An exceptional cause of duodenal obstruction detected antenatally: A compressive preduodenal portal vein

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Abstract

The authors report a case of a duodenal obstruction with an antenatal ultrasound suggestive of a duodenal atresia. At birth, the newborn was symptoms free. 3 weeks later, she was admitted for persistent nonbilious vomiting. Laparotomy, performed with the diagnosis of duodenal stenosis, demonstrated a compressive preduodenal portal vein (PDPV) which was the primary cause of the duodenal obstruction. Despite its exceptional occurrence, a compressive PDPV must be included in the differential diagnosis of a congenital duodenal obstruction. Its embryological study is discussed to heighten awareness of its identification and avoid the potential surgical difficulties. The discussion also focuses on the pre and postnatal ultrasound features in the assessment of the diagnosis of a PDPV. The recommended surgical procedure is the loose overbridging duodenoduodenostomy

1. Case report

Our patient was a full term girl weighing 3020 g with an Apgar score of 10. The antenatal ultrasound showed a double bubble suggestive of a duodenal atresia (Fig. 1). Physical examination after birth was normal and the abdominal x-ray showed no intestinal obstruction. She passed meconium at day 1. Feeding tolerance was correct and she kept a good condition during the stay. After multidisciplinary consultations, a mutual consensus was to discharge the baby at day 4 with clinical and ultrasound controls. Two weeks later, on the subsequent follow-up, the baby was doing well with a normal abdominal ultrasound.

At 27 days of life, she was transferred to our unit for persistent nonbilious vomiting and weight loss. The baby was slightly hypotonic and dehydrated. The abdomen was soft with no abdominal mass. The abdominal x-ray showed a double bubble evoking a duodenal stenosis with a little air beyond the duodenum. A barium meal (Fig. 2) confirmed the dilatation of the stomach and of the first part of the duodenum followed by a short stenotic zone and an intestinal malrotation. After rehydration, laparotomy was performed 3 days later. It was preceded by a gastroscopy which visualized a gastric dilatation, a normal pylorus, a dilatation of the first part of duodenum and especially a complete obstruction at the end level of the first part of duodenum. No duodenal web was seen. The laparotomy found a quiescent intestinal malrotation and a compressive preduodenal portal vein which was seen crossing in front of the end level of the first part of a distended duodenum (Fig. 3). A polysplenia was found. A longitudinal duodenotomy allowed eliminating an intrinsic obstruction. A Ladd's procedure

Abbreviations: PDPV, preduodenal portal vein; D1, first part of the duodenum; RVV, right vitelline vein; LVV, left vitelline vein.
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was performed followed by a loose overbridging duodenoduodenostomy with a wide loop preventing the portal vein from being obstructed by duodenum. Finally, the appendix was removed (Fig. 4). Postoperative period was uneventful. The baby was discharged at day 13. The barium meal at one month demonstrated the permeability of the anastomosis. Physical examination one year later showed a healthy child. The Doppler ultrasound found a permeable portal vein with a hepa-topetal blood flow. And the abdominal CT scan showed the anterior position of the portal vein (Fig. 5).

2. Discussion

The PDPV is an extremely infrequent condition. The understanding of the vascular anomaly requires its embryological study. The portal vein is built by the caudal part of the left vitelline vein, the retroduodenal anastomosis ultrasound and the cranial part of the right vitelline vein (Fig. 6) [1]. From the left vitelline vein forms also the superior mesenteric vein and splenic vein and they could be situated dorsally or ventrally to the fourth duodenum. Variations in the position of the portal vein may result from an anomaly of obliteration or an abnormal rotation.

Theoretically, the primary anomalies of development of PDPV are numerous with 22 types described and may be due to an abnormal obliteration of the middle anastomosis which requires the permeability of other segments. Two forms are mostly frequent. In case of persistence of the left vitelline vein, a pre-duodenal vein is displaced to the left in front of pylorus. If the right vitelline vein remains together with the inferior anastomosis, a preduodenal portal vein is displaced to the right lying in front of the descending part of duodenum. Among rotation anomalies, stomachal and duodenal situs inversus can induce a preduodenal location of the portal vein while the embryological development is normal.

The intestinal malrotation and situs inversus are the most frequently associated malformations. Both can be explained embryologically by a variable lack of rotation of the gastroduodenal

Fig. 1. Antenatal ultrasound at 36 weeks gestation shows the dilatation of the stomach and the duodenum.

Fig. 2. Upper gastrointestinal contrast study shows the dilatation of the stomach and of the first part of duodenum.

Fig. 3. Intra-operative view: a blood vessel passes over the first portion of duodenum (PDPV).

Fig. 4. The loose overbridging duodenoduodenostomy.
bloc and the intestinal loop \[2\]. It is not easy to distinguish whether the preduodenal position of the portal vein is due to regressions of the vitelline veins and their anastomoses, the result of abnormal rotation of the gastroduodenal or intestinal loop or if it is a combination of both.

In the malformation without situs inversus, it may be a primary abnormality of the portal vein associated with some degree of intestinal malrotation. This explanation may be applied to our case. Nakajima et al. \[3\] incriminate an autosomal recessive transmission of the PDPV on the basis of genetic studies in mice.

The interest of our case is to highlight the misguided ways in the assessment of the diagnosis of the PDPV in front of a duodenal obstruction. A review of literature acknowledges the same fact as, in all cases; the final diagnosis was delayed until the laparotomy. The exceptional occurrence of the PDPV as a primary cause of a duodenal obstruction, the misunderstanding of this vascular anomaly, the lack of specific prenatal, clinical and radiological features may explain the usual misdiagnosed etiologies in a duodenum obstruction due to a PDPV.

By itself, the PDPV is rarely the primary cause of a duodenal obstruction. The most common incidence is that of a newborn infant with duodenal obstruction secondary to an associated

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**Fig. 5.** CT scan showing the PDPV after duodenoduodenostomy.

**Fig. 6.** Schematic representation of the normal portal vein and its main anomalies. LVV, left vitelline vein; RVV, right vitelline vein. From Flament \[2\].
intestinal malrotation, duodenal atresia or duodenal web or annular pancreas. In a 25-year retrospective study in a single center, PDPV was found in five neonates. In all of them, the PDPV was asymptomatic and the duodenal obstruction was due to associated malformations [4]. In a review of literature from 1980 to 2012, we only collected 4 compressive PDPVs which were a clear evidence of lack of intrinsic duodenal obstruction [5–8] (Table 1). In all these cases, the preoperative diagnosis of duodenal stenosis or pyloric atresia was wrong.

A prenatal diagnosis of duodenal obstruction is suggested by polyhydramnios in the presence of a double bubble sign. Because duodenal atresia or stenosis, duodenal duplication, prepyloric antral diaphragm and PDPV may share these common sonographic features, the prenatal ultrasound cannot point out to the exact etiology of a duodenal obstruction. Nearly all of them were diagnosed as duodenal atresia which represents the most frequent etiology of duodenal obstruction.

The antenatal detection of a duodenal obstacle due to a compressive PDPV is exceptional, there is only one case reported [6]. In our case, the antenatal detection of dilatation of the fetal stomach and a duodenal bulb led to the diagnosis of duodenal atresia. At no moment the suspicion of compressing PDPV had been raised. At birth, the newborn was free of any symptoms and the abdominal x-ray showed no intestinal obstruction.

At 27-day of life, the baby was admitted for persistent non-bilious vomiting and dehydration. This evolution orientated to the diagnosis of an incomplete duodenal obstruction due to a partial duodenal web. This diagnosis was supported by the abdominal x-ray and an upper gastrointestinal radiological examination.

The preoperative ultrasound is helpful in the assessment of the diagnosis of a PDPV. The resultant portal vein courses anterior to the pancreas and duodenum. The diagnosis is made by identifying the prepancreatic course of the portal vein on transverse or sagittal images on Doppler ultrasound [10]. In all cases reported, the preoperative ultrasound was not performed, as the PDPV was not thought to be the primary cause of duodenal obstruction. This prepancreatic flow of the portal vein may also be visualized on CT scan or MR imaging [6–10].

In our case, the duodenal obstruction can be confidently described as being due to extrinsic pressure of the PDPV on the first part of duodenum. Some authors [2,4,11–13] denied the role of a low venous pressure within a thin-walled in the compression of the duodenum. Our case associated with others [5–8] confirm that the PDPV must be kept in mind in the differential diagnosis of a duodenal obstruction.

The preoperative assessment of PDPV as a cause of duodenal obstruction is the best but not an essential option, as, in all cases of duodenal obstruction, laparotomy is required. However, the understanding of this vascular anomaly is necessary in order to preserve the integrity of the vessel from an iatrogenic injury or a portal vein thrombosis.

Several procedures were advocated [2]: an end to end duodeno-duodenostomy transporting the portal vein to an ordinary retro-duodenal position, a gastrojejunostomy, a duodenojejunostomy and a Billroth II procedure.

The recommended surgical procedure is the loose overbridging duodeno-duodenostomy in order not to compress the blood flow of the portal vein. With the development of the laparoscopic surgery in neonates, this surgical correction may be safely done by minimally invasive procedures. Laparoscopic duodeno-duodenostomy can be successfully performed with an excellent, short-time outcome [14]. The Doppler ultrasound is recommended by Ohno et al. [15] to evaluate at follow-up the permeable portal vein with a hepatopetal blood flow.

3. Conclusion

Our case together with those reported in the literature demonstrate that, as exceptional as compressive PDPV may be, the surgeon has to be aware of it, as a cause of primary neonatal duodenal obstruction. A high level of suspicion is necessary. The use of preoperative Doppler ultrasound is helpful as it allows recognizing the vascular anomaly and preventing an operative iatrogenic damage from the PDPV.

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Conflict of interest

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Contributors statement

Antoine Mboyo: Dr Mboyo contributed to the surgical management of the patient, the conceptualization and design of the manuscript, reviewed and revised the manuscript, and approved the final manuscript as submitted. Soumeya Khadidja Khadir: Dr Khadir contributed to the conceptualization of the manuscript, reviewed and revised the manuscript, and approved the final manuscript as submitted Marie-Paule Guillaume: Dr Guillaume

Table 1

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<th>Symptoms</th>
<th>Imaging: abdominal x-ray, barium meal</th>
<th>Pre-operative diagnosis</th>
<th>Laparotomy</th>
<th>Associated malformations</th>
<th>Outcome</th>
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<tbody>
<tr>
<td>1. Georgacopulo</td>
<td>None</td>
<td>J0</td>
<td>Vomiting</td>
<td>Dilated stomach and D1</td>
<td>Duodenal stenosis</td>
<td>J4 compressive PDPV</td>
<td>Intestinal malrotation</td>
<td>Good</td>
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<td>(1980)</td>
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<td></td>
<td>Duodenoduodenostomy</td>
<td>Intestinal malrotation</td>
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<tr>
<td>2. Choi (1995)</td>
<td>Hydramnios</td>
<td>J0</td>
<td>Nasoantral tube; nonbilious clear fluid</td>
<td>Dilated stomach and D1</td>
<td>Duodenal stenosis</td>
<td>J4 compressive PDPV</td>
<td>Intestinal malrotation</td>
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<td>Double bubble</td>
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<td>Gastroduodenostomy</td>
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PDPV, preduodenal portal vein; D1, first of the duodenum.
contributed to the conceptualization of the manuscript, and approved the final manuscript as submitted. Richard Massicot: Dr Massicot contributed to the surgical management of the patient and approved the final manuscript as submitted. Vincent Flurin: Dr Flurin contributed to the medical management of the patient, reviewed and revised the manuscript, and approved the final manuscript as submitted. Abdelfetah Lalioui: Dr Lalioui contributed to the conceptualization of the manuscript, reviewed and revised the manuscript, and approved the final manuscript as submitted. Corina Zamfir: Dr Zamfir contributed to the conceptualization of the manuscript, reviewed and revised the manuscript, and approved the final manuscript as submitted. Marie-Thérèse Chevé: Dr Chevé contributed to the prenatal diagnosis of the patient, and approved the final manuscript as submitted.

References