conventional open procedures, laparoscopy is a safe diagnostic and therapeutic tool for MD, as the children recover quickly after laparoscopy. Considering the infant mentioned above, surgical procedure was performed as a duodenal obstruction was suspected, and AP was identified. Then owing to abdominal exploration, MD was observed near the ileocecal valve, which underlines the importance of abdominal exploration of the digestive tract during malformation surgery.

4. Conclusion

The combination of AP and MD is an extremely rare event and surgical intervention is needed. Our experience demonstrates that abdominal exploration is essential for children with gastrointestinal malformations.

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Author contributions

Conceptualization: Peng Cai.
Data curation: Peng Cai, Zhenwei Zhu, Jianlei Chen, Lulu Chen.
Funding acquisition: Peng Cai, Zhenwei Zhu, Jie Zhu.

Investigation: Peng Cai, Zhenwei Zhu, Jianlei Chen, Lulu Chen, Jiang Pan, Wenxian Zhi, Jie Zhu.
Methodology: Peng Cai, Wenxian Zhi, Bin Wu.
Software: Peng Cai, Jie Zhu, Bin Wu.
Writing – original draft: Peng Cai.
Formal analysis: Zhenwei Zhu, Jianlei Chen, Lulu Chen.
Supervision: Zhicheng Gu, Shungen Huang, Jian Wang.
Writing – review & editing: Zhicheng Gu, Shungen Huang, Jian Wang.
Project administration: Jian Wang.
Resources: Jian Wang.

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