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Intestinal Malrotation: Case Report

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Abstract: Intestinal malrotation is a malformation characterized by an anomaly in the rotation of the midgut around the axis of the superior mesenteric artery. Its diagnosis is difficult, given the rarity of the illness and the aspecificity and variability of the symptoms as in the case researchers have examined where the symptomatology was characterized by painful colic paroxysms together with closed bowels which passed spontaneously. A correct diagnostic-therapeutic approach together with a strong suspicion is therefore, fundamental for a complete resolution of the clinical picture. In the diagnostic investigation, the gold-standard is represented by a small intestine enema. Therapy is surgical and involves the Ladd technique.

Key words: Intestinal malrotation, duodenal obstruction, midgut, artery, small intistine, Italy

INTRODUCTION

Intestinal malrotation is a development anomaly resulting from an abnormal or incomplete rotation of the midgut during the development of the foetus. One out of every 500 live births in the USA has this malformation. However, the real incidence of malrotation is not known as it can be supposed that most of the conditions that remain asymptomatic during the pre-natal period remain so also later on and therefore are not diagnosed.

There are no significant differences in incidence between the two sexes. Around 60% of the cases are diagnosed within the 1st month of life, 20% between the 1st month and the 1st year of age and the rest after the 1st year of age with frequency decreasing as the age increases (Blanco-Gonzalez *et al*., 2011; Nardone *et al*., 2010).

In the paediatric period, duodenal obstruction or volvulus is most often responsible for the symptoms (Al-Jahdali *et al*., 2009; Applegate *et al*., 2006; Hagendoorn *et al*., 2011). This anomaly is rarely discovered after the first decade of life. The clinical picture is varied and the different manifestations depend on the phase in which the arrest of the morphogenetic process of the middle bowel occurs (Hanna and Akoh, 2010; Maciel *et al*., 2009).

Signs of embryo morphogenesis: The middle bowel is the segment that gives origin to the caudal part of the duodenum, the mesenterial small intestine, the cecum, the ascending colon and the proximal two thirds of the transverse colon. Initially, it is almost rectilinear. It then progressively undergoes an extension process that is faster in comparison to the body of the embryo forming a loopwith posterior concavity divided into two branches, superior and inferior from the course of the superior mesenteric artery. Between the V and the X week of intrauterine life, there is the extroflexion of the loop in the yolk sack. It is at this point that the process of rotation begins while the extension process continuous. After week XI, there is the re-entry of the middle intestine in the abdominal cavity and the completion of the process of anticlockwise rotation around the SMA axis which at the end is of 270°.

For this reason, the superior branch which lengthens more quickly in comparison to the inferior one is at first to be found on the right of the vascular axis and then passing below this, it migrates to the left, constituting the jejunum and the ileum; the inferior branch becomes first superior and then migrates to the right of the vascular axis, increasing in calibre to become part of the large intestine. In summary, researchers can divide this organogenetic process into three stages:

- The first stage occurs between the V and X week of intrauterine life and includes the herniation of the primitive loop of the yolk sac, the initial rotation (of 90°) and the re-entry in the abdominal cavity
- The second stage after the XI week characterized by the completion of the rotation process with the formation of the duodenal C placed posterior to the superior mesenteric artery with the ascending colon to the right, the transverse colon superior and the descending colon to the left

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- Finally, the third stage where there occurs the fixation of the mesentery, the descent of the cecum and the attachment to the posterior wall of the ascending and descending colon

If the morphogenetical process is blocked in the first phase, the outcome will be omphalocele. In the second stage, there can be:

Non-rotation: In which case, the small intestine will position itself in front and the colon at the back. Therefore, the duodenojejunal junction will not be found at the bottom and to the left of the SMA and the cecum will not reach the lower right quadrant. The result of all this will be that the mesentery root will be narrow, predisposing to volvulus.

Incomplete rotation: The duodenum will not go beyond the midline, continuing with the small intestine which is positioned to the right and the colon which is positioned to the left.

Therefore, there will be a normal fixation of the spleen angle with the left hypochondrium with the formation of a peritoneal band between the cecum and the right posterior abdominal wall (Ladd band) which passing in front of the duodenum will cause it to compress. Finally, the cecum will be set high against the root of the SMA, there will not be the duodenojejunal angle of Treitz and the mesentery root will be narrow. In this anatomical situation, a paraduodenal hernia can be formed if the rotation of the colon is not followed by that of the duodenojejunum.

Incomplete fixation: This will determine the formation of hernial pockets as there will not be a complete fixation of the folds of the right and left colon and the duodenum will not be fixed to the posterior wall of the abdomen. In this situation, a volvulus can occur due to the absence of the fixation of the cecum and of the ascending colon.

In the third stage, the block of the morphogenetical process will cause the absence of the fixation of the duodenum and the cecum will be mobile. For the sake of correctness, researchers would like to mention the anatomical-clinical classification of Mehall which identifies three variations:

- Typical: Ligament of Treitz absent or present to the right of the midline
- Atypical high: Treitz on the midline or to the left, above the XII thoracic vertebra
- Atypical low: Treitz on the midline or to its left, below the XII thoracic vertebra

The study cited which refers to 201 operations for malrotation on patients between the ages of 3 days and 20 years (with an average age, however <19 months) finds a normal position of the cecum in just 13% of the cases with a homogeneous distribution in the different age bands and in an almost superimposable association in the three different variations of the position of the ligament of Treitz.

The only significant difference that researchers want to underline would seem related to the relationship between age at the time of the operation and the position of the Treitz which is lower in patients with a tendency to a chronic symptomatology and therefore, operated on at a more advanced age. Finally, it is possible that malrotation regards only the duodenum with the positioning of the Treitz correspondent to the superior right quadrant of the abdominal cavity and the cecum positioned in its normal place even if often hypermobile. The symptomatological picture in adult patients varies and this is in relationship to the cause that determines it even if in a single case, it is not always easy to distinguish disturbances connected just to malrotation from those caused by the onset of a volvulus. The patient can submit himself to examination with acute abdomen or with a chronic clinical picture. Surgical treatment foresees the Ladd procedure described in 1936. The recent observation of a clinical case has induced us to evaluate a pathology that can be considered in the differential diagnosis of some cases of adult abdominal pain.

CASE REPORT

A 35 years old patient, male and non-smoker, non-drinker was examined by us because of recurrent abdominal colic with brief periods of alterations in bowel movement. This painful symptomatology of a colic nature was mainly located in the mesogastric region, coming post prandial and often with nausea and vomit.

During the anamnesis, the patient reported how this symptomatology occurred at least once a month, resolving itself spontaneously after a few hours or days and complaining about bowel disturbances in the same period characterized by diarrhoea or constipation without, however blood and/or mucus.

He had consulted several doctors about the problem, resulting in a long and difficult diagnostic investigation. First of all, an EGD was performed, leading to the diagnosis of a sliding hiatus hernia, grade A distal esophagitis and duodenitis. The examination of a bioptic sample showed slight but widespread atrophy, compatible with the hypothesis of celiac disease. For this reason, he was recommended to follow a gluten-free diet from which

the patient did not draw any benefit. A colonoscopy was therefore performed with multiple biopsies, revealing chronic active focal colitis. The 5-ASA was administered but without any benefit.

The tests for allergies and lactose breath tests were both negative. Suspecting that the problem was not organic in nature but psychosomatic, the patient underwent numerous visits to a psychiatry clinic and subsequently treated with benzodiazepine with a reported clear improvement in the clinical picture. It should be noted that in the clinical history of the patient, he underwent a video-laparocholecystectomy for gallstone cholecystitis, treated in another hospital. A few months after the operation, he developed an episode of acute edematous pancreatitis quickly resolved through medical therapy. He was finally examined by us for a new episode of widespread abdominal colic resistant to medical therapy. The patient had an emaciated aspect. In fact, he had not nourished himself adequately for some months. The objective examination of the abdomen revealed a moderate resistance of the abdominal wall in the higher quadrants, a slight increase in a-amylase (126 U I^{-1}) and in transaminases (60 U I^{-1}). Direct X-ray of the abdomen showed the absence of air-fluid levels and air crescent sign under the diaphragm; the ETG of the abdomen was negative for ecotomographic alterations of spleen, liver, pancreatic and renal parenchyma in the absence of free or loculated effusion.

A repetition of the EGD and the colonoscopy with pictures superimposable with the previous ones was felt to be useful. The diagnostic iter was completed with an enema of the small intestine that showed the right colon mobilized and placed alongside the left fold; this find was compatible with a wide mesentery that made us suspect a common mesenterium. Considering the repetition of the episodes of pain and their negative influence on the quality of life, it was thought opportune to proceed with an exploratory laparotomy. On opening the abdomen, an improper positioning of the cecum was seen, completely to the left and high, placed alongside the left fold (which was found to be positioned normally) with the loops of the small intestine completely to the right in respect to the ascending-cecum. The Ladd bands went from the cecum up to the parietal peritoneum in the right hypochondrium, these bands lay along the duodenum in a potentially compressive position. After the mobilization of the colon and the duodenum, there followed the lysis of the Ladd band and subsequently, the cutting of a ring compressing the superior mesenteric artery (Fig. 1 and 2). Having performed an appendectomy, it was not necessary to proceed with the resection of the loops undergoing Hypoxic suffering due to the normal resumption of their natural colour after the section of the ring. The post operative recovery was regular. He restarted eating per os from the 1st day and was discharged 3 days later. The follow up after 4 months did not reveal any significant symptomatology. The patient has started eating a regular diet and has achieved a slight gain in weight (3 kg in 1 month).

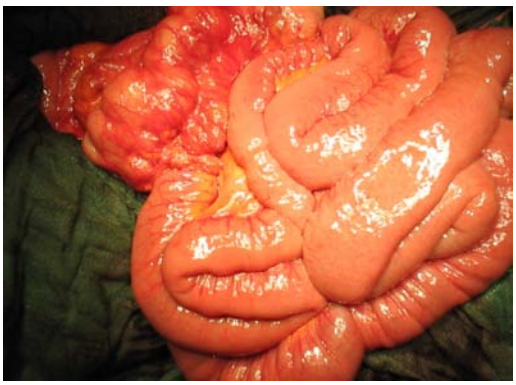

Fig. 1: Mesenterium commune

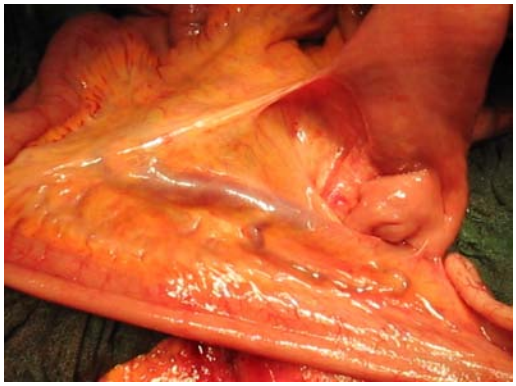

Fig. 2: Strangulated track superior mesenteric artery

RESULTS AND DISCUSSION

Intestinal malrotation is a pathology which is rarely found in adults. Its diagnosis in an adult is often difficult and delayed given the aspecificity and the variability of the symptoms. The revision of the literature (Hagendoorn *et al*., 2011; Nardone *et al*., 2010; Park *et al*., 2010; Ramirez *et al*., 2009) and the analysis of the observed case allow us to underline some aspects of the subject that researchers consider significant for tackling the anomalies of intestinal rotation. The rarity of the disease should not exclude the anomaly from being accurately looked for. This should be done through a detailed radiological examination with contrast medium

both of the higher sections (Enema of the small intestine) and the lower ones (Opaque enema) in patients with chronic abdominal pains which are not otherwise explainable; concerning this, it must be said that age cannot be a reason for excluding this anomaly given that markedly symptomatic cases submitted to operation in advanced age have also been reported therefore, symptoms such as recurrent abdominal pain, post prandial vomit, bilious, abdominal relaxation, gastroesophageal reflux variously coupled can justify a close radiological examination to search for signs of malrotation.

A radiological examination must be made to clarify the position of the duodenojejunal junction (normally to the left of the second lumbar vertebra) and of the cecum (right iliac fossa). All variations to these standards point towards the diagnosis. The radiological study is at the base of the anatomical overview of the patients. The scheme adopted by us based upon the position of the Treitz is carried out easily and allows us to foresee the disposition of the viscera in the pre-operating phase and to estimate the risk of a volvulus or internal hernia which is significantly higher in the typical malrotation in addition, it allows us to fairly approximately establish the probability of the surgical treatment being successful in fact, in the atypical forms of malrotation a return of the symptoms is reported in >10% of cases, compared to a complete absence of returning symptoms in the typical forms. This means that it should be carefully evaluated whether it is opportune to carry out the operation in subjects who, on the other hand have a lower risk of complications and whose symptoms could be not directly correlatable with the anatomical anomaly. The surgical indication does not find unanimous consent and seems to depend both on the type and entity of the malrotation and from the entity of the symptoms. There are reports of the treatment failing both in cases of non-rotation and in malrotation which regards only the duodenum.

The classification that researchers propose instead limits the failures only to atypical forms in 11-13% of cases while the typical formswith Treitz to the right of the midline or entirely absent do not have a return of symptoms after surgical intervention. It is unanimously accepted that the surgical technique to be adopted to deal with an intestinal malrotation is that described by Ladd (Raphaeli *et al*., 2010; Stanfill *et al*., 2010; Williams, 2007; Zhang *et al*., 2010; Chen *et al*., 2010):

- Laparotomy and reduction of the volvulus if present
- Section of the bands occluding the duodenum
- Positioning of the small and large intestine in a position of non-rotation (duodenum to the right, cecum to the inferior left quadrant) and possible fixation of the latter when necessary
- Aappendectomy of a preventive nature, given that this organ remains in an anomalous position and could therefore constitute an evident diagnostic problem in case of subsequent pathologies regarding it

CONCLUSION

In any case, the knowledge of the clinical aspects of intestinal malrotation deserves further close examination; the discovery of this anomaly especially in the adult, requires careful evaluation of the case in order to establish exactly whether surgical intervention is appropriate. The follow-up of the case that researchers observed (which can clearly be defined as a case of partial malrotation in the atypical form low with the Treitz corresponding to its usual Level (L II) (although on the midline) is still rather short. It seems, however that the disappearance of the symptomatology suggests that the problem can probably be resolved using a simple surgical technique that does not entail substantial risks.

REFERENCES

Al-Jahdali, A., G.M. Lees, D.P. Gay and R. Al-Sairafi, 2009. Colocolic intussusception in a preterm infant with intestinal malrotation. J. Pediatr. Surg., 44: e17-e18.

Applegate, K.E., J.M. Anderson and E.C. Klatte, 2006. Intestinal malrotation in children: A problem-solving approach to the upper gastrointestinal series. Radiographics, 26: 1485-1500.

Blanco-Gonzalez, A., E. Alvarez-Hornia Perez, E. Nachon Suarez and J.C. Blanco, 2011. Intestinal malrotation in the adult. Cir. Esp., (In Press). 10.1016/j.ciresp. 2010.04.012.

Chen, W.X., J.S. Ji, H. Zhang, J.D. Zhu and L.J. Qian, 2010. Value of spiral CT in diagnosing infantile intestinal malrotation. Zhonghua Yi Xue Za Zhi, 90: 1054-1056.

Hagendoorn, J., D. Vieira-Travassos and D. van der Zee, 2011. Laparoscopic treatment of intestinal malrotation in neonates and infants: Retrospective study. Surg. Endoscopy, 25: 217-220.

Hanna, T. and J.A. Akoh, 2010. Acute presentation of intestinal malrotation in adults: A report of two cases. Ann. R. Coll. Surg. Engl., 92: W15-W18.

Maciel, LC., F.O. Salan and G.C. Guisard, 2009. Obstructions intestinal for cecal volvulus, report case. Rev. Col. Bras Cir., 36: 99-100.

Nardone, A., N. Tamini, L. Nespoli and R. Pirovano, 2010. Volvolus in an adult patient due to intestinal malrotation. Case report and review of literature. Ann. Ital. Chir., 81: 377-381.

Park, H.N., J.J. Park, J.H. Cheon, W.J. Hyung and S.H. Cho *et al*., 2010. Nonrotation of the prearterial segment of midgut presenting as duodenal obstruction in a 60-year-old man. Korean J. Gastroenterol., 55: 252-255.

Ramirez, R., K. Chaumoitre, F. Michel, F. Sabiani and T. Merrot, 2009. Intestinal obstruction in children due to isolated intestinal malrotation. Report of 11 cases. Arch. Pediatr., 16: 99-105.

Raphaeli, T., C. Parimi, K. Mattix and P.J. Javid, 2010. Acute colonic obstruction from Ladd bands: A unique complication from intestinal malrotation. J. Pediatr. Surg., 45: 630-631.

Stanfill, A.B., R.H. Pearl, K. Kalvakuri, L.J. Wallace and R.K. Vegunta, 2010. Laparoscopic Ladd's procedure: Treatment of choice for midgut malrotation in infants and children. J. Laparoendoscopic Adv. Surg. Tech., 20: 369-372.

Williams, H., 2007. Green for danger! Intestinal malrotation and volvulus. Arch. Dis. Child Educ. Pract., 92: e87-e91.

Zhang, J.X., D.M. Li and J.S. Lin, 2010. Clinical analysis of 13 cases of congenital intestinal malrotation occurring after the neonatal period. Zhongguo Dang Dai Er Ke Za Zhi, 12: 64-65.