CASE REPORT

ACUTE APPENDICITIS: AN ATYPICAL PRESENTATION AS LEFT LOWER QUADRANT PAIN IN SETTING OF UNDIAGNOSED INTESTINAL MALROTATION WITH ASSOCIATED VASCULAR ANOMALIES AND LEFT ISOMERISM

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PJR October - December 2022; 32(4): 249-253

ABSTRACT

Acute appendicitis is one of the most frequent causes of acute right lower quadrant abdominal pain requiring emergency surgical intervention. Clinical examination and routine blood tests are sufficient for a provisional diagnosis, with CT scan as the most sensitive modality for diagnostic confirmation. Rare instances of appendicitis presenting as epigastric or left quadrant pain in cases of undiagnosed intestinal malrotation particularly in adults may present as a diagnostic dilemma and may at times even progress to catastrophic outcome. We report a case of 39 years old male patient with undiagnosed intestinal malrotation presenting with a recent history of left lower quadrant pain where acute appendicitis and associated left isomerism and aberrant vascular anomalieswere ultimately grasped on the abdominal CT examination.

Keywords: Acute Appendicitis, Intestinal Malrotation, Left Isomerism

Case Report

A 39 years old male patient was referred for an abdominal CT examination with a recent history of left lower quadrant pain which remained unresponsive to the routine gastritis treatment. Abdominal CT was performed with intravenous contrast in the portal venous phase. Examination showed differentially rotated small and large bowel segments with most of the small bowel clustered in righthemi abdomen, colonic segments clustered in left hemi abdomen and transverse colon placed vertically in the midline. An acutely inflamed appendix was appreciated lying in the midline in umbilical / hypogastric region with its tip directed upwards. There was loss of appendiceal intraluminal gas along with luminal distension and wall enhancement. Short axis dimension of inflamed appendix measured up to 10 mm. Significant peri-appendiceal mesenteric fat stranding was also appreciated signifying the inflammatory process. Enlarged adjacent mesenteric lymph nodes were visualized measuring up to 11 mm.

In addition the CT examination highlighted a spectrum of associated anomalies. Heterotaxy variant; left isomerism was appreciated in the form of polysplenia, comprising of multiple splenulesalong with a parent spleen.

Aberrant hepatic portal venous anatomy was also noted which was interpreted as preduodenal portal variant where an engorged branch of left portal vein was seen draining directly into the superior mesenteric vein. Further, its course was anomalous; taking a

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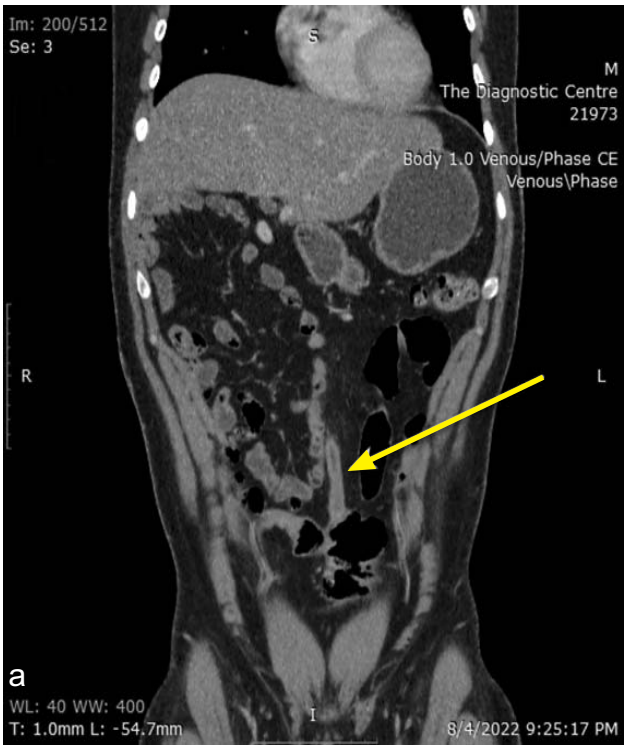
Submitted 17 October 2022, Accepted 8 November 2022

more extra capsular route anterior to both the duodenum and head of pancreas. Additionally the coeliac axis was absent and the splenic artery was taking direct origin from the aorta at the expected site of coeliac trunk. Superior mesenteric artery appeared hyperplastic with a dimunitive hepatic artery taking its origin from gastroduodenal artery.

Given these radiological findings, diagnosis of intestinal malrotation complicated by acute appendicitis in setting of associated spectrum of anomalies was inferred on the report. Primary surgeon was telephonically informed for the needs of an urgent intervention at the time of interpretation. Surgery was subsequently performed three days later via laparoscopic approach and histopathology confirmed acute appendicitis with periappendiceal inflammation and one reactive regional lymph node. Clinical follow-ups ascertained stable condition of the patient.



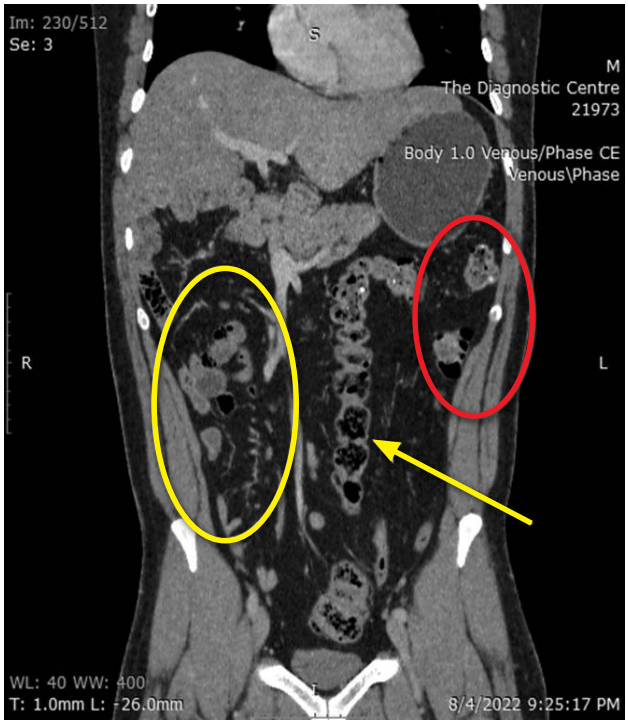
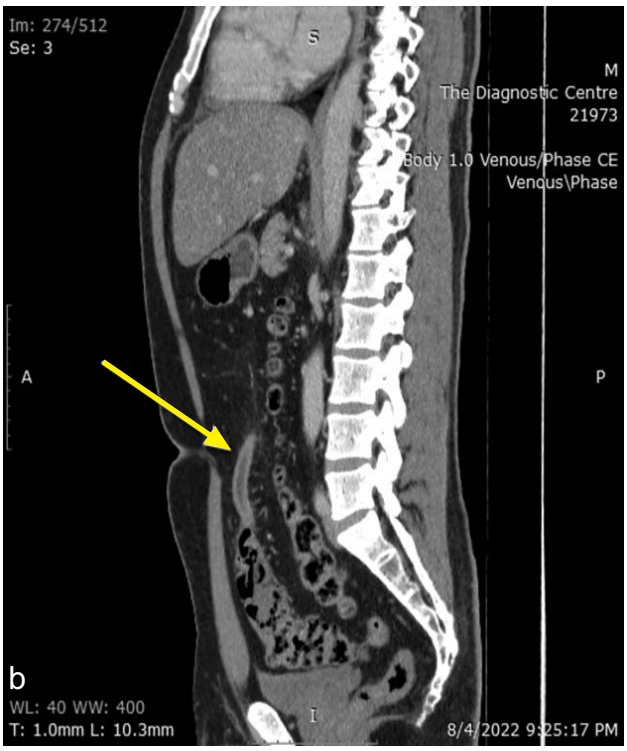


Figure 1: Coronal image of IV contrast-enhanced abdominopelvic CT demonstrates differentially rotated small and large bowel segments with most of the small bowel clustered in right hemi abdomen (yellow circle), colonic segments clustered in left hemi abdomen (red circle) and transverse colon placed vertically in the midline (yellow arrow).



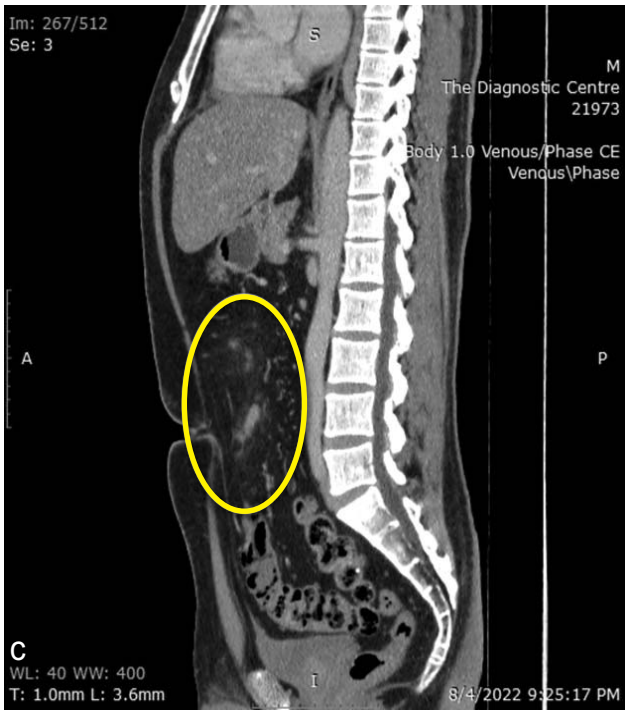
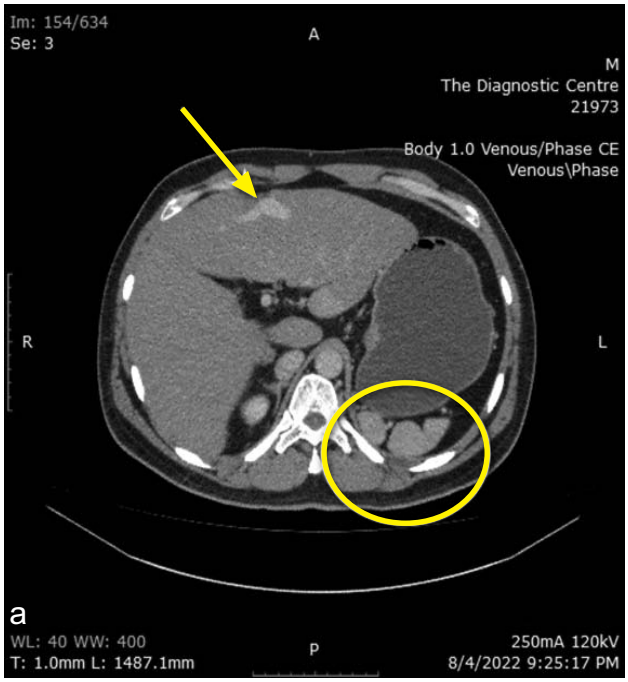


Figure 2: Coronal **(2a)** and sagittal **(2b, 2c)** images of IV contrast-enhanced abdominopelvic CT demonstrates an acutely inflamed appendix lying in the midline in umbilical/ hypogastric region with its tip directed upwards (yellow arrow 2a, 2b). Periappendiceal mesenteric fat stranding and enlarged adjacent mesenteric lymph nodes also appreciated (yellow circle 2c).



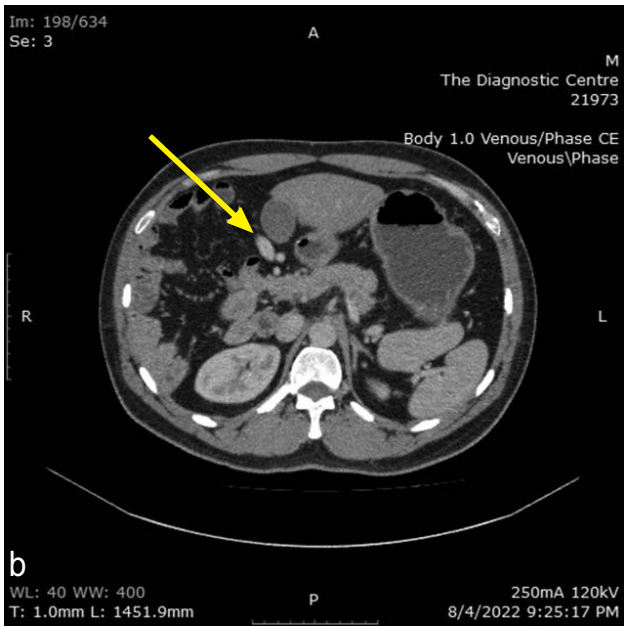


Figure 3: Axial images **(3a, 3b)** of IV contrast-enhanced abdominopelvic CT demonstrates splenules along with a parent spleen (yellow circle in 3a). Preduodenal portal vein variant also appreciated, taking an anomalous extra capsular route anterior to both the duodenum and head of pancreas (yellow arrows in 3a, 3b).

Discussion

Acute appendicitis is the acute inflammation of the vermiform appendix, a commonly encountered entity in routine clinical practice and one of the chief indications for immediate abdominal surgery in young patients. CT is the most sensitive modality to detect appendicitis.[1,9,10] The usual presentation is pain in the periumbilical and right lower quadrants with associated constitutional symptoms such as fever, nausea and vomiting. Physical examination reveals right lower abdominal tenderness, rebound tenderness, guarding and rigidity.[2,9] Routine labs show leukocytosis and raised inflammatory markers. The overall clinical and lab findings with a complimentary ultrasound scan are sufficient for the surgeons to proceed with an emergency surgical approach.[2]

Acute appendicitis at times may become difficult to diagnose because of an unusual clinical presentation secondary to atypical location of the appendix. Such anatomical variants may include intra pelvic or extra-peritoneal locations, herniated appendix in a strangulated femoral hernia or else the appendix may altogether be located in the left quadrants in cases

of associated intestinal malrotation, situs inversusor heterotaxy syndrome.[1,2,9]

Intestinal malrotation is a congenital anomaly that results from an abnormal rotation of the gut as it returns to the abdominal cavity during embryogenesis. During normal process of embryogenesis the bowel herniates into the base of the umbilical cord. On its returns to the abdominal cavity it undergoes a complex 270 degree counter-clockwise rotation resulting in the duodenojejunal flexure being typically located left of the midline, at the level of the L1 vertebral body, and the terminal ileum being located in the right iliac fossa.[6] In cases of intestinal malrotation there is a congenital anomalous rotation of the gut around the superior mesenteric artery axis with a resultant misplacement of the duodenojejunal junction to the right midline, right-sided location of the small bowel and left-sided location of the colon with the inversion of SMA/SMV relationship.[1,2,6] Individuals may live their entire lives with a malrotated bowel morphology without any associated symptoms, yet the abnormality does predispose to potential life-threatening complications such as acute appendicitis, midgut volvulus and obstructed internal hernias.[1,3,8,10]

Heterotaxy syndrome or situs ambiguusis described as an incomplete lateralization disorder that results in variable arrangement of the viscera across the left-right axis that is markedly different from their normal position and is considered as an entity separate form situs inversus which simply involves complete mirror imaging.[1,2,3,7,8] Left isomerism is a variant of heterotaxy syndrome comprising of polysplenia which by extension refers to the presence of multiple uniform sized splenules with either the absence or presence of a parent spleen.[3,7] A well-known association of intestinal malrotation exists with left isomerism with an estimated prevalence of up to 70 %.[1,3] Other anomalous conditions associated with left isomerism include bilobed lungs, midline liver, partial dorsal pancreatic agenesis or short pancreas , abnormalities of the portal vein formation and azygous or hemiazygos continuation of the IVC.[3,4,6,7]

In our case of a 39 year old male referred for an abdominal CT, presenting with a recent left lower quadrant pain, acute appendicitis was diagnosed in setting of congenital intestinal malrotation. The typical CT findings of acute appendicitis include inflamed thickened enhancing walls of the appendix with either an obliterated or else an oral contrast opacified lumen with theluminal diameter averaging more than 6 mm.[1,2,10] Regional mesenteric fat stranding and locoregional lymphadenopathy are associated features. In our case associated polysplenia was also present, along with congenital abdominal arterial and portal venous anomalies. The portal venous anomaly was narrowed down as the preduodenal portal vein variant, which passes ventral to the duodenum and the head of the pancreas.[3,5,7] However not all characteristic associations of the left isomerism syndrome, as mentioned previously in the literaturewere present in our case. Such rare syndromes usually remain asymptomatic are often discovered incidentally in patients imaged or operated for unrelated reasons.[3] Nevertheless, radiologists should be well acquainted with such patterns of anatomical variationswhilst remaining cautious for any superimposed critical complications, namely appendicitis or volvulus.[1]

Conclusion

This case highlights a crucial fact that the typical clinical presentation of acute appendicitis may be altered in setting of an undiagnosed intestinal malrotation, leading to a diagnostic dilemma and delay in the appropriate management. Intestinal malrotation remains a rare yet vital cause of left-sided acute appendicitis. Other associated congenital anomalies including polysplenia and vascular anomalies also remain asymptomatic and are mostly detected incidentally when patients are being evaluated for more obvious reasons. CT scan serves as a crucial problem solving imaging modality in such situations to ascertain the actual diagnosis. Radiologists must be well aware of the existence of such variational anomalies and must be able readily recognize them on imaging to avoid delay in management or surgical complications. This case happens to suggest a new diagnostic approach for referring physicians and radiologist altogether, with emphasis on the need for abdominal CT examinations in setting of confusing clinical picture yet with raised inflammatory markers.

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