Case report

A rare congenital anomaly of the duodenal shape incidentally found at duodenopancreatectomy: A case report and literature review

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ARTICLE INFO

Keywords:
Duodenum
Congenital duodenal anomalies
Duodenal-shaped anomaly
A case report

ABSTRACT

Introduction: An anomaly of the duodenal shape is one of the rare congenital anomalies and remains poorly known in many previous studies and the literature. The duodenum is formed by developing the terminal foregut and proximal midgut through four stages in the embryonic period. According to the anatomy, the duodenum is typically described as C-shaped, U-shaped, or even horseshoe-shaped.

Presentation of case: The patient was hospitalized for abdominal pain and jaundice and diagnosed with ampullary carcinoma. During surgery, we incidentally discovered that the duodenum was not a C-shape. The first part of the duodenum and proximal midgut half of the second part descended the head of the pancreas. However, the distal half of the second part bent to the right and ascended upwards to the upper-right margin of the pancreatic head. After that, the third part ran slantingly downward to the left and posterior of the pancreas and portal vein.

Discussion: During the fifth week, the ventral pancreatic bud moves around the duodenum’s posterior side and unites the dorsal pancreatic bud at the sixth week. The place of the distal half of D2 migrated abnormally after ventral pancreatic bud rotation finished. The rapid and premature elongation of the proximal midgut, the influence of a very fast enlarged liver, or the early return of the umbilical loop combine with insufficiently developed abdominal space. These reasons may have led to the abnormal folding of the D2 position.

Conclusion: Knowledge about this anomaly helps clinicians know the duodenal-anatomical abnormalities.

1. Introduction

The duodenum derives from the caudal foregut and the cephalic midgut [1–3]. C-Shaped or horseshoe-shaped structure of the duodenum is formed by the gut rotation to the right together with the rapid growth of the dorsal pancreas, which pushes the duodenum from its origin midline to the right side of the abdominal cavity [2,3]. Defects, which occur in this embryonic stage, lead to congenital anomalies of the duodenum [4,5]. The incidence of congenital duodenal anomalies was one in 20,000 to 40,000 births [6]. These anomalies include malrotation, duodenal web, duodenal atresia [4,5,7]. Most of them present with the symptoms of gastrointestinal obstruction. The rest has no signs and incidentally finds out [6,8]. The anomalies of duodenum shape are considered to be rare according to literature review as well as previous case studies. The first case realized that the second part of the duodenum cooled like a serpent and the second case described the anomaly as a “head over heels” position of the duodenum [9,10]. Especially, both cases were discovered in cadavers. These cases have not collected any symptoms, diagnosis, and management. Therefore, an understanding of duodenal anomaly in a real-life patient case may be useful for clinicians to provide reasonable interventions. We report a case of a congenital anomaly in the shape of the duodenum, which is discovered in duodenopancreatectomy by chance. This case report followed SCARE guidelines [11].

2. Case presentation

A 35-year-old woman had a history without remarkable diseases and gastrointestinal symptoms. The patient presented with abdominal pain, anorexia, malaise, and jaundice a few weeks before hospitalization. Clinical biochemistry showed pancreatic amylase and lipase of 165 U/L and 432 U/L, respectively. Liver-function tests altered with a high value...
in conjugated bilirubin of 55 μmol/L, a rise of 310 U/L in alanine transaminase, and 150 U/L in aspartate transaminase. These changes predicted an acute biliary obstruction. The patient has received an upper abdominal magnetic resonance cholangiopancreatography (MRCP) for tumor diagnosis. Esophagogastroduodenoscopy detected a Vater's ampulla tumor, and the biopsy confirmed malignant histology. The congenital anomalies of the duodenum were unrecognized by radiologists and surgeons before surgery. A duodenopancreatectomy with standard lymphadenectomy was performed.

During operation, an abnormal shape of the duodenum was found due to a change in the position of the distal half of the second part (D2) and the third part (D3). We have completely exposed the duodenum during duodenopancreatectomy, so that duodenal abnormality was clearly detected. The stomach, the first part of the duodenum (D1), the proximal half of D2, and duodenojejunal flexure were normal sizes and anatomical positions. D1 and the proximal half of D2 descended the head of the pancreas. However, the distal half of D2 bent to the right. And it ascended upwards to the upper-right margin of the head of the pancreas (Fig. 2). After that, the beginning with D3 ran slantingly downward to the left and posterior of the pancreas and portal vein (Figs. 1A, 2B). Then the fourth part (D4) kept entering between the superior mesenteric artery (SMA) and abdominal aorta artery (Fig. 2B). The image of a replaced right hepatic artery (RHA) from SMA, variant hepatic arterial anatomy also was revealed (Fig. 3). After this procedure, an en-bloc specimen was resected, including tumor, duodenum-jejunum, head of the pancreas, and common bile duct.

The patient was discharged from our hospital after 9 days of treatment and postoperative care. Follow-up after 1 and 3 months showed a stable function of the digestive system and normal laboratory tests.

3. Discussion

The duodenum develops early in the fourth week of embryologic development from both the terminal foregut and proximal midgut [2,3]. The junction of this foregut and midgut occurs in D2, just below the bile duct's origin [3]. In our case report, the duodenal anomaly also appeared in D2 and lower to the major papilla. The position of the stomach, D1 and proximal half of D2 was normal. However, the distal half of D2 bent to the right and ascended upwards to the head of the pancreas (Fig. 2). Therefore, the duodenum did not develop the typical C-shaped configuration. This duodenal anomaly of shape and position has also been introduced in two previous reports by Indian authors. In the first report, Lalwani R. evaluated that D2 coiled like a serpent [9]. Nayak SB. called this anomaly as “head over heels” position of the duodenum in the other report [10]. We evaluated that this abnormality was caused by folding the D2 position at the junction of this foregut, midgut, or proximal midgut.

The first formation of the stomach, duodenum, pancreas, and bile duct becomes apparent from the fourth week of gestation [1,2]. The presumptive stomach begins to expand further into a fusiform structure while the dorsal and ventral pancreas starts to grow from the duodenal bud. During the fifth week, the ventral pancreatic bud moves around the duodenum’s posterior side and unites the dorsal pancreatic bud at the sixth week [2,3,12]. These rotations of the stomach bend the duodenum into C-shape and push it to the right. In our case report, the position of the proximal half of D2, pancreas, and major papilla were normal. Hence, the place of the distal half of D2 migrated abnormally after ventral pancreatic bud rotation finished.

Saunders JB. had divided the development of the duodenum into four stages: Stage I (rudimentary stage), stage II (formation of primary curves), stage III (establishment of the duodenal loop), and stage IV (fixation: the final stage) [13]. This author concluded that the third stage was the most important period, which affected the duodenal anomaly of shape and position. We evaluated that the congenital duodenal anomaly in our study occurred at this third stage. The reasons for this anomaly may include the rapid and premature elongation of the proximal midgut, or the influence of a very fast enlarged liver, or the premature return of the umbilical loop [13]. Combined with insufficiently developed abdominal space, these reasons may have led to the abnormal folding of the D2 position.

In this case report, the congenital anomaly of D2 did not cause duodenal obstruction. Therefore, the patient had a history without remarkable gastrointestinal symptoms. The clinical signs were detected on hospitalization due to Vater's ampulla tumor. Imaging studies, MRCP, and esophagogastroduodenoscopy, were unrecognized the duodenal anomaly by radiologists and surgeons before surgery. Maybe we only focused on the proximal half of D2, pancreas, common bile duct, and Vater's ampulla, so we missed the anomaly of the distal half of D2. After surgery, we re-examined the MRCP images and recognized this duodenal abnormality (Fig. 1). In addition, the replaced RHA was detected on computed tomography image reconstruction (Fig. 3B). The replaced RHA, arising from SMA, was one of the most variations of the hepatic artery found in 10.1-13.2% of cases [14–16].

We only found the duodenal abnormality and a replaced RHA in this case report. During operation, we also evaluated the blood vessels. The superior pancreaticoduodenal artery, which supplied the proximal duodenum from the foregut, was a branch of the gastroduodenal artery. Whereas the inferior pancreaticoduodenal artery supplied the distal duodenum from the midgut. This artery was separated from the SMA and located just below the replaced RHA. Therefore, this duodenal abnormality did not lead to associated vascular abnormalities. In this patient with intraoperatively detected duodenal abnormality, we had to

![Fig. 1. Preoperative MRCP. (A) Post contrast T1-weighted axial image shows D3, at the level of D2 travels downward to the left, behind the head of the pancreas and enters between abdominal aorta and SMA; (B) T2-weighted axial image shows abnormal folding position of D2. D2 and D3: The second and third part of the duodenum; CBD: Common bile duct; HP: Head of the pancreas; IVC: Inferior vena cava; SMV: Superior mesenteric vein; SMA: Superior mesenteric artery.](image-url)
approach more carefully. However, D2 and D3 running posterior of the pancreas did not become strongly adherent to adjacent organs. So we did not encounter difficulties for mobilization of the duodenum.

4. Conclusions

An abnormal shape of the duodenum, which results from irregular migration of the distal half of D2 in the embryonic stage, is a rare congenital anomaly. This abnormality is not always symptomatic. Sometimes it is incidentally discovered due to other reasons in the case of adult patients. Therefore, imaging and clinical diagnosis are often missed. Knowledge about this anomaly helps radiologists, endoscopist doctors, or even surgeons know the anatomical abnormalities encountered in clinical practice.

Sources of funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Ethical approval

There is no ethical approval was obtained as it’s a case report but a written consent was taken from the patient.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Provenance and peer review

Not commissioned, externally peer-reviewed.

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Minh Duc Pham: concept and design of the manuscript, data collection, data analysis and writing the paper.
Phuoc Vung Doan: data collection and data analysis.
Minh Thao Nguyen: concept and design of the manuscript, data collection, drafting, revision.
All authors participated in the approval of the final version.

Declaration of competing interest

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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