FAIT CLINIQUE / CASE REPORT

Chronic volvulus on midgut malrotation leading to malnutrition and grow retardation: a case report of a 10-year-old boy.

Volvulus chronique sur malrotation du grêle conduisant à la malnutrition et à un retard de croissance: à propos d’un cas chez un garçon de 10 ans

BONG Yvonne Bih¹, NEOSSI GUENA Mathurin²³*, TAGNI Michele Sartre⁴, MOIFO Boniface¹⁵.

1: Faculty of Medicine and Biomedical Sciences, University of Yaoundé I (Yaoundé, Cameroon)
2: Department of Biomedical Sciences, Faculty of Sciences, University of Ngaoundéré (Ngaoundéré, Cameroon)
3: Imaging service, Ngaoundéré Regional Hospital (Ngaoundéré, Cameroon)
4: Medical Center la Cathedrale (Yaoundé, Cameroon)
5: Imaging service, Yaoundé Gyneco Obstetrics and Pediatrics Hospital (Yaoundé, Cameroon).

Mots-clés :
Volvulus, malrotation intestinale, transit gastroduodénal, échographie Doppler abdominale.

Keywords:
Volvulus, midgut malrotation, upper gastrointestinal seris, Doppler abdominal ultrasound.

*Auteur correspondant
Dr NEOSSI GUENA Mathurin
Service de Radiologie
Hôpital Régional de Ngaoundéré
BP 45 Ngaoundéré-Cameroun.
Email : mneossiguena@yahoo.fr
Tel : 00237 697692559

ABSTRACT
Midgut Malrotation (MM) with volvulus is a rare congenital pathology that usually presents in the neonatal period. It is unusual to come across this condition in the older child. However, cases of MM with chronic volvulus have been reported in older children. We present a case of MM with chronic volvulus in a 10 year old male who presented with recurrent episodes of generalised abdominal pain associated with bilious vomiting and constipation for 9 years. Diagnosis was made by upper gastrointestinal (UGI) series, B Mode and Doppler abdominal ultrasound. We therefore discuss the relevance and value of these technics in the diagnosis of this pathology in our milieu.

RÉSUMÉ
La malrotation intestinale avec volvulus du grêle est une pathologie congénitale rare qui se manifeste habituellement pendant la période néonatale. Elle est rare chez l’enfant plus âgé. Cependant, des cas de malrotation intestinale avec volvulus chronique ont été rapportés chez des enfants plus âgés. Les auteurs présentent ici un cas de malrotation intestinale avec volvulus chronique du grêle chez un jeune garçon de 10 ans présentant des épisodes récurrents de douleur abdominale généralisée associés à des vomissements bilieux et à une constipation pendant 9 ans. L’examen du transit gastroduodénal et l’échographie abdominale ont permis de poser le diagnostic. Nous discutons donc de la valeur de ces techniques dans le diagnostic de cette pathologie dans notre milieu.

1. Introduction
Midgut Malrotation (MM) is the failure of the midgut to rotate counter clockwise during embryological development of the gastrointestinal (GI) tract [1]. Arrest in midgut rotation at 180° leads to misplacement of the duodeno–jejunal junction to the right of the midline [1, 2]. In these cases, the small bowel mesentery has narrow vertical posterior attachments that make it prone to volvulus [3]. Not every patient with MM will develop midgut volvulus and therefore may be asymptomatic [2,4]. MM commonly presents in the first year of life, especially in the neonatal period, with symptoms of acute midgut volvulus; the likelihood of symptoms of acute
midgut volvulus decreases with age [5]. However, there are a few reported cases of chronic presentation, usually with non-specific symptoms making diagnosis challenging hence diagnostic delay is common [1]. Upper GI Series is the examination of choice for the diagnosis of midgut volvulus due to malrotation (~6). We report the case of a 10 year old male diagnosed of MM with chronic volvulus. The diagnosis was made by upper GI series, abdominal ultrasound and confirmed on laparotomy.

2. Observation
A 10 year old male was referred to us for recurrent episodes of generalized abdominal pain associated with bilious vomiting and constipation for 9 years. There was no history of fever during episodes, and he was well between episodes. His condition led to numerous consultations in different health facilities. The most recent episode had been treated with enema and amoebicides, but the persistence of symptoms led to referral to our health facility. Physical examination revealed a child who looked generally unwell and small for his age. The abdomen was mildly distended and soft, the percussion note tympanic and a succussion splash heard on auscultation. Rectal exams revealed faecalith in the rectum. Oesophagastroduodenoscopy (fibroscopy) was requested and the results revealed a marked gastric stasis without pyloric stenosis. Persistence of symptoms led to the request of an erect plain abdominal x-ray the following day which showed a double bubble (one gastric and the other at the bulb of the duodenum). After aspiration of gastric contents, erect plain abdominal x-ray showed a third airfluid level projecting over the spine corresponding approximately to the angle of Treitz (Figure 1).

An upper GI series was then requested indicated for suspicion of intestinal obstruction. The results revealed a prolongation of the stomach to the left iliac fossa (LIF), marked gastric distension and distension of the entire duodenal framework. Also the transverse diameter of the 2nd part of the duodenum was 8.3cm and the duodenum was well marked by mildly thickened transverse folds. There was duodenal opacification up to the angle of Treitz then progressive reduction of the intestinal caliber in a “rats tail” like manner and appearance of the twisting turn projecting over the left of the spine with a very mild passage at the twisting turn (Figure 2).

Complementary ultrasound in the B mode (Figure 3) put to evidence a prevertebral intestinal mass, malposition of the mesenteric vessels with marked dilatation of the mesenteric vein that had a varicose and tortuous aspect. Within this mass, followed from top to bottom, were images in "twisting turns". Real time images showed passage of the intestinal contents within the twisting turns.

Bases on these findings, the diagnosis of MM with chronic volvulus was therefore made.

This child was operated upon the same day. Intraoperative, was found numerous Ladd’s bands running from the right colic flexure to the small intestine; small intestinal and terminal ileum adhesions sandwiched in a massive felting thereby confirming the diagnosis. Also, the duodeno-jejunal angle was seen to the right of the spine and inversion of the mesenteric vessels seen too.
**Figure 2:** Upper GI Series: A: Posterior anterior view shows prolongation of stomach to LIF. B: Oblique view shows gastric distension and mildly thickened transverse duodenal folds and C: Oblique view shows intestinal caliber reduction in a "rats tail" like manner and the twisting turn.

**Figure 3:** B mode ultrasound: Transverse slice shows a prevertebral intestinal mass (A) and the dilated and tortuous SMV (B)

**Figure 4:** Transverse slice colour Doppler shows the SMV as it whirls around the SMA
3. Discussion
MM has been estimated to occur in approximately 1 in 500 live births [7] but not every patient with MM will develop midgut volvulus [2,4]. The incidence of volvulus decreases with age [8] and the clinical presentation varies with age. A majority of cases present during infancy with symptoms of acute volvulus including bilious vomiting, constipation, abdominal distension, diarrhoea and fever [5,7,8]. Chronic volvulus is rare and usually presents in late childhood or adulthood mainly with abdominal pain and less often with emesis or nausea [8,9]. Atypical clinical presentations with malabsorption-like symptoms such as chronic diarrhoea and failure to thrive have been reported [9]. These symptoms are non-specific to chronic volvulus and physical examination is usually not helpful. Diagnosis is therefore based on medical imaging results.

In this case, our patient was male. A male preponderance for chronic midgut volvulus has been reported [5] and there are several reported cases in males. Although some authors have reported an equal sex distribution [10].

The symptoms of chronic volvulus with MM are due to intermittent or partial twisting that results in lymphatic and venous obstruction. Several studies [5, 6, 8, 9, 10, 11] have reported intermittent bilious vomiting, abdominal pain and constipation as the main presenting symptoms in chronic volvulus with MM. Our patient presented with all of these symptoms.

The non-specific and intermittent nature of the symptoms pose a diagnostic challenge. For this reason diagnosis is based on a combination of clinical features and most especially medical imaging results. Upper GI series is the examination of choice [12] for the diagnosis of midgut malrotation with chronic volvulus. However, upper GI series has its drawbacks especially in less developed countries like ours where a remote controlled table and scopy are rarely available for the performance of a viable upper GI series in children. Abdominal ultrasound (US) with Doppler alone or in combination with upper GI series has been proven to be more adequate for the diagnosis in such circumstances [13]. Abdominal ultrasound will show: inversion of mesenteric vessels, abnormal pathway of mesenteric vessels, duplication of the mesenteric vein, anterior position of the mesenteric vein relative to mesenteric artery and the whirpwhool sign [14]. The presence of a whirpwhool sign on US is usually pathognomonic. In trained hands, US alone can be used to make the diagnosis of Midgut Malrotation with chronic volvulus.

4. Conclusion
Midgut malrotation with chronic volvulus is a rare pathology that is often missed due to its non-specific clinical presentation. The diagnosis is based on medical imaging including UGI series and abdominal US. It requires a high degree of suspicion in order for the clinician to recommend these exams. Despite the exposure to x-rays during UGI series and the difficulty of achieving this exam in our environment, the combination of UGI series, B mode US coupled to Doppler allow to assert the diagnosis of this pathology. We must systematically search the chronic volvulus in case of recurrent abdominal pains of the child.

Conflicts of Interest
The authors declare that there is no conflict of interests regarding the publication of this paper.

5. Références


9. Imamoglu M, Cay A, Sarihan H, Sen Y. Rare clinical presentation mode of intestinal malrotation after neonatal...

282


