Case report

Abnormal retroperitoneal ileum, undscended ileucecal junction and subhepatic appendix in a macrosomatic fetus: a case report

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Abstract

In the present paper a mixed variety of malrotation of lower mid gut like retroperitoneal fixed ileum, undescended ileocecal junction, subhepatic vermiform appendix and intraperitoneal ascending colon in a macrosomatic fetus is being reported. Earlier, subhepatic appendix was reported, but the associated anomalies like retroperitoneal ilium and intraperitoneal ascending colon have not been reported. Further, ontogenic explanation of the anomalies is discussed in detail.

Keywords: malrotation, fixed ileum, undescended ileocecal junction, subhepatic appendix, macrosomatic, fetus, malrotation of lower mid gut.

1 Introduction

Intestinal malrotations are applied to an extensive developmental abnormality that may lead potentially life-threatening conditions like obstruction and necrosis (STROUSE, 2004). As a developmental variation, intestinal malrotations are associated with many other anatomic anomalies and syndromes like Hirschsprung disease and chromosomal abnormalities (JAMIESON and STRINGER, 2000; APPLEGATE, ANDERSON and KLATTE, 2006). The rate of malrotation has been reported about 1 in 500 births (STROUSE, 2004). Regarding to the consequences of malrotation, delay in proper medical intervention may led to a wide range of emergency conditions and even death (3-5%) (JAMIESON and STRINGER, 2000).Here we are going to report series of malrotation of lower mid gut in a macrosomatic fetus.

2 Case report

A mixed diversity of anomalies of lower mid gut was encountered in a macrosomatic fetus (full term, male, donated 24 hours after death, weight: 8 kg, from diabetic mother) during the dissections carried out under a project in the department of Anatomy, Bandar Abbas medicine school, Hormozgan. After exposing the abdominal cavity, we found that the terminal part of the ileum developed retroperitoneal in right flank (paracolic gutter) and made an obtuse angle about 58° with dorsal aspect of the cecum (Figures 1 and 2). Further inspection revealed that the ileocecal junction and the vermiform appendix located in subhepatic region. The appendix was attached to the right side of the cecum and covered by a peritoneal fold. This fold formed a pouch in depth of 3 cm, which was directed toward the right side in indirect relation to the right hepatoreneal pouch (Morrison Pouch) (Figure 3).Furthermore the ascending colon with diameter less than 1cm located intraperitoneally (Figure 4).

3 Discussion

Macrosomatic fetus is defined as a birth weight of at least 4000 g. It has been reported diabetes mellitus increases risk of macrosomia. In spite of improvement of ultrasonograhpic equipment, detection of macrosomia has remained as a challenge (CHAUHAN, GROBMAN, GHERMAN et al., 2005). Although we couldn't find reports on macrosomia rate in Iran, but to the best of our knowledge this is the first report dealing with a macrosomatic fetus. The result of this study showed a mixed variety of intestinal malrotation in a macrosomatic fetus, which can be considered by clinicians. Malrotations of developing gut may lead to a variety of complications like acute or chronic volvulus, and bowel necrosis (APPLEGATE, ANDERSON and KLATTE, 2006). One of the interesting finding in our case was retroperitoneal fixed ileum. Ileum is the terminal part of the small intestine and develops intraperitoneally. The terminal part of ileum joins to the right side of cecum. Normally this junction is located in right inguinal region (STANDRING, 2005). This case can be explained by referring to gut development. As the primary intestinal loop herniates into umbilicus, it also rotates around the axis of the superior mesenteric artery by 90 degree counterclockwise so that the cranial limb moves caudally and to the embryo' s right, and the caudal limb moves cranially and to the embryo's left. This rotation is complete by eight week. During 10th weeks the intestinal loop reenters the abdomen, it undergoes an additional 180 degree counterclockwise rotation. The dorsal mesentery of the ascending colon shortens and folds, causing this part of large intestine come into contact with body wall. During 11th weeks the cecum is displaced inferiorly and pulling down ascending colon (APPLEGATE, ANDERSON and KLATTE, 2006; AWAPITTAYA, PATTANA-ARUN, TANSATIT et al., 2007; LARSEN, 2001). So it can be concluded, the displacement of the ascending colon didn't occur during 11th week and it suspended by a meso in

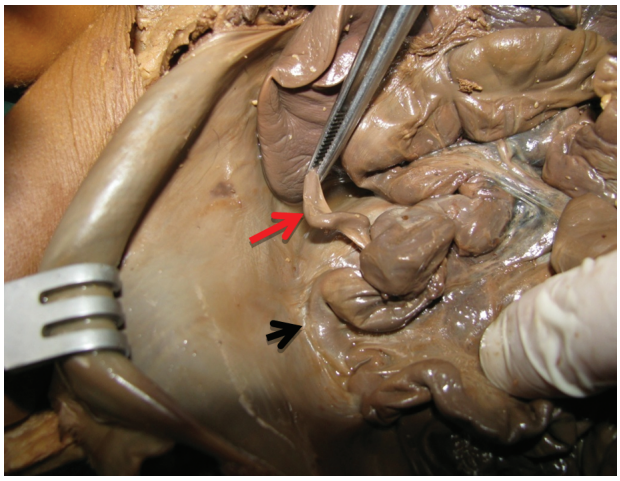

Figure 1. The vermiform appendix in subhepatic position (Red arrow), distal part of ilium lied retroperitoneally (Black arrow).

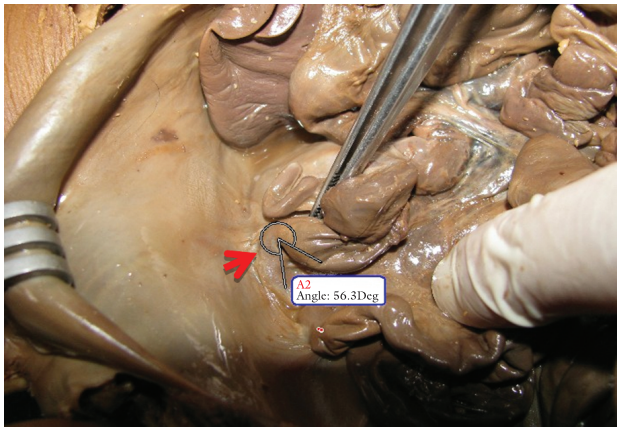

Figure 2. Iliocecal junction .the angle of iliocecal junction measured 56.3 degree (arrow).

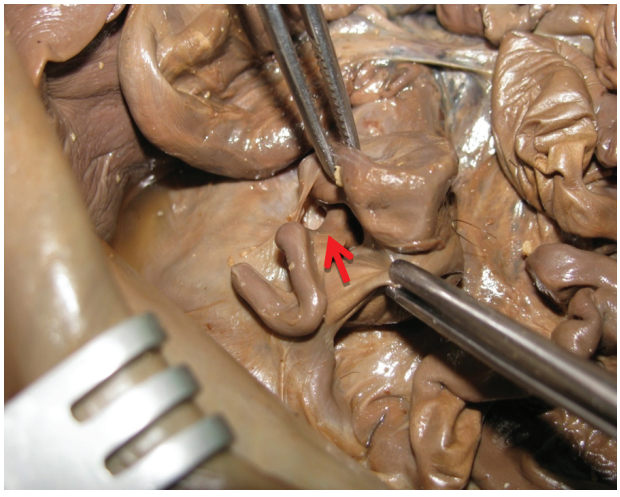

Figure 3. The vermiform appendix in peritoneal pouch, which its entrance faces laterally (arrow).

greater sac. The distal part of the ileum also, was pushed toward right lumbar fossa, as secondary retroperitoneal organ. The anatomy of the ileocecal junction (IC) and ileoceal valve concept has drawn the attention of surgeon to

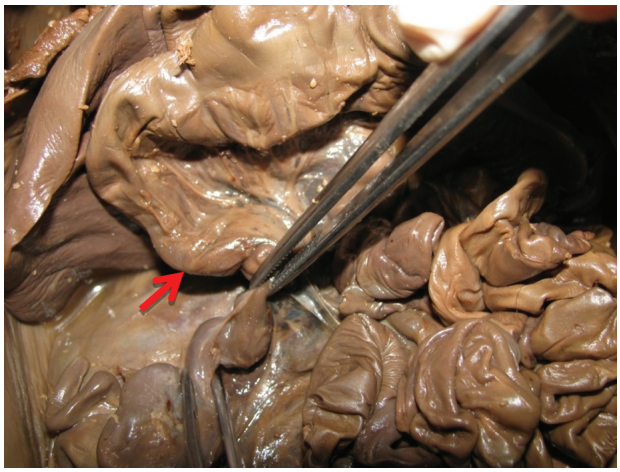

Figure 4. The ascending colon lied intraperitoneally in subhepatic region (arrow).

itself. Study has shown that terminal intussuscepted ileum into the cecum can acts as physiologic valve and sphincter (AWAPITTAYA, PATTANA-ARUN, TANSATIT et al., 2007). So any deviation from normal development may interfere with normal valve functions. Although we didn't histological examination on this part due to some limitations, but it seems histological examination of such variation could be useful. Additionally Undescended cecum in this case was associated with subhepatic vermiform appendix. The vermiform appendix is a narrow, vermian tube which arises from the posteromedial cecal wall, 2 cm below the end of the ilium (STANDRING, 2005). It is the only organ in the body that has no constant position. The various positions are retrocecal (65.3%), pelvic (31%), subcecal (2.3%), preileal lateral pouch, mesocoeliac, left-sided (associated with situs viscerum inversus), intraherniary and lumbar (1%) and postileal (0.4%). The rarer types include subhepatic,lateral pouch, mesocoeliac, left-sided (associated with situs viscerum inversus), intraherniary and lumbar appendicitis (appendix is posterior, lying against the peritoneum behind or below the caecum) (TUCKER, RASHID AL-FAGIH, EL-AMIN et al., 2002). It has been reported subhepatic appendix with frequency of 0.09% (PALANIVELU, RANGARAJAN, JOHN et al., 2007). Although subhepatic appendix position reported as a rare case it has not been pointed out to the peritoneal pouch in which appendix partially hidden. This kind of abnormal position of the appendix is of clinical and surgical importance. Inflammation of a subhepatic appendix can mimic cholecystitis and perforation of a subhepatic appendix can mimic liver abscess (KULVATUNYOU and SCHEIN 2001; PATEL, LAKSHMAN, HAYS et al., 1996). In general, although we couldn't find the relevant reports, but our little case report can be considered by surgeons and radiologist.

4 Conclusion

Based on the result of this study we recommend more study on the mechanisms by which maternal hyperglycemia may lead to developmental abnormalities like intestinal malrotations.

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