Case 723

Intestinal malrotation and volvulus - a case report of a 5 year old girl

Published on 28.01.2001

DOI: 10.1594/EURORAD/CASE.723
ISSN: 1563-4086
Section: Abdominal imaging
Imaging Technique: Ultrasound
Imaging Technique: Ultrasound-Colour Doppler
Imaging Technique: Digital radiography
Case Type: Clinical Cases
Authors: A. Malich*, S. Voigt*, S. Giggel**, U. John***, WA Kaiser*
Patient: 5 years, female

Clinical History:
Dystrophic child, severe intermittend spastic pain of the abdomen and intermittend vomiting

Imaging Findings:
A 5 years and 6 months old girl was admitted because of unclear severe acute intermittend vomiting and severe spastic abdominal pain. These intermittent symptoms were evaluated in 4 other hospitals before without a definite diagnosis. As a result of these only an intolerance versus lactosis was detected. On time of admittance the girl was without any signs of viral or bacterial infection but having acetonaemic foetor ex ore and intestinal pain. Clinical investigation did not show any signs of meningitis. In the exploration the patient’s parents reported about intermittend severe vomiting since the age of 2 years, mostly associated with severe abdominal pain, not associated with infections. During the stay at hospital the girl vomited 3-6 times biliary substances a day. The girl was severely dystrophic (14.8 kg; 112 cm; body mass index 11.8 kg/m2; <3. Percentile using normative data of german children). Ultrasound The high resolution ultrasound B-mode examination showed normal parenchymal organs of the upper abdomen. In the right and left periumbilical region mesenterical lymph nodes, oval structured, hilar perfused, and enlarged up to 14 mm were visible. In the upper right quadrant an enlarged swollen liquid-filled intestinal part, probably jejunum was visible. In the upper right quadrant an enlarged swollen liquid-filled intestinal part, probably jejunum was visible. Medial from that bowel another intestinal structure, enlarged, fluidly distended, surrounded by a dilatated distal superior mesenteric vein was visible. Bowel loops around the superior mesenteric artery (whirlpool effect) was demonstratable. There was almost no intestinal movement. The ultrasound examination revealed normal findings on structure and size but not location (left upper quadrant) of the coecum and colon and no free intraabdominal liquid. Upper G.I.series Application of Micropaque®. Fast transportation through esophagus and stomach. Normal pass of the pylorus. Jejunal parts were filled in a spiral down-descending way. The whole jejunal parts are located in the lower right abdominal quadrant. An intermittend duodenal obstruction inferior to the papilla was detected. Full intestinal contrast enhancement was severely prolonged, after 1 ½ hours. Due to overlapping contrasts it was not possible to localize the ileocoecal region accurately, which was probably located in projection on the 2nd vertebral body. The D-J flexure was inferior to the pylorus. Surgical therapy First a diagnostic laparoscopy was performed, obviously showing the volvulus. Because of frustran tries of derotation of the intestine the surgeons had to convert to an open procedure. A clockwise torsion of 480° of the bowel was found and reduced. Within the mesenterium extremely enlarged veins were visible. The Ladd’s band was divided and broadened, because Ladd’s band obstructed the lumina of the duodenum hardly and, consequently, induced the intermittet...
vomiting. Finally an appendectomy was performed.

Discussion:

Definition: Malrotation is defined as any disturbance of the physiological anteclockwise rotation of the bowel during embryonal development. Volvulus means a torsion the lumina of the bowel and in addition a strangulation of surrounded vessels, mainly caused by an inverse rotation of the bowel. Most common reason is a missed fixation and an abnormal flexibility of the intestine. Embryology / Pathology: During embryonal development, the hindgut undergoes a process of growth, lengthening and rotation. 1. Herniation into the umbilical cord along the axis of the superior mesenteric artery. 2. Rotation of 270° in an anticlockwise direction. 3. Reduction of the hindgut. 4. Fixation.

Abnormalities due to the arrest of rotation may occur at any phase and, consequently, may lead to a number of variations of malrotation (1). Three grades of malrotation are described. Grade 1: (nonrotation): 90° rotation, bowel in the right upper quadrant. Grade 2: 180° rotation without final 90° movement. Grade 3: Inverse second rotation with or without correct third 90° rotation. Malrotation often leads to obstruction of the duodenal part of the intestine and to ileus-symptoms induced by peritoneal bands, anchoring the cecum to the retroperitoneum across the duodenum in the right upper quadrant. Children having an esophageal atresia or tracheo-esophageal fistula show in 4.4% a malrotation in addition. In general the incidence rate of malrotation is given as 1 in 500 live births. the incidence of non rotation varies in different publications and is given as 1 of 6000 live births (2). Diagnosis: Typical clinical signs of malrotation are vomiting and abdominal distension (3, 4). As in our case, in most other cases the "whirlpool" sign (bowel loops around the superior mesenteric artery) is a typical sonographic feature of malrotation (89%, 5). Other sonographic signs are: duodenal dilatation with tapering configuration, fixed midline bowel, dilatation of the distal superior mesenteric vein (5). Most of these signs, especially the duodenal dilatation and the loops are visible in X-ray-based examinations precisely. However, in gastrointestinal contrast studies a false positive rate of up to 15% is described (4). Gastointestinal contrast studies are useful to support the diagnosis after an ultrasound performance. Today most institutions rely more on the upper G.i.series but contrast enema may be useful as well. A typical sign of malrotation is the location of the D-J flexure inferior to the pylorus. Other findings are volvulus, duodenal obstruction and the position of the cecum (4). Plain abdominal radiographs show a gaseous distension of the duodenum but often the appearance is nonspecific or normal. CT-examinations are rarely required in clinical setting but valuable if sonographic features are equivocal. Differential diagnosis: Regarding to clinical signs · Intussusception · Acetonaemic vomiting · Meningitis · Viral infection · Cyclic vomiting syndrome. Regarding to radiological signs · Intussusception · Appendicitis retrocoecally located. Prognosis and Treatment: The evidence supporting mandatory correction of malrotation in asymptomatic children is weak (4). Surgical treatment, however, is effective and suggested in all cases with clinical signs. In those cases, clinically inapparent, a surgical therapy should be done, however, in all children older than two years of age. Laparoscopic treatment is currently the state of the art, however, this performance is often difficult. A conversion to an open procedure is often necessary. In the literature severe complications of the malrotation are described, such as superior mesenteric venous thrombosis and acute ischemic infarction of the bowel. Pruritus, cholangitis and a secondary pancreatitis were rarely found as well as ascites, too. As a postoperative risk obstruction of the bowl was reported. After surgical therapy most patients are healthy, the recurrence rate is low (1.8%-5% (4).

Differential Diagnosis List: Chronic Volvulus and malrotation of the bowel with twisted and enlarged v. mesenterica superior causing intermittend mechanical ileus, Type 1

Final Diagnosis: Chronic Volvulus and malrotation of the bowel with twisted and enlarged v. mesenterica superior causing intermittend mechanical ileus, Type 1

References:


Figure 1

**Description:** rotated and inverted structure in the upper right quadrant of abdomen

**Origin:**
Description: bowel, rotated spirally, dilated and congested venous vessel, whirlpool sign

Origin:
Description: Jejunal intestinum, enlarged, swollen, liquid-filled, without movement, surrounded by superior mesenteric vein, Whirlpool-sign Origin:
Description: Application of Micropaque®. Fast transportation through esophagus and stomach. Normal transportation of contrast agents through the pylorus. Jejunal parts were filled in a spiral descending way. The whole jejunal parts are located in the lower right abdominal quadrant. Origin:
**Description:** Spiral down descending intestinum, atypically located, fast transportation. Duodenum dilatated with intermittent duodenal stenotic parts **Origin:**
Description: Jejunal parts were filled in a spiral down-descending way. The whole jejunal parts are located in the lower right abdominal quadrant. Due to overlapping contrasts it was not possible to localize the ileocecal region accurately, which was probably located in projection on the 2nd vertebral body. The D-J flexure was located inferior to the pylorus. Origin: