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Case Report

Delayed presentation of duodenal diaphragm and annular pancreas in a 10-year-old girl: Case report ☆

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ABSTRACT

Congenital duodenal obstruction is a common cause of bowel obstruction. It is relatively easy to diagnose in the neonatal period. However, if the obstruction is due to a duodenal diaphragm, diagnosis may be delayed until later in infancy or even adulthood, depending on the size of the aperture in the diaphragm. Congenital duodenal obstruction may be associated with other gastrointestinal and biliary anomalies. The association of a duodenal diaphragm and a partial annular pancreas is a rare clinical entity. We present an unusual case of late presentation of duodenal diaphragm with partial annular pancreas in a 10-year-old girl with a 3-month history of abdominal distension and vomiting. The plain abdominal X-ray showed the classic picture of a double bubble. The CT images showed narrowing of the third duodenal portion and dilatation of the stomach and duodenum. The pancreatic tissue incompletely surrounded the second part of the duodenum. Intraoperatively, both the duodenal diaphragm and the partial annular pancreas were confirmed. Excision of the diaphragm and duodenoplasty were performed. A duodenal diaphragm should be suspected in patients with a history of abdominal distension and vomiting, even in late childhood. Associated gastrointestinal and biliary anomalies should always be excluded.

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Introduction

Congenital duodenal obstruction is an embryological defect due to failure of the duodenum to recanalize between the 6th and 8th week of pregnancy [1]. It is classified into 3 types [2]. Type I or mucosal diaphragm has been found to be the cause in up to 92% of cases [3–5]. Its early diagnosis remains a challenge as it can be missed both preoperatively and intraoperatively [6]. Preoperatively it may remain undiagnosed until beyond infancy depending on the size of the aperture in the mucosal diaphragm, and intraoperatively it may be missed in the absence of external wall deformity [7–9]. Duodenal obstruction may also be caused by extrinsic compression due to malrotation, preduodenal portal vein or annular pancreas [10–12]. Double duodenal obstruction due to both intrinsic and extrinsic causes is quite rare [13]. The association of a duodenal diaphragm with an incomplete annular pancreas is exceptional [14]. The pancreatic tissue extends from the head of the pancreas to partially encircle the descending duodenum [15]. We report a case of a duodenal diaphragm associated with a partial annular pancreas in a 10-year-old girl.

Case report

We report the case of a 10-year-old female patient with no medical or surgical history who presented with progressive abdominal distension and multiple episodes of non-bilious vomiting over a 3-month period with normal bowel transit.

She had significant abdominal distension with no evidence of organomegaly. She was well oriented and her vital signs were stable. Her labs, including a complete blood count, basic metabolic panel and liver function tests, were normal.

The initial plain abdominal radiography showed dilatation of the stomach and duodenum with the classic double bubble image (Fig. 1). An abdominal and pelvic CT with intravenous and oral contrast was performed. It showed an incomplete duodenal obstruction due to an endoluminal membrane of the third part of the duodenum causing a significant dilatation of the stomach and duodenum (Fig. 2). The contrast was passed into the bowel through a central aperture. Pancreatic tissue was seen partially surrounding the second part of the duodenum (Fig. 3). Malrotation was excluded. Based on the imaging findings, the diagnosis of a duodenal diaphragm associated with a partial annular pancreas was made. Upper gastrointestinal contrast study and esophago-gastro-duodenoscopy (EGD) were not performed.

The patient underwent surgery. Laparotomy confirmed the diagnosis of partial annular pancreas and normal bowel rotation. The duodenal external wall deformity provided guidance as to where to perform the duodenotomy. This revealed a diaphragm with a central aperture which was successfully excised. A nasogastric tube and a trans-anastomotic tube (TAT) were both inserted. A surgical drain was left in the peritoneal cavity near the duodenum before closure. There were no significant intraoperative difficulties. Histopathologic examination of the excised diaphragm revealed a duodenal mucosa.

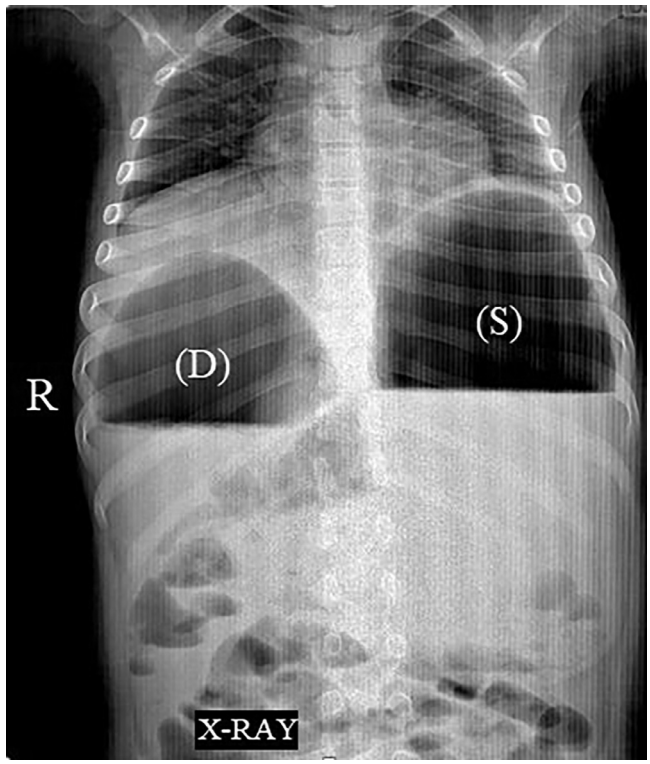

Fig. 1 – Plain abdominal radiography showing dilatation of the stomach (S) and the duodenum (D) (double-bubble image).

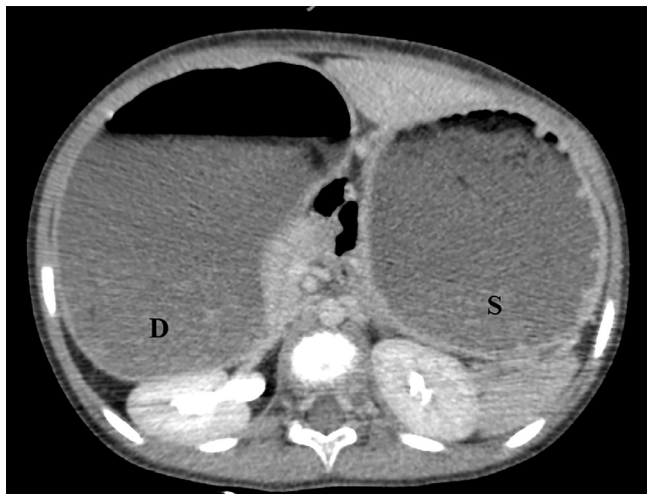

Fig. 2 – Axial CT scan showing significant dilatation of the stomach (S) and duodenum (D).

Postoperatively, our patient did well. She started oral feeding on day 10 and was sent home three days later. Her first follow-up at one month showed no complications. She gained 2 kg of weight.

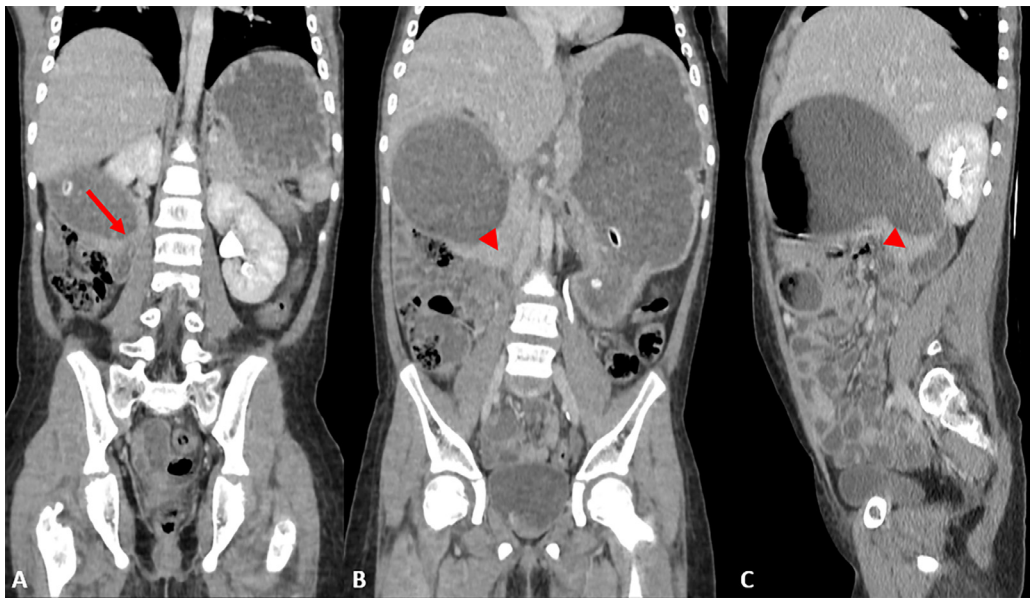

Fig. 3 – Coronal (A and B) and sagittal (C) CT scans showing the association of the duodenal diaphragm (arrow) and the annular pancreas surrounding partially the second part of the duodenum (arrowhead).

Discussion

The incidence of congenital duodenal obstruction in general has been reported by various authors to be around 1 in 6000 to 10000 live births [16]. The duodenal diaphragm has been reported to be the cause in up to 92% of cases [3,5]. As seen in our patient, a fenestrated duodenal membrane may delay diagnosis until late infancy or sometimes adulthood [16]. The duodenal diaphragm is associated with other anomalies in up to 78% of cases [17,18]. An association with Down's syndrome or abdominal situs invertus is well known, although not present in our patient [19–21]. However, a duodenal diaphragm and a partially annular pancreas is an unusual association and to our knowledge only a few cases have been reported in the literature [15]. The association of non-bilious vomiting and abdominal distension is reported to be the most common clinical presentation [22],23. The most common location of the diaphragm is between the first and second part of the duodenum, reported in 85% of cases [23]. In our patient, the membrane was located in the third portion of the duodenum, which is a rare case [24].

Although the classic image of a double bubble on plain abdominal radiography is pathognomonic, it is not specific [25]. Upper gastrointestinal contrast studies and esophago-gastroduodenoscopy (EGD) are important tools [26]. However, the possibility of an associated gastrointestinal and biliary abnormality should always be considered and further investigations should be performed accordingly. In fact, partial annular pancreas is often poorly recognized and may go undetected, hence the importance of advanced imaging techniques such as abdominal and pelvic CT with intravenous and oral contrast. This allows contrast examination of the gastrointestinal tract and adjacent abdominal structures.

Surgical excision of the duodenal diaphragm and duodenoplasty are the mainstay of treatment [25]. Although endoscopic excision is possible, it is not yet widely used [6].

The use of a trans-anastomotic tube (TAT) is controversial [16], but recent studies have shown that it allows early institution of oral feeding [27,28]. This was the case in our patient, who received a TAT, allowing oral feeding on postoperative day 10.

The survival rate of duodenal obstruction has improved to 90% in developed countries [11]. However, there are no studies from developing countries, where a long list of factors contributes to mortality, such as associated major congenital anomalies, prematurity, nosocomial infections and poor infrastructure [16].

Conclusion

Duodenal diaphragm and partial annular pancreas is a rare combination with delayed presentation due to incomplete duodenal obstruction. Associated gastrointestinal and biliary abnormalities should always be excluded, hence the importance of advanced imaging techniques.

Patient consent

An informed consent was obtained from the patient's parents

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