Case Report

A rare case of idiopathic congenital megaduodenum in adult misinterpreted during childhood: case report and literature review

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\textbf{A B S T R A C T}

Intestinal malformations are common disorders in newborn and favorable outcomes have been reported for such conditions. Although, if the patient is treated in a not experienced center, misinterpretation of the clinical and radiological findings may lead to errors in treatment and possible complications in adulthood. We report a case of a congenital megaduodenum which was misinterpreted as an intestinal malrotation resulting in late complications. The patient underwent a successful surgical resection of the duodenum with improvement of his clinical symptoms and nutritional status.

This case report emphasizes the importance of considering megaduodenum in the differential diagnosis of patients with feeding impairment, even during adulthood. Early diagnosis and treatment may improve patients’ outcome and reduce morbidity.

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\textbf{Introduction}

Megaduodenum can occur due to mechanical or functional abnormalities. In childhood, mechanical megaduodenum is frequently caused by congenital duodenal stenosis; however, there are other causes of mechanical megaduodenum including congenital bands, annular pancreas, and superior mesenteric artery syndrome [1,2]. Idiopathic megaduodenum in children is a rare disease defined as a massive

\textbf{Abbreviations:} CT, computed tomography; CT, computed tomography; DJB, Duodenal-jejunal bypass; DP, duodenoplasty; m, months; PR, partial resolution of the symptoms; R, resolution of the symptoms; UGS, upper gastrointestinal series; Y, years.

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dilation of the duodenum without mechanical obstruction [2–6]. Considering its rarity, idiopathic megaduodenum may be misdiagnosed and mimic other intestinal disorders, especially at nonexperienced centers. The aim of this study is to describe a case of an adult patient with idiopathic megaduodenum which was previously diagnosis as intestinal malrotation.

**Case report**

A 41-year-old man was referred to our clinic for evaluation of a chronic abdominal pain, nausea, vomiting, diarrhea, abdominal distention, and unintentional weight loss. The patient had a diagnosis of intestinal malrotation, which was detected in childhood during his first surgical procedure with 40 days old. Thereafter, the patient underwent several surgical procedures, including partial gastrectomy and gastrojejunostomy, with no improvement of his symptoms. The operating room report of the last surgery described that a tube duodenostomy was performed and almost 5 L of duodenal fluid was drained. He did not have comorbidities and did not use any medications. On physical examination, the patient was malnourished, his abdomen was distended, the bowel sounds were reduced in the upper quadrants and intestinal loops were palpable. Physical examination was otherwise normal. His family history was unremarkable.

Computed tomography scan with oral and intravenous iodinated contrast media was requested to evaluate his symptoms. Computed tomography scan demonstrated a marked dilation of the duodenum which occupied almost all abdominal cavity (Figs. 1A–D) and displaced the adjacent organs, including stomach (arrows) and liver (dashed arrow).
Fig. 2 – Illustration demonstrating the surgical technique. Patient already had a gastrectomy with a Roux-en-Y jejunal loop (A). Patient underwent a radical duodenectomy (B) and a small portion of the duodenum with Vater’s papilla was maintained and reimplanted in a Roux-en-Y jejunal loop (C).

organs, including the stomach (Figs. 1A and C) and the liver (Fig. 1C). The stomach was not distended due to previous gastrojejunostomy and there was no vascular compression or other mechanical compression of the duodenum. There is also a paucity of subcutaneous fat, supporting the clinical impression of malnutrition. A diagnosis of idiopathic megaduodenum was suspected and surgical treatment was indicated.

The patient underwent radical duodenectomy and a small portion of the duodenum with Vater’s papilla was maintained and reimplanted in a Roux-en-Y jejunal loop (Figs. 2 and 3). On pathologic examination, the duodenum demonstrated ganglionic cells in the submucosa and myenteric nervous plexus, excluding the diagnosis of aganglionosis, and the absence of other pathologic findings suggested the diagnosis of idiopathic cause. After the surgery, the patient gained weight and reported improvement of his symptoms. Hepatobiliary scintigraphy demonstrated biliary clearance and radiotracer activity in the small bowel with no abnormal accumulation (Fig. 4). The patient was revaluated 15 months after surgery with complete resolution of his symptoms.
Megaduodenum can occur due to mechanical or functional abnormalities. In childhood, congenital malformations are the main cause of mechanical megaduodenum, including congenital stenosis and bands. Complications of surgery are also a possible cause. The main causes of mechanical megaduodenum in adulthood are annular pancreas, adhesions, tumors, inflammatory lesions, aneurysms, and superior mesenteric artery syndrome [2,6–11]. Some other less common causes of mechanical megaduodenum are foreign bodies, parasites, and webs. Nevertheless, there are some cases without any mechanical cause that cause functional megaduodenum.

The etiology of primary functional megaduodenum remains unknown; however, some theories divide in 3 main causes as following: (1) paralytic, such as postvagotomy and in patients with Chagas disease; (2) neuropathic, including vitamin deficiency, congenital aganglionosis, porphyria, and diabetes mellitus; and (3) myopathic, such as collagen diseases [3–6,12,13]. In the presented case, there was no mechanical cause of obstruction and the histopathologic analysis of the duodenum did not demonstrate any significant abnormality in the duodenal wall and the myenteric plexus were normal.

Table 1 summarizes the main cases of megaduodenum described in the literature [1,4–25]. Patients with megaduodenum often have abdominal distention, nausea, vomiting, diarrhea, and malnutrition, especially in chronic cases and late presentation. Some patients also have atypical symptoms such as hematemesis, steatorrhea, and recurrent acute pancreatitis. Physical examination demonstrates distention with palpable bowel loops and, depending on the grade of the distention and the nutritional status of the patient, the megaduodenum can be visible. Considering that the clinical manifestations may be unspecific, it can result in a delay in the diagnosis [14,20]. The main radiological finding is dilatation of duodenum. In cases of mechanical megaduodenum, the case of obstruction may be detected; however, the functional ones no anatomic cause of obstruction is demonstrated. The treatment depends on the underlying cause and the degree of distention of the duodenum. In our case, the duodenum was extremely dilated and radical duodenectomy was performed with good results.

This case report emphasizes the importance of considering megaduodenum in the differential diagnosis of patients with feeding impairment, even during adulthood. Early diagnosis and treatment may improve patients' outcome and reduce morbidity.
| Author                        | Year | n  | Gender | Age (y) | Symptoms                                      | Imaging | Cause                      | Treatment | Outcome |
|------------------------------|------|----|--------|---------|-----------------------------------------------|---------|----------------------------|-----------|---------|
| Stutervant, et al [25]       | 1939 | 3  | 2F, 1M | 18, 34, 57 | Dyspepsia, vomiting, Abdominal pain, vomiting | UGS     | Duodenal obstruction       | -         | -       |
| Barnett, et al [16]          | 1954 | 1  | F      | 30      | Abdominal pain, vomiting and hematemesis | UGS     | Congenital duodenal stenosis | DJB       | R       |
| George, et al [1]            | 1967 | 1  | M      | 19      | Vomiting                                      | UGS     | Idiopathic                 | DJB       | PR      |
| Eaves, et al [12]            | 1985 | 3  | 2M, 1F | 17, 20, 23 | Abdominal pain, vomiting, and steatorrhea | UGS     | Duodenal web and duodenal obstruction | -         | -       |
| Barmeda, et al [20]          | 1966 | 6  | -      | Newborn | -                                             | -       | Duodenal atresia           | DP and DJB | R       |
| Mansell, et al [11]          | 1991 | 1  | M      | 29      | Vomiting                                      | UGS     | Myopathy (polymyositis)    | DJB       | R       |
| Buchman, et al [18]          | 1994 | 1  | F      | 21      | Nausea, abdominal pain                        | UGS     | Anorexia                  | Enteral probe | PR      |
| Weston, et al [13]           | 1998 | 2  | -      | -       | -                                             | -       | Myopathy (scleroderma)     | -         | -       |
| Endo, et al [19]             | 1998 | 2  | M      | 2, 8    | Vomiting, weight loss                         | UGS     | Duodenal web               | Duodenectomy | R       |
| Boeckxstaens, et al [14]     | 2002 | 1  | F      | 30      | Abdominal pain, vomiting, weight loss         | UGS     | Abnormal distribution of cell of Cajal | Jejunostomy | -       |
| Nichol, et al [22]           | 2004 | 1  | M      | 41      | Vomiting, gastric plenitude                   | UGS     | Myopathy (scleroderma)     | DJB       | R       |
| Barmeda, et al [22]          | 2006 | 1  | F      | 60      | Vomiting, gastric plenitude                   | UGS     | Idiopathic                 | DJB       | R       |
| Kudoh, et al [9]             | 2007 | 1  | F      | 48      | Vomiting, weight loss                         | UGS and CT | Myopathy (scleroderma)     | DJB       | R       |
| Lytras, et al [7]            | 2011 | 1  | F      | 75      | Episodes of acute pancreatitis, History of duodenal atresia | UGS and CT | Myopathy (scleroderma)     | DJB       | R       |
| Zaraa, et al [4]             | 2012 | 1  | F      | 29      | Vomiting, diarrhea, weight loss, peliagra     | -       | Duodenal atresia incompletely treated during childhood | Jejunostomy | -       |
| George, et al [15]           | 2012 | 1  | M      | 47      | Abdominal pain, vomiting, gastric plenitude   | UGS     | Idiopathic                 | Clinical treatment | R       |
| Zhang, et al [4]             | 2012 | 4  | 3F, 1M | 3 m, 10 m, 7, 12 | Vomiting, distention, palpable mass, weight loss | UGS     | Idiopathic                 | Duodenoplasty | R       |
| da Silva APC, et al [10]     | 2015 | 1  | M      | 32      | Vomiting, weight loss, hematemesis            | UGS     | Strongyloidiasis            | Partial gastrectomy, duodenectomy | R       |
| Park, et al [17]             | 2015 | 1  | M      | 77      | Abdominal pain, gastric plenitude             | UGS, CT | Idiopathic                  | Bezoar removal and change in eating habits | R       |
| Contreras, et al [23]        | 2015 | 1  | F      | 46      | Abdominal pain, nausea, vomiting              | UGS     | Myopathy (scleroderma)     | Jejunostomy | R       |
| Papis, et al [24]            | 2016 | 1  | M      | 20      | Vomiting                                      | UGS, CT | Familiar myopathy          | DJB       | R       |
| Yu, et al [5]                | 2017 | 1  | F      | Teenager| Abdominal pain, vomiting                      | UGS, CT | Chronic obstruction (bezoar) | DJB       | R       |
Authors contribution

NH: Data curation; Formal analysis; Methodology; Project administration; Supervision; Visualization; Roles/Writing - original draft; Writing - review & editing.
VBB: Data curation; Methodology; Roles/Writing - original draft; Writing - review & editing.
ESA: Data curation; Methodology.
RBSM: Data curation; Methodology.
MCCM: Conceptualization; Data curation; Methodology; Project administration; Resources; Supervision; Roles/Writing - original draft; Writing - review & editing.

Supplementary material

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2019.04.016.

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