CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 7 (2015) 134–136

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Incidental midgut malrotation detected during second laparotomy: Case report and literature review

Veli Vural[a], Mehmet Akif Türkoğlu[a,*], Gulnur Karatas[b]

[a] Department of General Surgery, Akdeniz University School of Medicine, Antalya, Turkey
[b] Private Vitale Hospital, Antalya, Turkey

ARTICLE INFO

Article history:
Received 8 July 2014
Received in revised form 3 December 2014
Accepted 3 December 2014
Available online 8 December 2014

Keywords:
Intestinal malrotation
Relaparotomy
Loop hepaticojejunostomy

ABSTRACT

INTRODUCTION: Intestinal malrotation is defined as intestinal nonrotation or incomplete rotation around superior mesenteric artery (SMA), involving anomalies of intestinal fixation as well. The patients may be recognized incidentally during other surgical procedures or at autopsy. Here in, we present a case of midgut malrotation which was diagnosed incidentally during hepaticojejunostomy procedure for benign biliary stricture.
PRESENTATION OF CASE: A 46 years old male patient was referred to our clinic with failed surgery for biliary stricture due to extensive adhesions. Prior to our surgery, intestinal malrotation was not reported and noticed by the diagnostic tools. When the patient underwent relaparotomy, midgut malrotation was observed.
DISCUSSION: Distruption in the normal embryological development of bowel is the cause of intestinal malrotation. Various anatomic configurations and anomalies resulting from rotation anomalies of midgut. Adult patients are usually asymptomatic and the anomaly is discovered only at autopsy or incidentally at surgery. The role of additional surgery especially in patients with asymptomatic disease related to malrotation is debated.
CONCLUSION: Performing loop hepaticojejunostomy with Braun enteroenterostomy is feasible and acceptable option rather than Roux-N-Y hepaticojejunostomy in case of intestinal malrotation.

1. Introduction

Intestinal malrotation is defined as intestinal nonrotation or incomplete rotation around superior mesenteric artery (SMA), involving anomalies of intestinal fixation as well [1,2]. The incidence of intestinal malrotation is 1/200–1/500 living births but it is estimated to be symptomatic in only 1/6000 live births [3–5].

Malrotation is usually symptomatic through the first twelve months of life since it is the result of abnormal embryological development. It is classically defined as a pediatric disease [6]. Acute bowel obstruction, intestinal ischemia with a midgut or cecal volvulus, chronic vague abdominal pain, duodenal obstruction and internal herniation are the presenting symptoms in symptomatic patients [7]. However in patients who do not present the acute symptoms of the disease the diagnosis is difficult. Barium studies, CT scans, angiography, and often emergent laparotomy are the several modalities described in diagnosis findings of the disease.

On the other hand, many adult patients are never diagnosed through the life time since they remain asymptomatic so these cases are recognized usually incidentally during other surgical procedures or at autopsy [8]. Here, this paper presents a case of intestinal midgut malrotation which incidentally was found during laparotomy for benign biliary stricture.

2. Presentation of case

A 46 years old male patient was referred to our clinic with the symptoms of nausea, vomiting, and abdominal pain after meals. The symptoms had started six months ago. The pain was centered over epigastrium and right subcostal regions and sometimes reflecting to back of the patients. In his medical history, he had a cholecystectomy operation performed eleven years ago due to acute pancreatitis. When the symptoms occurred, he admitted to outside hospital. Since the patient had elevated liver function test in spite of normal bilirubin and amylase levels, postoperative benign biliary stricture was suspected. Magnetic resonance cholangiopancreatography (MRCP) reported an operated gallbladder and dilated intra- and extra hepatic biliary ducts.

The choledoc was 13 mm in its largest diameter. There was a suspicious filling defect at the distal part of the choledoc. Although

* Corresponding author. Tel.: +90 5068864003; fax.: +90 2422278837.

http://dx.doi.org/10.1016/j.ijscr.2014.12.004

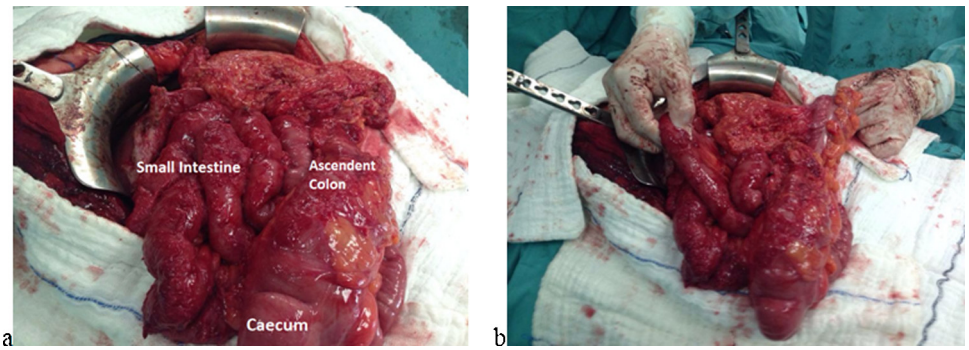

Fig. 1. The small intestines locate at the subhepatic region and the cecum and ascending colon are in the midline (a). The ileum entered the cecum from the right side (b).

several trials for performing endoscopic retrograde colangiopancreaticography (ERCP) had failed due to anatomical variations, his ultrasonographic examination and computed tomography (CT) scans did not report any pathology about the malrotation. The surgery had been planned for benign biliary stricture in outside hospital. Two trials for surgery had been failed due to extensive adhesions in the abdominal wall. The patient was then referred to our clinic. The patient underwent relaparotomy. During relaparotomy, it was observed that there were extensive adhesions between the intestinal layers and the liver. With a successful entry into the abdomen, we noticed that the small intestines were laying on the right side of the abdomen and at the subhepatic region. Hepatic and splenic flexure, the transverse colon were located at a lower location than expected and also cecum was located at the central of the abdominal cavity. Curiously, duodenojejunal junction was placed superior to the transverse mesocolon and ileum entered the cecum from the right (Fig. 1). There were no signs of acute emergencies like volvulus, ischemia, perforation or appendicitis related to intestinal malrotation.

During the operation the adhesive bands were lysed, the duodenum was mobilized. After then the choledoc was isolated from the surrounding tissues and a loop hepaticojejunostomy with a enteroenterostomy was performed. The patient was discharged smoothly 7 days after operation.

3. Discussion

Disruption in the normal embryological development of bowel is the cause of intestinal malrotation [9]. Clinical features depend on the stage of disruption and are discussed as follows. It is important to understand the normal development which will help in understanding the etiology of malrotation.

In normal embryological development, SMA is the axis around which that the normal rotation is taking place. The proximal end to the axis is the duodenojejunal loop and the distal end is cecocolic loop. The normal rotation is divided into 3 stages [9]. Both loops make a total of 270° in rotation during normal development. Both loops start in a vertical plane parallel to the SMA and end in a horizontal plane. Stage I occurs between the 5 and 10 week's of gestation. It includes extrusion of the midgut into the extraembryonic cavity, a 90° counterclockwise rotation, and return of the midgut into the fetal abdomen. Stage II occurs in 11th week of gestation completing a 270° counterclockwise rotation within the abdominal cavity. With this completion of this rotation the duodenal 'c' loop forms behind the SMA with the ascending colon to the right, the transverse colon above, and descending colon to the left. Stage III lasts from 11 weeks' gestation until term which involves the descent of the cecum to the right lower quadrant and fixation of the mesenteries. Nonrotation results from arrest in development at Stage I. Subsequently, the location of duodenojejunal junction is not at the inferior and to the left of the SMA, and the cecum is not in the right lower quadrant. The mesentery in turn forms a narrow base as the gut lengthens on the SMA without rotation, and this narrow base is prone to clockwise twisting leading to midgut volvulus. Incomplete rotation results from cessation in Stage II and is most likely results in duodenal obstruction. Obstruction is usually in the third portion of duodenum due to the compression by peritoneal bands running from misplaced cecum to the mesentery. Potential hernial pouches form when the mesentery of the right and left colon and the duodenum do not become fixed retroperitoneally. It includes incomplete fixation. İf the descending mesocolon between the inferior mesenteric vein and the posterior parietal attachment remains unfixed, the small intestine may strangulate toward unsupported area as the bowel migrates to the left upper quadrent.

Various anatomic configurations and anomalies resulting from rotation anomalies of midgut [10]. Nonrotation is the most common malrotation variant [11,12] and results when the caecal bud is the first segment to return into the abdomen and comes to rest in the left lower quadrant. Adult patients are usually asymptomatic. Many may live without any complaint, and the anomaly is discovered only at autopsy or incidentally at surgery. Some may present with chronic and unexplained abdominal discomfort and vague pain but only a small proportion patients complain of acute episodes abdominal pain.

Radiographic studies may help in the diagnosis of rotational anomaly. Plain X-ray is helpful in the diagnosis of malrotation in case of obstruction or volvulus. But the absence of cecal gas shadow with additional location of small intestinal loops predominantly in the right side should also arouse the suspicion of malrotation [13]. CT may report abnormal anatomic location of small bowel and colon, abnormal relationship of the superior mesenteric vessels, and aplasia of the uncinate process, but CT scanning is not well developed for diagnosing midgut malrotation [14,15]. CT had reported no rotational midgut anomaly in our case, supporting above findings. Ultrasonography has been shown to be very sensitive (approximately 100%) in detecting neonatal malrotation in the hands of experienced ultrasonographers. Ultrasonographic examination of our patient did not report any pathology about malrotation.

Surgery is recognized as the appropriate treatment for symptomatic disease directly related to malrotation, however, the role of surgery especially in patients with asymptomatic disease is debated [16,17]. Surgery will provide complete relief of symptoms in two-thirds of patients and partial relief in the rest [18]. When the patients are left untreated surgically, the life time risk of having emergency surgery for acute volvulus or ischemia may be up to 20%. When malrotation incidentally detected during operation,

additional surgery brings more postoperative complications thus, it is suggested that additional operation should be avoided in asymptomatic cases [17,19].

4. Conclusion

Asymptomatic patients with intestinal malrotation are usually diagnosed incidentally during surgical procedures or at autopsy. Loop hepaticojejunostomy with Braun enteroenterostomy may be applied in case of midgut malrotation due to the anatomic feasibility. Also this procedure is acceptable and safety as seen in our patient. In case of adult patients with asymptomatic intestinal malrotation observation is enough since surgical procedures do not bring additional benefits for the patient.

Conflict of interest

The authors declare that, they have no conflict of interest.

Sources of funding

Any study sponsors had no such involvement in the research.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Turkoglu MA: study design, data collections, data analysis, writing.

Vural V: study design, data collections, writing.

Karatas G: data collections, writing.

Key learning points

- Asymptomatic patients with intestinal malrotation are usually diagnosed incidentally during surgical procedures or at autopsy.
- Loop hepaticojejunostomy with Braun enteroenterostomy may be applied in case of midgut malrotation due to the anatomic feasibility.
- In case of adult patients with asymptomatic intestinal malrotation observation is enough since surgical procedures do not bring additional benefits for the patient.

References

[1] H.C. Lee, S.S. Pickard, S. Sridhar, S. Dutta, Intestinal malrotation and catastrophic volvulus in infancy, J. Emerg. Med. 43 (July) (2012) 49–51.
[2] A. Zellos, D. Zarganis, S. Ypsiladis, D. Chatzis, G. Papaioannou, C. Bartsocas, Malrotation of the intestine and chronic volvulus as a cause of protein-losing enteropathy in infancy, Pediatrics 129 (February) (2012) 515–518.
[3] B. Warner, Malrotation, in: K.T. Oldham, P.M. Colombani, R.P. Foglia (Eds.), Surgery of Infants and Children: Scientific Principles and Practice, Lippincott Williams & Wilkins, Philadelphia, 1997, p. 1229.
[4] A.V. Dilley, J. Pereira, E.C. Shi, et al., The radiologist says malrotation: does the surgeon operate? Pediatr. Surg. Int. 16 (2000) 45.
[5] C.L. Berseth, Disorders of the intestines and pancreas, in: W.H. Taeusch, R.A. Ballard (Eds.), Avery's Diseases of the Newborn, 7th ed., WB Saunders, Philadelphia, 1998, p. 918.
[6] M. Von Flüe, U. Herzog, C. Ackermann, P. Tondelli, F. Harder, Acute and chronic presentation of intestinal nonrotation in adults, Dis. Colon Rectum 37 (1994) 192–198.
[7] S.D. Hsu, J.C. Yu, S.J. Chou, H.F. Hsieh, T.H. Chang, Y.C. Liu, Midgut volvulus in an adult with congenital malrotation, Am. J. Surg. 195 (2008) 705–707.
[8] A.P. Ladd, J.A. Madra, Congenital duodenal anomalies in the adult, Arch. Surg. 136 (2011) 576–584.
[9] V. Martin, C. Shaw-Smith, Review of genetic factors in intestinal malrotation, Pediatr. Surg. Int. 26 (2010) 769–781.
[10] S.A. Kapfer, J.F. Rappold, Intestinal malrotation–not just the pediatric surgeon's problem, J. Am. Coll. Surg. 199 (2004) 628–635.
[11] A.W. Sizemore, K.Z. Rabbani, A. Ladd, K.E. Applegate, Diagnostic performance of the upper gastrointestinal series in the evaluation of children with clinically suspected malrotation, Pediatr. Radiol. 38 (2008) 518–528.
[12] W.L. Schey, J.S. Donaldson, J.R. Sty, Malrotation of bowel: variable patterns with different surgical considerations, J. Pediatr. Surg. 28 (1993) 96–101.
[13] P. Garg, M. Singh, S. Marya, Intestinal malrotation in adults, Ind. J. Gastroenterol. 10 (1991) 103–104.
[14] R. Zissin, V. Rathaus, A. Oscadchya, et al., Intestinal malrotation as an incidental finding on CT in adults, Abdom. Imaging 24 (1999) 550–555.
[15] D.M. Nichols, D.K. Li, Superior mesenteric vein rotations (a CT sign of midgut malrotation), AJR 141 (1983) 707–708.
[16] E.T. Durkin, D.P. Lund, A.F. Shaaban, M.J. Schurr, S.M. Weber, Age-related differences in diagnosis and morbidity of intestinal malrotation, J. Am. Coll. Surg. 206 (2008) 658–663.
[17] A.W. Moldrem, H. Papaconstantinou, H. Broker, S. Megison, D.R. Jeyarajah, Late presentation of intestinal malrotation: an argument for elective repair, World J. Surg. 32 (2008) 1426–1431.
[18] R.T. Maxson, P.A. Franklin, C.W. Wagner, Malrotation in the older child: surgical management, treatment, and outcome, Am. Surg. 61 (1995) 135–138.
[19] M.R. McVay, E.R. Kokoska, R.J. Jackson, S.D. Smith, The changing spectrum of intestinal malrotation: diagnosis and management, Am. J. Surg. 19 (2007) 712–717.