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Case Report

Congenital Duodenal Obstruction with Extra pancreatic Wirsung Duct

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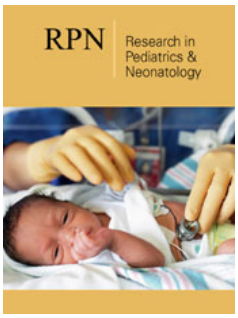
ISSN: 2576-9200



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Submission: April 27, 2023
Published: May 09, 2023

Volume 7 - Issue 5

How to cite this article: Shchapov Nikolay*, Ekimovskaya Ekaterina and Kulikov Denis. Congenital Duodenal Obstruction with Extra pancreatic Wirsung Duct. Res Pediatr Neonatol. 7(4). RPN. 000671. 2023. DOI: 10.31031/RPN.2023.07.000671

Abstract

An annular pancreas may cause congenital duodenal obstruction (CDO) or remain asymptomatic. In our practice, we came across atypical extra pancreatic location of the Wirsung duct in a newborn patient with CDO. Such duct malformation was unique in our practice and has not been described in literature yet. We report this case and speculate on embryology mechanisms that could lead to it.

Materials: From 2017 to 2021, 27 children with duodenal obstruction were treated in our department an intraoperative complication occurred in one child-it was the injury to atypically located Wirsung duct. A male child, born on 35 weeks of gestation had abdominal distention after birth. The examination revealed duodenal obstruction, an atrial septal defect and Down's syndrome. The child underwent laparoscopic procedure on his 6th day of life. During blunt division of transverse-duodenal ligament the abnormally located duct was damaged as it was located in the ligament tissue but not within the pancreatic parenchyma. We performed Kimura duodenal anastomosis with suturing the duct into it.

Result: After surgery the child suffered from infection with metabolic disorders and gastric bleeding, on the 5th postoperative day anastomosis leak occurred, which demanded further duodena-pancreato-neuroanastomosis with a Roux-en-Y loop and cholecystectomy. No surgical complications were seen afterwards. We consider this case is worth discussing regardless its poor outcome (the patient died on the 16th day after the last operation) as it is a unique observation of pancreatic duct anomaly.

Conclusion: We report a rare variant of the anomalous location of the Wirsung duct. Unawareness of its possible extra pancreatic location could lead to its rupture and further demand of reconstructive operation. To explain the complex embryogenesis of annular pancreas and pancreatic ducts malformations further research is needed.

Keywords: Congenital duodenal obstruction; Annular pancreas; Extra pancreatic Wirsung duct; Pancreatic embryogenesis; Children

Abbreviations: CDO: Congenital Duodenal Obstruction; AP: Annular Pancreas; CRP: C-Reactive Protein; MRI: Magnetic Resonance Imaging

Introduction

Congenital Duodenal Obstruction (CDO) is a rare disease that occurs with a frequency of 1: 5000-10000 newborns [1]. The main causes of blocked intestinal passage are duodenal membrane, duodenal atresia, and duodenal stenosis caused by annular pancreas (AP) [2]. To restore passage via duodenum a large number of various techniques have been developed with both their advantages and disadvantages. The anastomosis proposed by Kimura [3] made it possible to change the results of treatment of children with CDO radically and then it became the gold standard for this disorder. Such a success of the operation is explained by the fact that a bypass anastomosis is applied. It can be used for all types of obstruction, without involving either the pancreas, or its ductal system, or the bile tract. Since 2001, this procedure has been performed laparoscopically [4] and now it is the operation of choice for many pediatric surgeons all over the world. In our practice, we had one case of intraoperative complication due to atypical extra pancreatic location of the Wirsung duct. Such a malformation was unique in our experience and it has not been described in the world literature until now.

Case Report

From September 2017 to December 2021, 27 children with duodenal obstruction were treated in the Neonatal Surgery department of the Moscow regional center for maternal

and childhood healthcare. The cause of obstruction was duodenal membrane in 13 cases (48%), AP in 10 patients (37%) and in 4 cases (15%) we saw duodenal atresia with diastasis between the proximal and distal parts of duodenum. All children underwent Kimura procedure, in 4 children it was an open surgery, and in 23 children-laparoscopic one. Intraoperative complication occurred in one child-it was an injury to the abnormally located Wirsung duct. A male child from a 37 years old woman with no somatic disorders (IX pregnancy, VI childbirth). During pregnancy she had toxicosis, colpitis (cured), acute respiratory syndrome at 30 weeks (without antibiotic therapy). Antenatal Ultrasound did not find any malformation. The woman had spontaneous delivery at 35 weeks of gestation with weight 2560g, length 50cm, head 32cm, chest circumference 30cm, Apgar score 7/8. The child had a phenotype with mongoloid eyes, flattened nose, Gothic palate, low-set ears, his abdomen was large due to distention. Chest and abdomen x-ray showed duodenal obstruction, which was proved by abdomen ultrasound exam. Echocardiography revealed an atrial septal defect. Karyotyping revealed chromosome 21 trisomy. A laparoscopic Kimura duodena-duodenal anastomosis was performed. During duodenum mobilization an abnormally located duct was damaged. It was not located in the pancreatic parenchyma, but passed in front of the head of the pancreas on its anterior surface from medial to lateral side as part of transverse duodenal ligament and merged with choledochal to the right of the duodenum, opening with a common aperture above the obstruction area (Figure 1). When we bluntly dissected the ligament, the duct was ruptured, since it was impossible to differentiate it within the ligament tissue. The duct took a tubular shape after the rupture. As no bleeding was observed we suspected it was the pancreatic duct (Figure 2). So, we made anastomosis with suturing the duct into it. The posterior lip of the duodeno-duodenoanatomosis was formed with a continuous suture (PDS 6/0). Then the proximal part of the pancreatic duct was sutured into the medial angle of the anastomosis with interrupted sutures. The operation was completed by suturing the anterior lip of the duodeno-duodenoanastomosis. We placed abdomen drainage to the site of the operation.

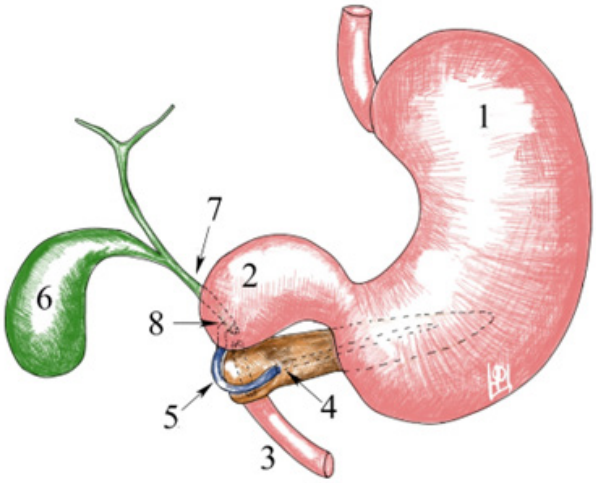

Figure 1: Scheme of the malformation: 1-Stomach, 2-Proximal duodenum, 3-Distal duodenum, 4-Pancreas, 5-Extrapancreatic Wirsung duct, 6-Gallbladder, 7-Choledochus, 8-Common orifice of choledochal and pancreatic duct.

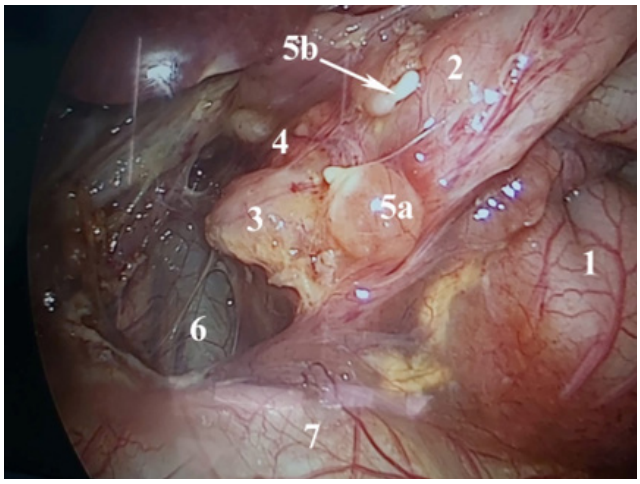

Figure 2: Intraoperative picture after damage of the pancreatic duct: 1-Stomach, 2-Proximal duodenum, 3-Distal duodenum, 4-Annular pancreas, 5(a)-Proximal (extended) part of the Wirsung duct, 5(b)-Distal (narrow) part of the Wirsung duct, 6-Inferior vena cava, 7-Transverse colon.

Result

The operation time was 115 minutes. The child had severe gastric bleeding on the first day after the operation, and his blood tests showed increase of infection (neutrophilia, metamyelocytes, increase of C-reactive protein) and further acute renal injury. We observed anastomosis leak on the 5th postoperative day. Further operation included duodena-pancreato-jejunoanastomosis with a Roux-en-Y loop and cholecystectomy. Unfortunately, due to severe infection the child developed multiple organ failure with lethal outcome on the 16th postoperative day after the last procedure.

Discussion

The pancreas is formed from the two buds: dorsal (located behind the duodenum, along its mesentery) and ventral (formed from the bile duct), and each bud has its own duct. When the duodenum rotates, the ventral bud moves dorsally, appears behind the dorsal pancreatic bud and they merge. While the duct of the ventral pancreatic bud becomes the main one (Wirsung), the dorsal duct is either reduced or persists as an additional duct (Santorini). Sometimes, during duodenum rotation, the bile duct and the proximal part of the ventral bud move to the right, while the distal part of the bud "moves" in the opposite direction and appears in front of the dorsal bud. Merging dorsal and ventral buds ring the duodenum [5]. AP occurs in less than 1% of the world's population [6]. Under certain conditions, this type of pancreas can compress the duodenum and lead to its total obstruction. This cause of duodenal obstruction occurs in about 8% of cases [2]. Also, AP can be an incidental finding in adult patients [6-8]. So, Alkhayyat et al. [9] in their study identified AP in 210 people among 6,162,600 patients who underwent abdominal examination over a 5-year period. However, we did not find any reports on extra pancreatic location of the Wirsung duct either in medical literature or in other studies of human and animal pancreatobiliary system. Discussion of this anomaly with colleagues (pediatric surgeons and experts of pancreatic surgery in adulthood) proved that our observation was unique.

In order to explain phenomenon of the AP and malformation embryology, we have two hypotheses. The first is that excessive growth in the distal direction violates the rotation of the ventral pancreatic bud. The rotation of the duodenum does not allow to overcome the inertia of the distal segment. The latter wraps around the duodenum, merging with the dorsal bud, its proximal and distal ends. The second hypothesis is similar to liver embryogenesis, when endothelial cells migrate to the vessels coming from the yolk sac, which eventually become the liver parenchyma. We can suggest that pancreatic ductal system is formed firstly, and only after that endothelial cells migrate and form pancreatic parenchyma. Then we can assume that, under certain conditions, migration occurs only on one side of the main duct of the ventral pancreatic bud, which further leads to extra parenchymal location of the Wirsung duct (Figure 3). This hypothesis needs further research, but has strong potential to explain the observed anomaly.

In most cases preoperative examination of children with CDO is limited to radiography, which demonstrate "double bubble sign" [2,10]. If this sign is not clear, it is possible to insert 30-60ml of air through the gastric tube and repeat an X-ray [2], or a contrast GIT examination can be performed [10]. An ultrasound exam may add important information: wide proximal part of the duodenum with thick walls, as well as annular pancreas [11], but it is not so important if Kimura procedure is going to be used. However, the possibility of intraoperation trauma of an extra parenchymal Wirsung duct makes us to look for preoperative imaging of pancreatic duct anatomy and Magnetic Resonance Cholangial-Pancreatography (MRCP) is the best method. It shows the ductal system in children with annular pancreas and provides more information for detailed preoperative plan. In our practice, we were not able to use MRCP, so we performed a precise blunt dissection of transverse duodenal ligament with special attention paid to any tubular structures to identify Wirsung duct possible anomalous location.

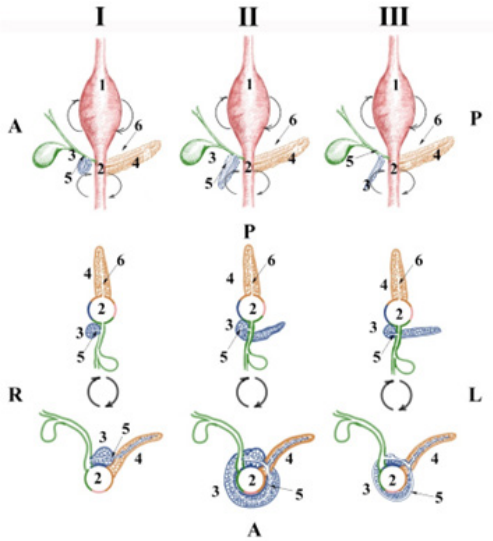

Figure 3: Scheme of pancreatic embryogenesis: I-Normal, I-Annular pancreas, III-Extra panreatic Wirsung duct; sides: A-Anterior, P-Posterior, R-Right, L-Left; 1-Stomach, 2-Duodenum, 3-Anterior pancreatic bud, 4-Posterior pancreatic bud, 5-Wirsung duct, 6-Santorini duct; ↻ ↺-stomach and pancreatic rotation direction (90° clockwise around longitudinal axis).

Nowadays, mortality rates in patients with CDO decreased up to 5-10%, due to better surgery techniques, innovative materials and therapy strategies [12]. It is no longer the malformation itself, but a concomitant pathology, particularly cardiac anomalies cause lethal outcomes [2,10,12]. However, sometimes we see a vicious practice to exclude patients with lethal outcomes from study cohorts because these patients suffered and died from other severe conditions with no regard to CDO. We argue that an in-depth analysis of adverse outcomes and complications may provide for new scientific discoveries and surgical approaches. In our case, the combination of CDO with Down's syndrome, heart disorder, severe infection worsen postoperative period. But we think this case is worth to consider regardless its poor outcome.

Conclusion

We report a rare variant of the anomalous location of the Wirsung duct. Unawareness of its possible extra pancreatic location could lead to its rupture and further demand of reconstructive operation. To explain the complex embryogenesis of annular pancreas and pancreatic ducts malformations further research is needed.

Funding Statement

This research did not receive any specific grant from funding agencies in the public, commercial or not-for-profit sectors. The authors have no conflicts of interest to declare.

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