A diagnostic dilemma: Left-sided appendicitis in a 10 year old boy with previously undiagnosed intestinal malrotation. A case report

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ABSTRACT

INTRODUCTION: Intestinal malrotation is a congenital rotational anomaly that occurs as a result of an arrest of normal rotation of the embryonic gut, said to occur in 1 in 6000 live births. Due to the abnormal caecal and appendix position, diagnosis of acute appendicitis becomes more challenging, thus leading to diagnostic and operative intervention delays. Our aim is to highlight the diagnostic challenges in this clinical scenario.

PRESENTATION OF CASE: We present a case of a 10 year old boy with previously undiagnosed intestinal malrotation with a left sided acute appendicitis. Initial symptoms lead to a treatment for gastroenteritis, however, due to ongoing pain a CT abdomen was done which showed the malrotation and appendicitis. He required a laparoscopy converted to open appendicectomy due to an appendiceal mass.

DISCUSSION: Historically, intestinal malrotation was thought to be a disease of infancy with infrequent occurrence after the age of one year. However, recent analysis has shown an increase in presentations after one year of life into adulthood. Thus, the prevalence of malrotation in children and adults over the age of one year appear to be higher than initially presumed.

CONCLUSION: Left sided acute appendicitis is a diagnostic dilemma, thus often leading to management delays. It is pertinent to remember that malrotation of the gut is more common than previously thought, and not just a disease of infancy. It is advisable to consider imaging studies while balancing the risk-benefit ratio of radiation exposure, especially in paediatric cases to cinch the diagnosis.

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1. Introduction

Acute appendicitis continues to be a difficult condition to diagnose and has a high risk of missed and delayed diagnosis. Left sided appendicitis therefore poses an even greater diagnostic conundrum [1]. Majority of cases with left sided appendicitis have an associated midgut malrotation, situs inversus totalis or an abnormal position of a long appendix. Due to the ambiguity of symptoms, diagnosis is often challenging and thus may lead to an increase in morbidity and mortality.

Intestinal malrotation is a rare congenital abnormality and symptomatic cases are estimated to occur in 1 in 6000 live births [2]. Historically, intestinal malrotation was considered primarily a disease of infancy with infrequent occurrence past one year of life. However, that view is changing with increased documented presentations above the age of one year [3,4].

We present a case of a 10 year old boy with previously undiagnosed intestinal malrotation with a left sided acute appendicitis and aim to highlight the diagnostic challenges faced.

2. Case presentation

A 10 year old boy was brought into the Caboolture Hospital Emergency Department at 2 pm with a 24 hour history of abdominal pain and vomiting. The pain was of gradual onset, in the left lower quadrant, commencing after his afternoon meal. He had 4 episodes of vomiting prior to presentation with no change in his bowel movement, no fevers and no urinary symptoms. His childhood immunizations were up to date. He had a past medical history of asthma which was well controlled with salbutamol inhalers when required.

On review, he was noted to be afebrile, BP 138/78 mmHg and HR of 113/min. Abdominal examination revealed a soft abdomen, with tenderness localized to the left lower quadrant. There was no flank tenderness. Respiratory, cardiovascular and genital examination were normal. Urine dipstick was noted to have moderate blood without leucocytes or nitrates.

Full blood examination revealed a Hb of 14.0 g/dL, Hct of 41%, WCC of 12 × 10⁹/L, platelet count of 260 × 10⁹/L. Urea, Electrolytes and creatinine were within normal range. C- Reactive Protein (CRP) was markedly elevated at 126 mg/L. On Ultrasound of the abdomen, the appendix could not be visualized, both kidneys appeared normal, there was no intra-abdominal free fluid noted and both testes were normal.

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weeks post op and made a good recovery. Histology of the appendix revealed a perforated acute suppurative appendicitis.

4. Discussion

Intestinal malrotation is a congenital rotational anomaly that occurs as a result of an arrest of normal rotation of the embryonic gut. During normal abdominal development, the foregut, midgut and hindgut herniate out from the abdominal cavity, where they undergo a 270 degree counter clockwise rotation around the superior mesenteric vessels. Following the rotation, the bowels return into the abdominal cavity, with fixation of the duodenojejunal loop to the left of the midline and the cecum in the right lower quadrant. Intestinal malrotation refers to any variant in this rotation and fixation of the GI tract during fetal development.

Historically, intestinal malrotation was thought to be a disease of infancy with infrequent occurrence after the age of one year. However, recent analysis has shown an increase in presentations after one year of life into adulthood. Thus, the prevalence of malrotation in children and adults over the age of one year appear to be higher than initially presumed.

Due to the rarity of cases and ambiguity of presenting complaints, often times the diagnosis of a left sided appendicitis is challenging and delayed. This child presented with abdominal pain and vomiting. He was of school going age and the initial presumption of a gastroenteritis is a sound differential. Also, the presence of a false dipstick hematuria and an elevated blood pressure during his Emergency Department presentation, also added a level of complexity to the clinical picture.

Another element that delayed diagnosis was the decision to proceed with a repeat ultrasound abdomen instead of a computed tomography scan during the second day of admission. Radiation exposure in the paediatric group requires careful consideration given that there is a probable increased risk of future carcinogenesis [5,6]. Radiation was limited to a low dose CT scan in this patient. Imaging studies are important to diagnose atypical cases such as this one and it is pertinent to know the options available [7]. The most extensive study on left sided acute appendicitis reviewed 64 reports regarding 95 cases. Incidence of acute appendicitis associated with midgut malrotation was reported at 24.2% and incidence with situs inversus totalis was 69.4%. The study concluded that because the appendix is located abnormally, acute appendicitis represents a diagnostic dilemma and that ultrasound and computed tomography can provide useful information [8,9].

Other differentials that need to be considered in the paediatric age group are groin hernias or testicular torsion with referred pain, constipation, mesenteric adenitis, urinary tract infection, Meckel’s diverticulum, bowel obstructions possibly secondary to volvulus or intussusception, undescended testes, diabetic ketoacidosis, rare causes such as epiploic appendagitis [10,11]. Henoch-Schönlein

On the basis on ongoing abdominal pain and elevated inflammatory markers, he was referred to the surgical team. Surgical team reviewed the patient in the emergency department and subsequently referred the case to the paediatric team in view of haematuria and an elevated BP on the basis that there could be an underlying glomerulonephritis. Formal urine microscopy showed no haematuria or proteinuria. He was therefore admitted to the paediatric ward for observation.

Over the next 24 hours, he had ongoing abdominal pain and started developing watery diarrhoea. Repeat blood investigations showed an increase in inflammatory markers, with a WCC of \(24.4 \times 10^9/L\) and a CRP of \(206\, \text{mg/L}\). He was commenced on IV antibiotics. At this stage, a repeat Ultrasound was ordered which revealed a dilated tubular structure in the left iliac fossa with a small amount of free fluid around the tip. This tubular structure could not be followed to the caecum. It was at this stage that a low dose CT abdomen was ordered which revealed a severely inflamed appendix that was significantly dilated at 2.06 cm in the left iliac fossa. It demonstrated a 9.7 mm appendicolith and a smaller 5 mm appendicolith. Intestinal malrotation was noted with the small bowel in the right abdomen, the large bowel in the left and an inversion of the normal Superior Mesenteric Artery/Superior Mesenteric Vein relationship Figs. 1–3.

3. Outcomes and follow up

He was referred to the surgical team and underwent a laparoscopy converted to Laparotomy and Appendicectomy. Intraoperative findings showed an appendicular mass with omentum surrounding the appendix. His caecum was in the left upper quadrant, with a long appendix that was perforated near the base. There was an associated localized pus collection.

He was in hospital for a further 4 days for IV Ceftriaxone and IV Metronidazole. He was discharged on Day 4 post operatively with a course of oral antibiotics. He was seen in the outpatient clinic 2
5. Conclusion

Left sided acute appendicitis is a diagnostic dilemma, thus often leading to management delays. It is pertinent to remember that malrotation of the gut is more common than previously thought, and not just a disease of infancy. Other differentials should also be considered due to ambiguity of presenting symptoms. It is advisable to consider imaging studies while balancing the risk-benefit-ratio of radiation exposure, especially in paediatric cases to cinch the diagnosis.

Conflict of interest

None.

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None

Ethical approval

No ethical approval was required or requested for this case report.

Informed consent

Verbal consent was obtained from patient’s father.

Author Contributions

Ashvini Shekhar—Case report author, literature review, correspondence author.

Rasika Hendahewa—Involved in the care of patient, secondary author, editor, supervisor

Guarantor

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