

Visceral variations in adult intestinal malrotation: A case-series study

Erişkin bağırsak malrotasyonunda iç organ varyasyonları: Olgu serisi çalışması

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Abstract

Aim: Intestinal malrotation (IM) is a very rare rotational anomaly of the midgut. Although uncommon, failure to diagnose IM may lead to devastating consequences. It is important that radiologists and surgeons are made aware of the diagnosis of adult IM and accompanying variations. Since IM is usually reported as a case report, it is important to note the frequency of the accompanying variations. It is aimed to present radiological findings of intestinal malrotation, increase awareness of abdominal variations and pathologies accompanying adult IM and reduce diagnostic failure.

Methods: Thirty-two patients aged over 18 years, who underwent computed tomography (CT) and received a diagnosis of IM, were retrospectively evaluated in terms of CT findings. Aplasia or hypoplasia of the uncinate process of the pancreas, focal thickening areas that may cause suspicion of a pseudomass, location of the superior mesenteric vein relative to the superior mesenteric artery, localization of the cecum, and the frequency of a redundant sigmoid colon were determined.

Results: The frequency of IM in adults undergoing CT was 1/5,375. In two of 32 cases, the cause of acute abdomen was identified while the other cases were detected incidentally. Aplasia or hypoplasia of the uncinate process of the pancreas and a redundant sigmoid colon were detected in 21 (65.6%) and 23 (71.9%) cases, respectively. The cecum was in the normal location in eight patients (25%), and there was a normal superior mesenteric artery-vein relationship in six (18.8%).

Conclusion: Intestinal malrotation is rarely encountered in adults and most cases are incidentally detected. IM is often accompanied by aplasia or hypoplasia of the uncinate process of the pancreas. A normal superior mesenteric artery-vein relationship and the cecum in the normal position are seen in one out of every four to five cases.

Keywords: Intestinal malrotation, Computed tomography, Redundant colon, Uncinate process

Öz

Amaç: İntestinal malrotasyon (IM), midgutta çok nadir görülen rotasyonel bir anomali dir. Nadiren de olsa, IM tanısı koyamamak yıkıcı sonuçlara yol açabilir. Radyologların ve cerrahların erişkin IM tanısı ve eşlik eden varyasyonlar hakkında bilgilendirilmesi önemlidir. IM genellikle bir vaka raporu olarak bildirildiğinden, eşlik eden varyasyonların sıklığını not etmek önemlidir. İntestinal malrotasyonun radyolojik bulgularını sunmak, erişkin intestinal malrotasyona eşlik eden abdominal varyasyonlara ve patolojilere farkındalığı artırmak ve tanı başarısızlığını azaltmak amaçlanmaktadır.

Yöntemler: Bilgisayarlı tomografi çekilen ve intestinal malrotasyon tanısı alan 18 yaş üzerindeki 32 olgunun BT'leri retrospektif olarak değerlendirildi. Pankreasta unsinat proses aplazisi veya hipoplazisi, psödomass şüphesi uyandırabilecek fokal kalınlaşma alanları, süperior mezenterik venin süperior mezenterik artere göre konumu, çekumun lokalizasyonu, sigmoid kolon redunsansisinin sıklığı belirlenecektir.

Bulgular: Bilgisayarlı tomografi çekilen erişkinlerde intestinal malrotasyon sıklığı 1/5375'tir. 32 olgunun 2'sinde akut batın nedeni saptanmış olup diğer olgular insidental olarak saptanmıştır. Pankreas unsinat proses aplazi veya hipoplazisi 21(%65,6) olguda ve sigmoid kolon redunsansisi 23 (%71,9) olguda rastlandı. Normal konumda çekum 8 (%25) olguda ve normal superior mesenterik arter-ven ilişkisi 6 (%18,8) olguda saptandı.

Sonuç: Erişkin intesitnal malrotasyona nadir olarak rastlanmaktadır. Karşılaşılan olguların büyük kısmına insidental olarak rastlanmaktadır. Erişkin intesitnal malrotasyona sıklıkla pankreas unsinat proses aplazi veya hipoplazisi eşlik etmektedir. Normal superior mesenterik arter-ven ilişkisi ve normal konumda çekum 4-5 olguda bir görülmektedir.

Anahtar kelimeler: İntestinal malrotasyon, Bilgisayarlı tomografi, Redundan kolon, Unsinat proses

Introduction

Any rotation of the midgut of less than 270 degrees around the superior mesenteric pedicle is defined as intestinal malrotation (IM) [1], which also presents with malfixation and predisposes patients to various conditions, such as volvulus and intestinal obstruction. In childhood, IM is often diagnosed based on the volvulus and intestinal obstruction it causes [2]. IM is rarely encountered and the true incidence of IM is not known in adults. In the diagnosis of IM, the gold standard is to see the duodenojejunal junction in abnormal localization during the imaging of the upper gastrointestinal tract [3]. However, in suspicious cases in daily practice, the superior mesenteric vein (SMV) localized on the left of the superior mesenteric artery (SMA) or vertical on ultrasound and the misplaced cecum on direct radiography suggests the presence of IM, but can also cause false positive and negative diagnoses.

A redundant sigmoid colon is defined as the presence of the sigmoid colon over bilateral iliac crests on barium colon radiographs [4]. Defined nearly a century ago, this condition has once more gained importance due to the difficulties it causes during colonoscopy [5]. In addition, it predisposes patients to sigmoid colon volvulus [6]. As in IM, malfixation is considered in a redundant sigmoid colon. Since the rotation of the duodenojejunal loop covers the pancreatic primordium, IM can also be accompanied by the hypoplasia of the uncinate process of the pancreas [7]. The coexistence of heterotaxy syndrome and IM has also been reported [8].

Adult IM is usually described in case reports [9-11]. The current study aimed to evaluate the incidence of adult IM in patients that underwent computed tomography (CT), identify the causes of concomitant acute abdomen, and the incidence of abdominal abnormalities accompanying IM.

Materials and methods

The abdominal CT images of the patients aged over 18 years were screened from the radiology archives of Adiyaman and Evliya Celebi Education and Research Hospitals between January 2013 and April 2020. Among 172,000 CT images of adults obtained from the two centers, those of 39 cases diagnosed with IM were reevaluated. Seven patients with a history of surgery were excluded from the study. The localization of the cecum, presence of a redundant sigmoid colon, location of SMV relative to SMA, the morphology of the pancreatic uncinate process were evaluated for all patients included in the study. In addition, variational conditions, such as heterotaxy and polysplenia, and other factors that could cause abdominal pain were noted. All cases were assessed by two radiologists with 10 and 11 years of experience with consensus.

Ethical statement

The study protocol was approved by Adiyaman Clinical Research Ethics Committee (Date: 01/14/2020, No: 2020/1-36), and conducted in accordance with the principles of Declaration of Helsinki.

Statistical analysis

For the statistical analyses, SPSS for Windows (IBM Corp. Released 2012. IBM SPSS Statistics for Windows, Version 21.0, Armonk, NY) was used. The continuous variables

were expressed as mean and standard deviation (SD), and the categorical variables, as frequency and percentage.

Results

In this study, IM was detected in one of every 5,375 adult cases according to the abdominal CT findings. The mean age of the cases was 45.53 (21.69) years, ranging from 18 to 89 years. Seventeen patients were male and 15 were female. While eight patients presented to the emergency department with the complaint of abdominal pain, 24 cases were incidentally detected during an abdominal CT performed for other reasons. Internal herniation, midgut volvulus, polysplenia, and heterotaxy syndrome were determined in one patient each. In the remaining 28 cases, no pathology was found. The cecum was located on the right in eight cases (25%), in the middle in 14 (43.8), and on the left in 10 (31.3), (Figure 1). A redundant sigmoid colon was found in 23 (71.9%) of cases while the remaining nine patients (28.1) had normal sigmoid colon (Figure 2). SMV was located to the left of SMA in 11 (34.4%) cases, to the immediate anterior of SMA in 15 (46.9%), and to the right of SMA in six (18.8%), (Figure 3). Aplasia and hypoplasia of the uncinate process of the pancreas were seen in 17 (53.1%) and four (12.5%) cases, respectively while uncinate process was normal in the remaining 11 (34.4%) cases (Table 1). An increase in the thickness, which gave the appearance of a pseudomass, was present in the pancreatic head in 10 cases (31.3%) and pancreatic tail in four (12.5%). In addition, 6 cases (18.8%) presented with a short pancreas.

Table 1: Localization of cecum and superior mesenteric vein and uncinate process morphology in cases with intestinal malrotation

Cecum			Localization of SMV			Uncinate process		
Right	Middle	Left	Right	Anterior	Left	Normal	Hypoplasia	Aplasia
8	14	10	6	15	11	11	4	17

SMV: Superior mesenteric vein

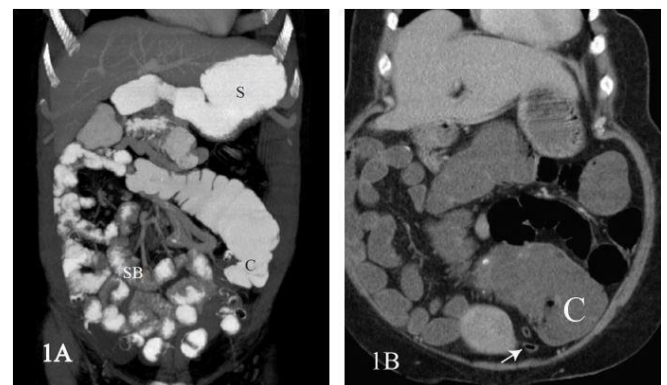


Figure 1: A: Oral and IV-contrast enhanced CT performed on a 36-year-old male patient due to abdominal pain, coronal reformatted MIP images shows cecum on the left side and small bowel loops on the right side of the abdomen, B: Abdominal non-contrast CT performed due to renal colic in a 51-year-old woman shows cecum and appendix (arrow) in the left side of the abdomen (S: stomach, SB: small bowel, C: cecum, IV: intravenous, MIP: maximum intensity projection)

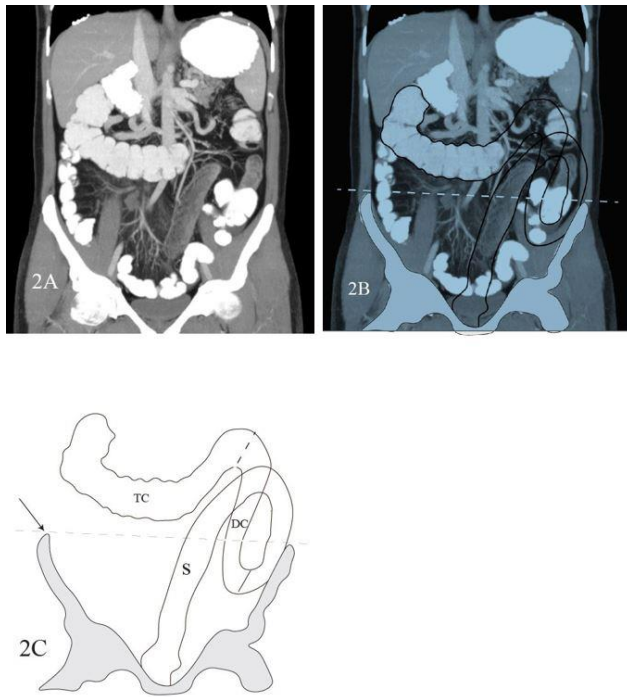


Figure 2: The coronal reformatted MIP images of a 41-year-old male patient who received oral and IV contrast enhanced abdominal CT after unsuccessful colonoscopy, the sigmoid colon (S) goes on the line passing through the bilateral iliac crests (arrow) (IV: intravenous, CT: computed tomography, MIP: maximum intensity projection, DC: descending colon, TC: transverse colon).

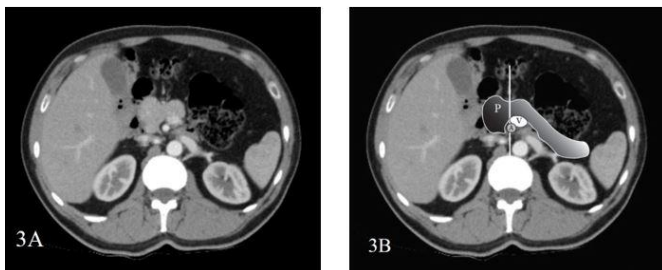


Figure 3: In contrast-enhanced abdominal CT performed to a 29-year-old male patient due to lymphoma, the SMV (V) is located to the left of the SMA (A). Uncinate process aplasia in the pancreas (P), there is no pancreatic tissue on the posterior of SMV at the left of the imaginary line crossing the right edge of the SMV.

Discussion

Intestinal malrotation is seen in one of every 200-500 births [12]. In this study, the incidence of IM among adult cases that underwent CT was one in 5,375. Of the 32 diagnosed cases, only two had acute abdomen, six had abdominal pain and the other cases that were incidentally detected. The incidence of IM was 72% in children with heterotaxy [13] while the frequency of heterotaxy in adult patients with IM is 6.2%.

It is reported that the cecum is in the right lower quadrant in 20% of cases with IM [7]. In the current study, consistent with the literature, the cecum was found in its normal localization in 25% of cases. The rate of a redundant sigmoid colon has been shown to range from 16% to 20.5% [5,14,15], and it was determined as 71.9% in the current study, which also probably suggests that the malfixation of the sigmoid colon is also a common condition accompanying IM.

The location of SMA is fixed but that of SMV changes according to the localization of the small intestine [7]. The starting point for success in diagnosing IM is the presence of an abnormal SMV-SMA relationship. However, there may be exceptions that are not few in number. In a previous study, a normal SMV-SMA relationship was found in 30% of pediatric

IM cases [16]. In the current study, a normal SMV-SMA relationship was detected in 18.8% of the adult IM cases.

Pancreatic abnormalities may be encountered in IM as a result of the close developmental relationship between the pancreas and small intestines. Aplasia or hypoplasia of the uncinata process of the pancreas has been identified in 86% of cases with IM [17]. In the current study, aplasia or hypoplasia of the pancreatic uncinata process was detected in 65.6% of the cases, which is lower than previously reported. Shape variations that may cause suspicion of a mass at the pancreatic head have been described in 35% of the general population [18]. In a study evaluating patients with IM, the contour abnormality of the pancreatic head was 48% [17]. In the current study, the contour abnormality of the pancreatic head was determined as 31.3%, and there was no significant difference compared to the values reported for the general population in the literature.

Limitations

The value of this study was limited by its retrospective design, which may have resulted in overlooking mild IM cases.

Conclusion

IM is rarely encountered in adults and most cases are incidentally detected. Adult IM is often accompanied by the aplasia or hypoplasia of the pancreatic uncinata process. A normal SMA-SMV relationship and the cecum in the normal position are seen in one out of every four to five cases.

References

- Pickhardt PJ, Bhalla S. Intestinal malrotation in adolescents and adults: spectrum of clinical and imaging features. *AJR Am J Roentgenol.* 2002;179(6):1429-35.
- Long FR, Kramer SS, Markowitz RI, Taylor GE, Liacouras CA. Intestinal malrotation in children: tutorial on radiographic diagnosis in difficult cases. *Radiology.* 1996;198(3):775-80.
- Ballesteros Gomez E, Torremade Ayats A, Duran Feljubadalo C, Martin Martinez C, Caro Tarrago A. Intestinal malrotation--volvulus: imaging findings. *Radiologia.* 2015;57(1):9-21.
- Raahave D, Christensen E, Loud FB, Knudsen LL. Correlation of bowel symptoms with colonic transit, length, and faecal load in functional faecal retention. *Dan Med Bull.* 2009;56(2):83-8.
- Kantor JL. The Common Affections of the Colon, their Origin and their Management. *Bull N Y Acad Med.* 1929;5(8):757-88.
- Valsdottir E, Marks JH. Volvulus: small bowel and colon. *Clin Colon Rectal Surg.* 2008;21(2):91-3.
- Strouse PJ. Disorders of intestinal rotation and fixation ("malrotation"). *Pediatr Radiol.* 2004;34(11):837-51.
- Cupers S, Linthout CV, Desprechins B, Rausin L, Demarche M, Seghaye MC. Heterotaxy syndrome with intestinal malrotation, polysplenia and azygos continuity. *Clin Pract.* 2018;8(1):1004.
- Bhatia S, Jain S, Singh CB, Bains L, Kaushik R, Gowda NS. Malrotation of the Gut in Adults: An Often Forgotten Entity. *Cureus.* 2018;10(3):e2313.
- Eccleston JL, Su H, Ling A, Heller T, Koh C. Gastrointestinal: Adult presentation of intestinal malrotation. *J Gastroenterol Hepatol.* 2016;31(8):1382.
- Haak BW, Bodewitz ST, Kuijper CF, de Widt-Levert LM. Intestinal malrotation and volvulus in adult life. *Int J Surg Case Rep.* 2014;5(5):259-61.
- Kotze PG, Martins J, Rocha J, Freitas C, Steckert J, Fugita E. Ladd procedure for adult intestinal malrotation: Case report. *ABCD Arq Bras Cir Dig.* 2011;24(1):89-91.
- Ryerson LM, Pharis S, Pockett C, Soni R, Fruitman D, Guleserian KJ, et al. Heterotaxy Syndrome and Intestinal Rotation Abnormalities. *Pediatrics.* 2018;142(2).
- Cuda T, Gunnarsson R, de Costa A. The correlation between diverticulosis and redundant colon. *Int J Colorectal Dis.* 2017;32(11):1603-07.
- Tan CK. Observations on some anomalies of the sigmoid colon. *Singapore Med J.* 1972;13(3):146-50.
- Menten R, Reding R, Godding V, Dumitriu D, Clapuyt P. Sonographic assessment of the retroperitoneal position of the third portion of the duodenum: an indicator of normal intestinal rotation. *Pediatr Radiol.* 2012;42(8):941-5.
- Chandra J, Grierson C, Bungay H. Normal variations in pancreatic contour are associated with intestinal malrotation and can mimic neoplasm. *Clin Radiol.* 2012;67(12):1187-92.
- Ross BA, Jeffrey RB, Jr., Mindelzun RE. Normal variations in the lateral contour of the head and neck of the pancreas mimicking neoplasm: evaluation with dual-phase helical CT. *AJR Am J Roentgenol.* 1996;166(4):799-801.

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