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Case Report

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

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ABSTRACT

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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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

Abbreviations: 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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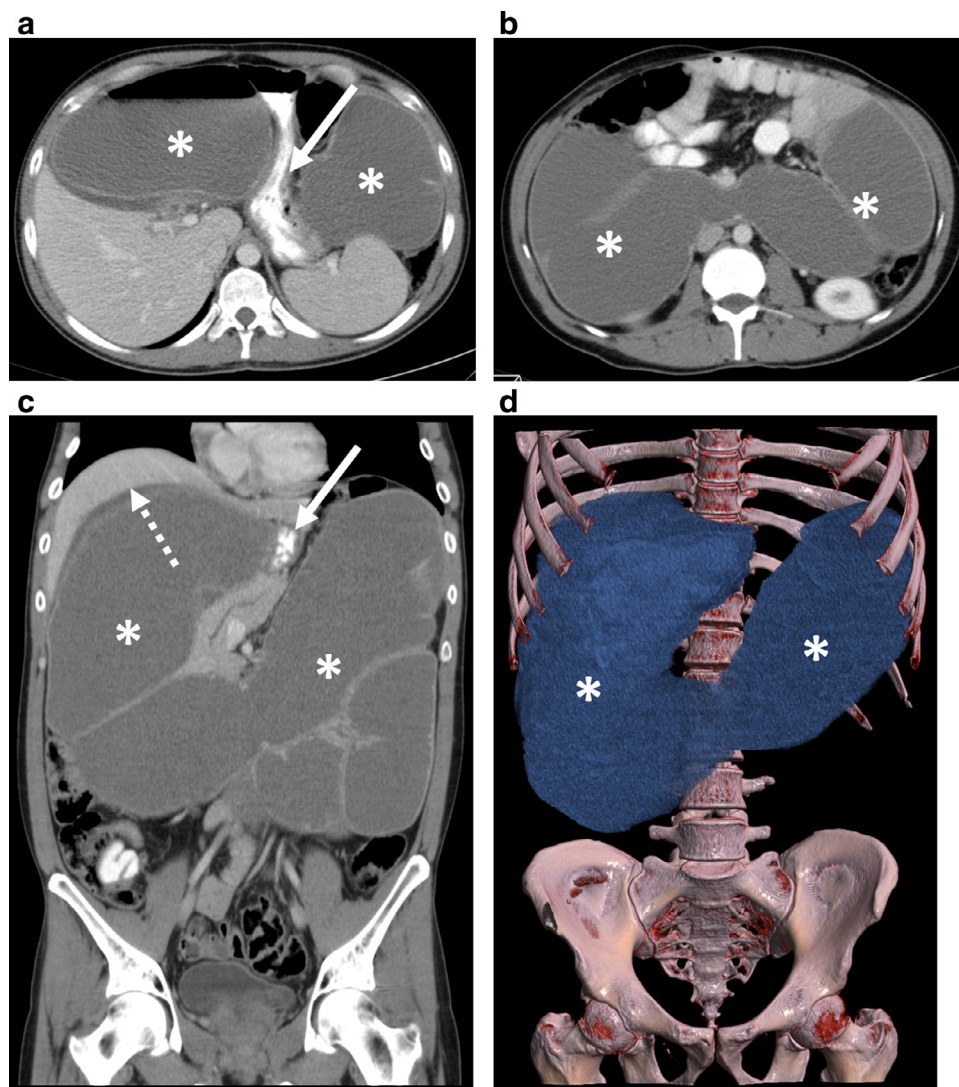


Fig. 1 – A 41-year-old man, with megaduodenum. Computed tomography scan with oral and intravenous iodinated contrast media in axial (A and B) plane, coronal plane (C) and 3-dimensional reconstruction (D) demonstrate marked dilation of the duodenum (asterisks). The duodenum occupies almost all abdominal cavity and displaces adjacent organs, including stomach (arrows) and liver (dashed arrow).

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

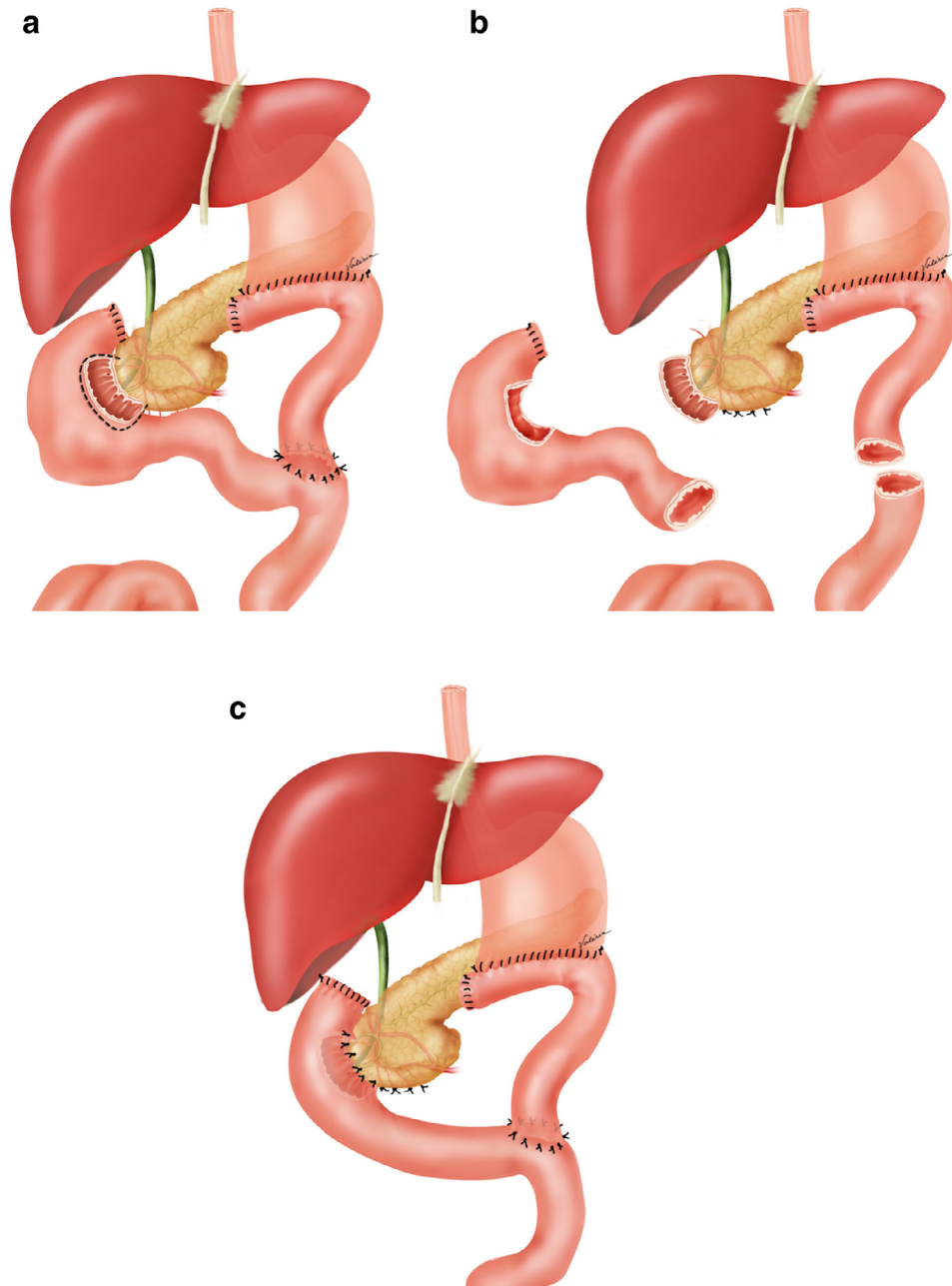


Fig. 2 – Illustration demonstrating the surgical technique. Patient already had a gastrectomy with a Roux-en-Y jejunal loop (A). Patient underwent a radical duodenectomy (B) and a small portion of the duodenum with Vater's papilla was maintained and reimplanted in a Roux-en-Y jejunal loop (C).

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 reevaluated 15 months after surgery with complete resolution of his symptoms.

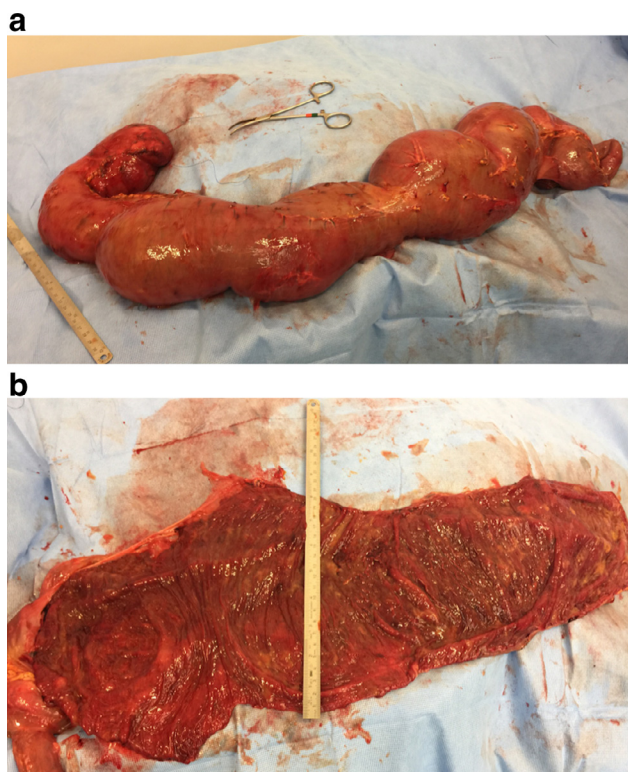


Fig. 3 – Surgical specimen demonstrates marked dilatation of the entire duodenum.

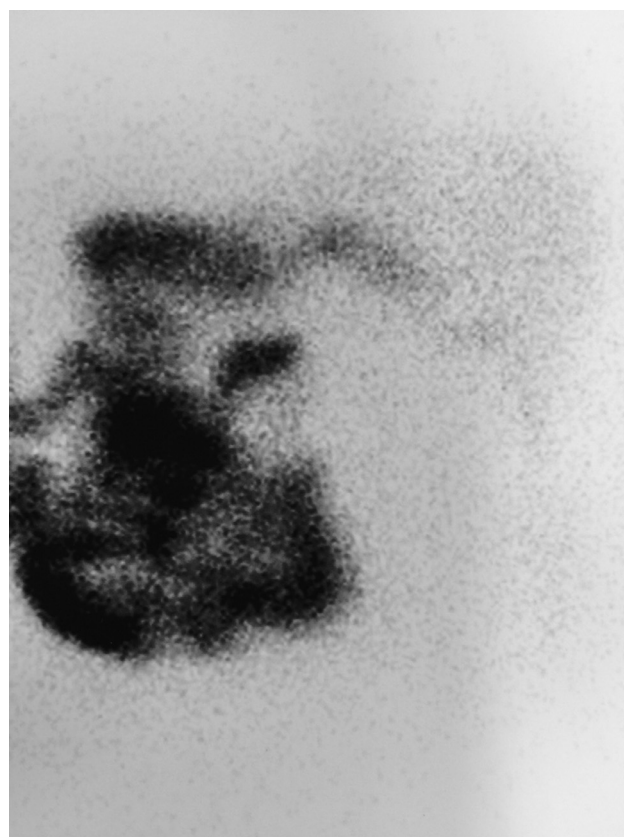


Fig. 4 – Hepatobiliary scintigraphy showed biliary clearance and radiotracer activity in the small bowel with no abnormal accumulation.

Discussion

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.

Table 1 – Summary of cases of megaduodenum described in the literature.

Author	Year	n	Gender	Age (y)	Symptoms	Imaging	Cause	Treatment	Outcome
Stutervant, et al [25]	1939	3	2F, 1M	18, 34, 57	Dyspepsia, vomiting	UGS	Duodenal obstruction	-	-
Barnett, et al [16]	1954	1	F	30	Abdominal pain, vomiting and hematemesis	UGS	Aganglionosis	DJB	PR
Chandler, et al [6]	1967	1	M	19	Vomiting	UGS	Congenital duodenal stenosis	DJB	R
Eaves, et al [12]	1985	3	2M, 1F	17, 20, 23	Abdominal pain, vomiting, and steatorrhea	UGS	Idiopathic	DJB	PR
Adzick, et al [21]	1986	6	-	Newborn	-	-	Duodenal atresia	DP and DJB	R
Mansell, et al [11]	1991	1	M	29	Vomiting	UGS	Myopathy (polymyositis)	D and jejunostomy	PR
Buchman, et al [18]	1994	1	F	21	Nausea, abdominal pain	UGS	Anorexia	Enteral probe	PR
Weston, et al [13]	1998	26	-	-	-	-	Myopathy (scleroderma)	-	-
Endo, et al [19]	1998	2	M	2,8	Vomiting, weight loss	UGS	Duodenal web and duodenal obstruction	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 [20]	2004	1	M	41	Steatorrhea, osteoporosis	UGS	Idiopathic	Duodenoplasty and DJB	R
Barmeda, et al [22]	2006	1	F	60	Vomiting, gastric plenitude	UGS	Aganglionosis	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	CT	Duodenal atresia incompletely treated during childhood	Excision of duodenal band and duodenectomy	R
Zaraa, et al [4]	2012	1	F	29	Vomiting, diarrhea, weight loss, pellagra	-	Idiopathic	-	-
George, et al [15]	2012	1	M	47	Abdominal pain, vomiting, gastric plenitude	UGS	Acromegaly	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 (not confirmed)	Partial gastrectomy, duodenectomy	R
Park, et al [17]	2015	1	M	77	Abdominal pain, gastric plenitude	UGS, CT	Idiopathic (associated with bezoar)	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	Familial 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.

PCCV: 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](https://doi.org/10.1016/j.radcr.2019.04.016).

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