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CASE REPORT

Appendiceal intussusception presenting as a caecal mass

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Abstract

Introduction: The differential diagnosis of caecal mass is broad and the inclusion of appendiceal pathologies is an important element. *Case Report:* We report a 37-year-old woman with recurrent right iliac fossa pain. Computed tomography scan revealed a caecal mass suggesting complete inversion or intussusception of the appendix, which was confirmed by pathologic microscopic examination. This case report discusses appendiceal intussusception with emphasis on diagnosis and treatment options. *Discussion:* Appendiceal intussusception is a rare entity and the complete type typically presents as a polypoid lesion located at the appendiceal orifice in the caecum. It is imperative to include this entity in the differential diagnosis of caecal mass, especially during colonoscopy, as the removal of this polypoid lesion can result in a devastating caecal perforation or haemorrhage.

Keywords: Appendix, caecum, intussusception, appendiceal intussusception.

INTRODUCTION

Intussusception of the appendix is a rare condition with a reported incidence of 0.01%.[1] It is more common in adults, with a female predilection[2] and in patients with cystic fibrosis.[3] Clinical manifestations vary, from asymptomatic patients to patients with acute abdomen that mimics acute appendicitis. However, chronic and intermittent abdominal symptoms are the most common clinical scenarios.[1] Due to its rarity and vague presentation, this condition often creates a diagnostic dilemma for radiologists,[1] but when the diagnosis is considered, computed tomography (CT) scan and ultrasound are the modalities of choice.[4-7] Complete inversion or intussusception can mimic a polyp endoscopically.[3,8,9] Therefore, gastroenterologists and surgeons should be aware of this condition as simple polypectomy carries a high risk of perforation and bleeding.[9]

CASE REPORT

The patient was a 37-year-old female who presented with recurrent right iliac fossa pain. She was known to have hypertension and diabetes mellitus (on insulin and an angiotensin-converting enzyme inhibitor), with no relevant surgical or family history. The patient had numerous hospitalisations in the past for intractable nausea, vomiting and right lower quadrant abdominal pain and was diagnosed with gastroparesis secondary to diabetic neuropathy.

On physical examination, the abdomen was soft and bowel sounds were normal. She exhibited no abdominal distension or palpable masses. There was mild right lower quadrant tenderness without rebound tenderness or guarding.

CT scan showed a few prominent right lower quadrant mesenteric lymph nodes. Pelvic organs were normal in size and appearance. There was no evidence of bowel obstruction, pneumoperitoneum, or calcific or obstructive uropathy. The appendix was not visualised. However, a 10 mm mass arising from the wall of the caecum and at the base of appendix was found (FIG. 1). Therefore, appendiceal inversion or intussusception was suspected and the possibility of accompanying mucinous neoplasm or mucocele could not be ruled out.

As the exact aetiology of her pain and nature of this mass was still unclear, the patient was sent for colonoscopy. In the caecum, a lesion was noted arising from the caecal wall with intact overlying mucosa. It was soft and mobile, consistent with what appeared to be a submucosal polyp (FIG. 2). No other evidence of abnormalities was noted.

Subsequently, the patient underwent robotic ileocecectomy and the macroscopic examination

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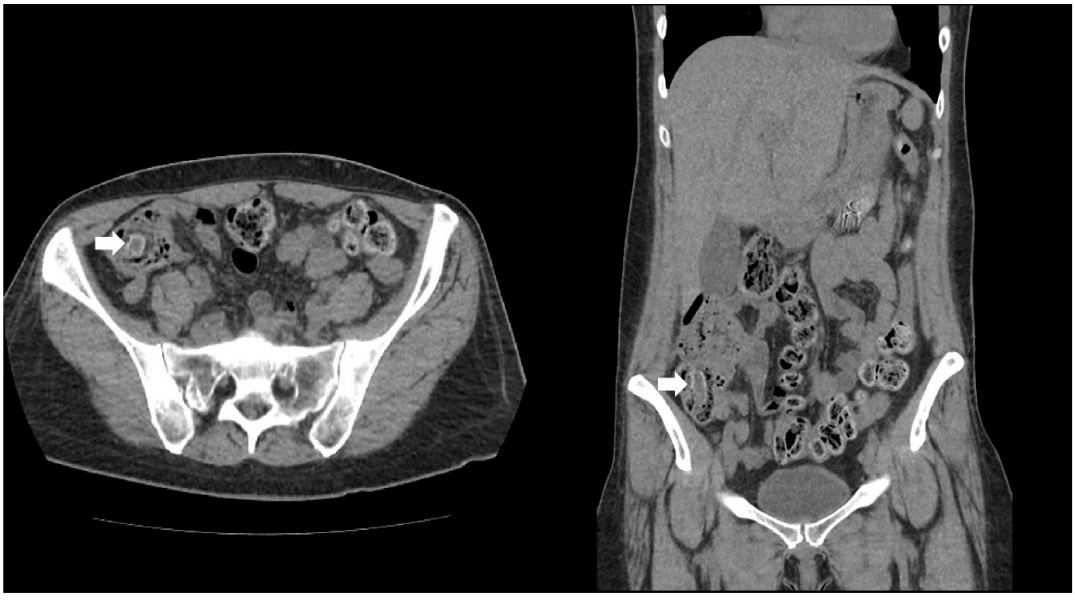

FIG. 1: Computed Tomography (CT) showing the cecal mass/appendix intussusception (arrow).

revealed the following: a 10.0 x 6.0 cm segment of caecum with an attached 7.5 x 4.5 cm segment of ileum was received. The serosa was pink-tan, smooth and intact. The specimen was opened to reveal pink-tan, normally folded mucosa remarkable for a 2.0 x 1.0 x 1.0 cm luminal protrusion at the appendiceal base, forming a polypoid mass.

Microscopic examination demonstrated a polypoid lesion composed of unremarkable fibromuscular stroma admixed with adipose tissue and scattered ganglion cells. The lesion was lined by normal-appearing colonic mucosa with prominent lymphoid follicles (FIG. 3). The overall findings were compatible with an inverted appendix/appendiceal intussusception.

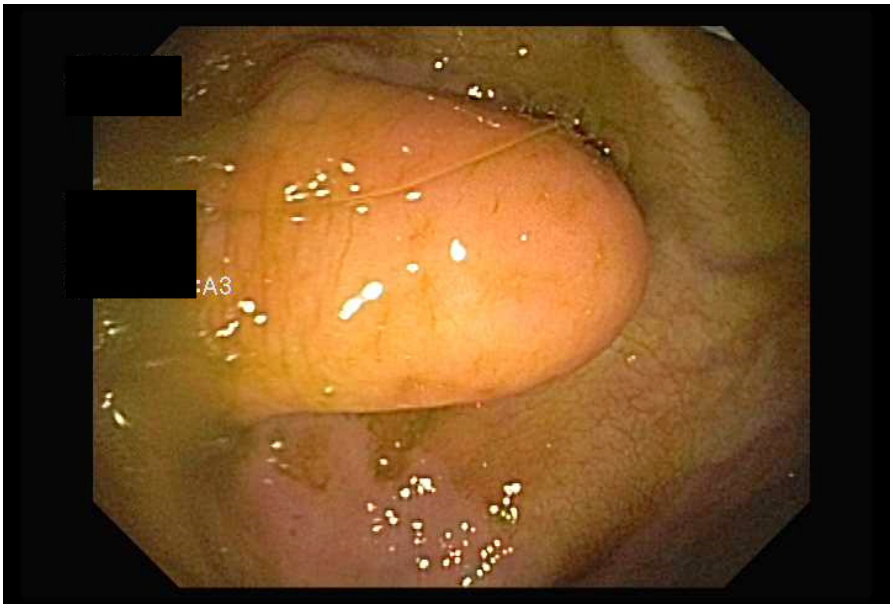

FIG. 2: Colonoscopy showing a lesion arising from the cecum at the appendiceal orifice with intact overlying mucosa.

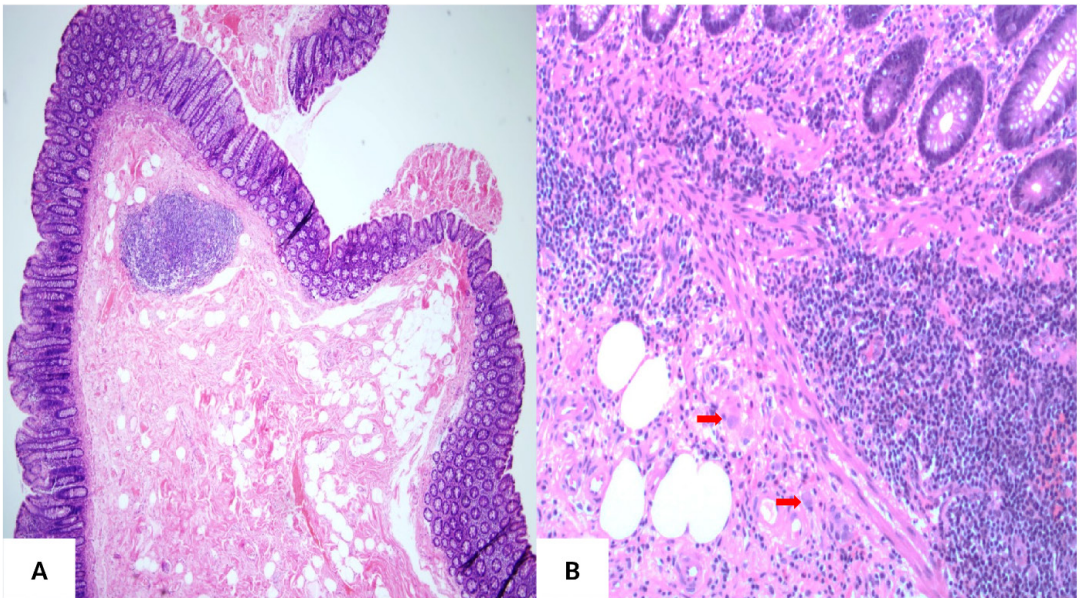

FIG. 3: A. Section of the mass showing fibrofatty and muscular stroma lined by unremarkable colonic mucosa. Note the presence of a lymphoid follicle (H&E stain, 4x). B. higher power view showing scattered ganglion cells (arrows), typically seen in appendix (H&E stain, 20x).

Her postoperative course was uncomplicated except for mild nausea related to her gastroparesis. The patient reported complete resolution of her right lower quadrant pain and overall significant relief of gastrointestinal symptoms after a year from the surgery.

DISCUSSION

Intussusception is defined as telescoping of a bowel segment (intussusceptum) within another (intussuscipiens). While colonic intussusception is a well-established entity, especially in the paediatric population, appendiceal intussusception is a rare entity that was first reported in 1859 by McKidd.[10]

Appendiceal intussusception is classified by McSwain[11] into 5 types: Type I: The tip of the appendix invaginates into the proximal appendix; Type II: Part of the appendix (not the tip) invaginates into the proximal appendix; Type III: Part of the proximal appendix invaginates into the caecum; Type IV: The proximal appendix invaginates into the distal appendix - retrograde invagination; and Type V: Complete inversion/ invagination of the appendix into the caecum.

Our case represents an example of a complete invagination of the appendix into the caecum (Type V) with the formation of a pseudomass in the caecum. Interestingly, Type V appendiceal intussusception can involve the entire colon and protrude from the anus.[12]

Radiologically, diagnosing appendiceal intussusception is difficult. However, the presence of a coiled spring sign along with absence of appendiceal filling during contrast enema is suggestive of intussusception of the appendix.[4] However, contrast enemas are rarely indicated nowadays. Donut or target signs are other helpful signs in ultrasonography.[13] In our case, the diagnosis of appendiceal intussusception was suggested by the CT scan; it showed a mass arising from the appendix base within the caecum and the actual appendix was not visualised. CT scan remains the best modality for diagnosing appendiceal intussusception and typically shows a target-like lesion within the caecum.[14]

Alternatively, colonoscopy can support the diagnosis of appendiceal intussusception especially in cases of complete inversion. In these cases, the endoscopic picture often mimics a polyp or a neoplastic lesion, as in our case. The presence of a polypoid lesion with a central depression located at the appendiceal orifice in the caecum is highly correlated with intussusception of the appendix.[15,16]

Chaar *et al*.[2] reviewed the pathologic findings of 151 cases of reported appendiceal intussusception and found that intussusception was associated with inflammation (29%), endometriosis (26%), mucocele (18%), adenoma (9%), carcinoid (6%), adenocarcinoma (5%), and other rarer findings including hamartoma,

papilloma, mucosa-associated lymphoid tissue lymphoma, juvenile polyp, Crohn's disease and melanosis coli (6%).

Resection is the treatment of choice in symptomatic cases, using either an endoscopic approach (endoloop removal of the appendix) or a surgical approach. The latter varies in complexity from simple appendectomy to right hemicolectomy.[14] Chaar *et al*.[2] favours appendectomy with the inclusion of a caecal cuff for two reasons: first, to eliminate the possibility of another intussusception occurring within the appendiceal stump, which already has been reported in the literature[17]; and second, to create an adequate margin for any potential malignancy arising from the appendix. However, in cases of prior and well-documented malignancy, right hemicolectomy is the recommended approach.[18]

CONCLUSION

It is imperative to include intussusception of the appendix in the differential diagnosis of caecal mass, especially during colonoscopy, as removal of this polypoid lesion can result in a devastating caecal perforation or haemorrhage, necessitating further surgical exploration and increasing patient morbidity.

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