Case report

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Appendicitis associated with intestinal malrotation: imaging diagnosis features. Case report

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Abstract

Intestinal malrotation is a rare pathological situation consisting of non-rotation or incomplete rotation of the primitive intestine. Due to the abnormal caecal position inflicted by malrotation, diagnosis of acute appendicitis is difficult. Ultrasonography (US) and Computed Tomography (CT) are relevant and complementary imaging techniques for establishing an otherwise elusive diagnosis

We present the case of 54 year old male presenting with nonspecific abdominal complaints in which US (standard and contrast enhanced) and CT scans identified acute appendicitis associated with malrotated caecum and ascending colon, located in the left hipocondrum.

Keywords: malrotation, appendicitis, contrast enhanced ultrasonography

Introduction

Intestinal malrotation is a rare congenital anomaly comprising non-rotation and incomplete rotation [1] of the primitive intestinal tube around the axis of the superior mesenteric artery (SMA). In adults, the diagnosis is mostly incidental, based on investigations carried out for unrelated symptoms [2,3,4].

Due to the abnormal caecal position inflicted by malrotation, patients associating appendicitis will demonstrate atypical symptoms, with pain projected to the left of the middle line [5]. The specialist may be confronted with a diagnosis dilemma due to the patient's symptoms which mimic a wide spectrum of conditions.

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Ultrasound (US) is generally the first evaluation technique employed for acute abdominal symptoms. Contrast enhanced ultrasonography (CEUS) brings added value in establishing the differential diagnosis. Computed Tomography (CT) can provide accurate information in patients where US examination encounters limitations.

We present the case of a patient with an abnormal caecal position and associated appendicitis where the above mentioned US and CT procedures played an important role in complementary imaging techniques for establishing an otherwise elusive diagnosis.

Case report

A 54 year old patient with aortic valve prosthesis, arterial hypertension and chronic obliterative arteriopathy of the inferior right limb, was admitted with diffuse abdominal pain in the left hipocondrum, altered intestinal transit, and loss of appetite. No previous surgical interventions on the abdomen were noted. The patient was under oral anticoagulant and anti-hypertension medication.

The physical examination revealed tenderness over the left upper quadrant and distended abdomen. The

white blood cell count was 14.700/mm^3 with predominance of neutrophils (76.2%). No anemia or renal insufficiency was noted. The laboratory results indicated a proper anticoagulant treatment, with INR value of 3.09.

The US examination (Logiq 7; General Electric, Milwaukee, WI, USA) depicted a bowel conglomerate at the left hipochondrum, with inflammatory hyperemia at the color Doppler investigation. Furthermore a fluid filled distended small gut loop was identified (fig 1). Characteristic stenotic alterations were noted. No free liquid was visible between the intestinal segments or in the pelvis.

CEUS in pulse-inversion mode at a mechanical index of 0.09, after 1.6 mL bolus injection of the contrast agent SonoVue (Bracco, Milano, Italy) of the intestinal segment in the left upper quadrant, depicted transparietal contrast enhancement of the enlarged wall in the arterial phase. Peri-intestinal marked epiploon congestion was also noted (fig 2).

The plain abdominal radioscopy pointed to the small bowel air-liquid levels revealed in the right flank and periumbilical region.

Native and i.v. contrast enhanced CT scans confirmed the presence of small bowel inflammatory segments in the left upper quadrant. The distended small bowel loop presented a slightly enlarged and iodophile wall. The adjacent peritoneal fat's edematous infiltration was noted. The CT described caecum and ascending colon present in the left upper quadrant and furthermore identified a thickened and inflamed appendix with coprolite images in the caecum and the appendicular lumen. Examination of the superior mesenteric vein (SMV) and artery (SMA) revealed a reversed position, with the artery present at the right side of the vein (fig 3).

These findings were consistent with acute appendicitis accompanied by suboclusive alterations of the small bowel, superimposed on the malrotated gut.

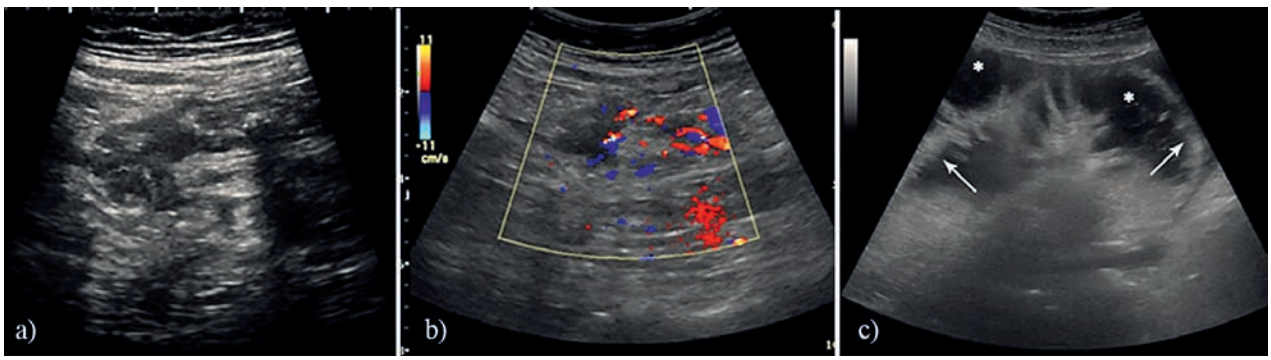


Fig 1. Ultrasonographic examination a) Gray scale ultrasound depicting bowel conglomerate; b) Color Doppler imaging indicating hyperemia; c) Gray scale ultrasound of distended gut. Asterisk in distended lumen, arrows indicating wall.

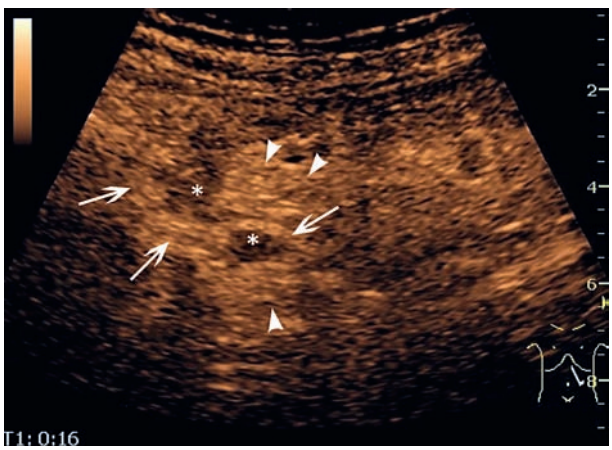


Fig 2. CEUS of inflamatory segments. Arterial phase. Asterisk in lumen. Arrows pointing towards gut wall enhancement. Arrowheads pointing towards peri-intestinal epiploon enhancement

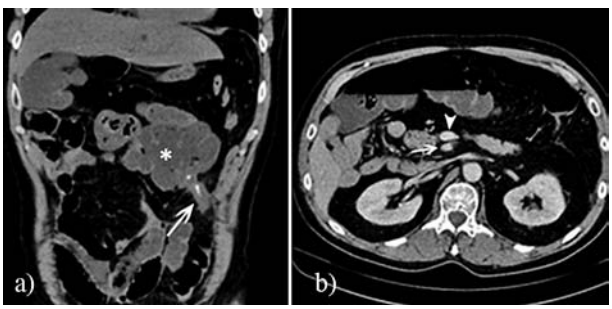


Fig 3. CT examination a) Native CT Coronal reconstruction. Left upper quadrant caecum and modified appendix with coprolite. Arrow pointing at appendix. Asterisk in caecum lumen. b) i.v. contrast enhanced CT venous phase, transvers scan. SMA (arrow) located to the right of the SMV (arrow head)

Based on the US, CEUS and CT results, the patient was scheduled for a surgical direct appendicectomy. The operation was delayed until safe coagulation parameters were achieved. Thus the oral anticoagulant was suppressed and the patient was switched on subcutaneous low molecular weight heparin injections. The supportive therapy comprised of large spectrum antibiotics and antalgics. Careful monitoring of the patient and repeated ultrasound examinations were carried out.

The patient's symptoms easily improved under the supportive therapy, but US follow up noted the appearance of free fluid between small bowel loops.

During surgical procedure an inflammatory conglomerate was identified in the left upper quadrant, consisting of small bowel loops, epiploon, caecum and ascending colon. The macroscopically appearance of the appendix was characteristic for gangrenous alteration, perforation and consolidation by small bowel loops. The position and particular mesocolon of the caecum and ascending colon confirmed the diagnosis of gut malrotation. A direct appendectomy was consequently performed. The patient's evolution was favorable and he was discharged five days later. The pathological examination of the appendix confirmed the gangrenous appendicitis and associated perforation.

During the follow up, the patient's evolution complicated with pseudomembranous colitis, diagnosed by colonoscopy with biopsies. The US examination and the CT scan showed features of extended pancolitis. Under antibiotic treatment the evolution was favorable.

Discussions

It is estimated that rotation anomalies of the primitive intestine appear in one of every 500 newborns [6]. Eighty percent of the cases are symptomatic in the first two weeks of life.

The right colon and the small intestine are most frequently affected by development anomalies. Rotation may be incomplete, with caecum and ascending colon positioned at the left of the midline but with a normal positioned transverse colon. These particular settings often coexist with fixation deficits, creating the foundation for possible future volvulus [7].

Generally, adult patients with malrotation can exhibit three different clinical presentations [8]. Some patients will express symptoms and signs of acute obstruction, while others will relate to chronic abdominal manifestations such as pain and intermittent obstruction. In contrast with these groups, another category can complain of atypical symptoms, not related to intestinal malrotation as in the case of the above presented patient.

Transabdominal ultrasound is a widely available technique, which frequently is the first imaging examination in emergency situations [9]. Both color and power Doppler ultrasound are applied to demonstrate intramural vessels in the bowel wall and to detect hypo- or hypervascularisation [10,11].

The primary criterion for sonographic diagnosis of appendicitis is a non-compressible, 6 mm, blind-ended, tubular structure at the base of the cecum with absent peristalsis [12-14]. Secondary signs include circumferential hyperemia in transverse section by color-flow Doppler, peri-appendiceal fluid collections, visible appendicolith, and interruption of the echogenic submucosa (indicating perforation) [15,16]. An ovoid shape in the transverse incidence along the appendiceal length can reliably exclude appendicitis. Using these criteria, reports have evidenced sensitivities of 75 to 90% and specificities of 86 to 100%, with positive predictive values of 91 to 94%. [17].

New sonographic technologies combined with the application of contrast agents allow the monitoring of intramural perfusion employing a low mechanical index without Doppler mode (contrast harmonic imaging) [18]. CEUS can bring valuable information on the localization and extent of inflammation, bowel wall thickening, intramural and also peri-intestinal inflammatory reaction. Moreover, the technique is successful in identifying intestinal fistulas, abscesses or ileus [19].

The main usefulness of CEUS resides in the diagnosis of intestinal ischemia. Diminution or absence of enhancement in the bowel wall are highly suggestive for intestinal ischemia. Hamada et al. reported the sensitivity of this method at 94.1% and the specificity at 100% [20]. In the present case, CEUS was able to accurately exclude intestinal ischemia.

The patient's atypical localization of the appendix and superimposed inflammatory gut segments constituted a drawback in the establishing of acute appendicitis.

Indirect signs of an acute abdominal process were revealed on standard B mode and Doppler scans. CEUS added superior diagnostic quality to the examination, in terms of process localisation, intestinal wall alteration and periintestinal affects and ruled out conditions as intestinal ischemia, inflammatory bowel disease, and ulcerative colitis. As a consequence US and CEUS managed to identify an acute abdominal condition and contributed to narrowing the spectrum of otherwise invaluable and possibly invasive examinations.

Thus CT scan became the examination of choice in the present case and revealed the congenital gut malrotation associated with acute appendicitis.

The most extensive study upon left sided appendicitis

is attributed to Sami et al [21] who reviewed 64 reports regarding 95 cases. Incidence of acute appendicitis associated with midgut malrotation was reported at 24.2% and pain localization in the left upper quadrant was as rare as 7.3%. As in the current presentation, the study concluded that because the appendix is located abnormally, acute appendicitis represents a diagnostic dilemma and that US and CT can provide quite useful information.

We found no report about the use of CEUS in similar cases. In our opinion, CEUS provides relevant information and brings a significant contribution in ruling out other concurrent conditions in patients with intestinal complaints.

Conclusions

The abnormal caecal position inflicted by intestinal malrotation may give atypical symptoms in cases of associated appendicitis making the diagnosis very difficult. Many imaging techniques may be necessary for assessment of the diagnosis.

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