

## CASE REPORT

# Endometriosis-induced intussusception of the caecal appendix

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Accepted 17 November 2014

## SUMMARY

Appendicular intussusception is an uncommon entity, with a reported incidence of 0.01%. The diagnosis is difficult and often only performed at the time of surgery. Intussusception has multiple causes including tumours, foreign bodies and polyps. The definitive treatment is surgical, and the extent of resection is determined by the underlying pathology and degree of invagination. Endometriosis is a rare cause of appendicular intussusception, with 194 cases described in the English literature. We report a case of a 42-year-old woman who presented with chronic abdominal pain in the lower right quadrant. A mass at the caecum was identified during investigations for renal stones by CT. Colonoscopy showed a polypoid lesion, with presumed origin in the appendix. Ileocaecal resection was performed because an appendicular tumour was suspected. Pathological examination identified endometriosis of the appendix and associated peritoneum with invagination of the caecum. The patient was discharged 7 days after surgery and is currently asymptomatic.

## BACKGROUND

Appendicular intussusception was first described by McKidd in 1858 in a 7-year-old child and has an incidence of 0.01%, according to a recent long-term cohort study of 71 000 participants over 40 years.<sup>1–3</sup> This appendicular variation is found in 0.1% of patients undergoing appendectomy.<sup>4</sup>

The invagination of the appendix occurs most often in adults (76%), having a chronic presentation in about 63% of cases. Endometriosis is the underlying cause in about one-third of cases.<sup>2</sup> However, in 1.35% of cases, the intussusception has its origin in a primary tumour of the appendix, which is why a careful assessment of the situation is necessary to choose the appropriate treatment.<sup>5</sup>

Endometriosis, originally described by Von Rokitsky in 1860, is characterised by the presence of foci of endometrial tissue outside the uterine cavity and musculature. The tissue can be identified in ectopic locations such as the intestine, heart and lungs. It can be associated with symptoms such as dysmenorrhoea, pelvic pain and infertility, but an asymptomatic presentation is not uncommon.<sup>3–6</sup> Endometriosis is a disease that affects 8–15% of all women of reproductive age. Intestinal involvement is the extragenital location affected most often, with an incidence that ranges from 3% to 37%.<sup>6–7</sup> Rectum and sigmoid colon are the locations where ectopic tissue is most frequently found. The appendix is involved in 0.08% of cases.<sup>1–6–7</sup>

## CASE PRESENTATION

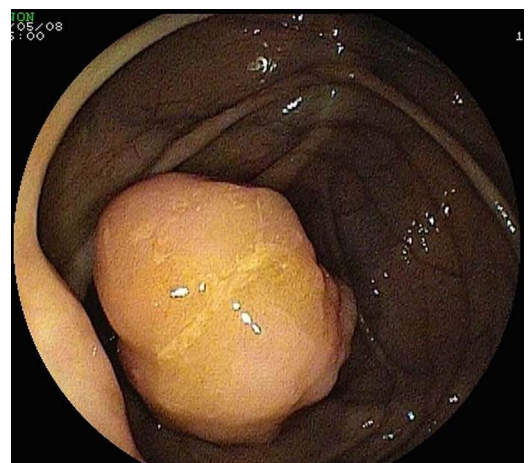
A 42-year-old woman was monitored in urology appointments for chronic abdominal pain in the right lower quadrant and right lower back, associated with right renal lithiasis with hydronephrosis. The patient had a history of bronchitis and hypertension.

## INVESTIGATIONS

An abdominal CT scan was performed, which detected a polypoid lesion in the caecal lumen, raising the suspicion of a caecal neoplasm. The complete blood count (CBC), biochemistry and tumour markers were within analysis reference ranges. Colonoscopy was performed, which identified an exophytic formation on the caecum (figure 1). Biopsies were performed, all of which featured only non-specific inflammatory changes. A virtual colonoscopy was also performed, which did not identify any further lesion but did not exclude the possibility of appendicular tumour (figure 2). In addition, somatostatin receptor scintigraphy was performed preoperatively, but was negative.

## TREATMENT

On the basis of investigations performed it was decided to undertake an exploratory laparotomy. The caecum was approached and no appendix was identified. On palpation of the caecum a mass was palpable within the lumen (figures 3 and 4) and an ileocaecal resection performed.



**Figure 1** Polypoid image, apparently originating from appendiceal orifice, visible on colonoscopy.



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**To cite:** Costa M, Bento A, Batista H, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-200098



**Figure 2** Virtual colonoscopy, demonstrating an area topographically compatible with the appendix, not filled by oral contrast due to its occlusion.

### OUTCOME AND FOLLOW-UP

The postoperative period was uneventful and the patient was discharged after 7 days. Histopathological examination revealed endometriosis of the ileocaecal appendix, with invagination into the caecum (figure 5).

The patient was followed up as an outpatient approximately 1 month after surgery and was asymptomatic.

### DISCUSSION

Appendicular intussusception can be secondary to intrinsic or extrinsic causes. Anatomical factors such as a fully mobile appendix, a thin and narrow mesoappendix, a large proximal appendiceal lumen, a partially fixed caecum or hyperperistalsis may be involved in invagination of the appendix into the caecum.<sup>1 5</sup> However, there are changes secondary to the appendix itself that may be the cause of this phenomenon, such as

polyps, mucous cysts, parasites, endometriosis, lymphoid hyperplasia, faecoliths, foreign bodies, adenocarcinoma or carcinoid tumours.<sup>1 4</sup>

Most cases of intussusception are diagnosed during surgery (57%), with preoperative diagnosis being achieved in 32% of cases. This number has increased with the progressive development of diagnostic methods.<sup>2</sup>

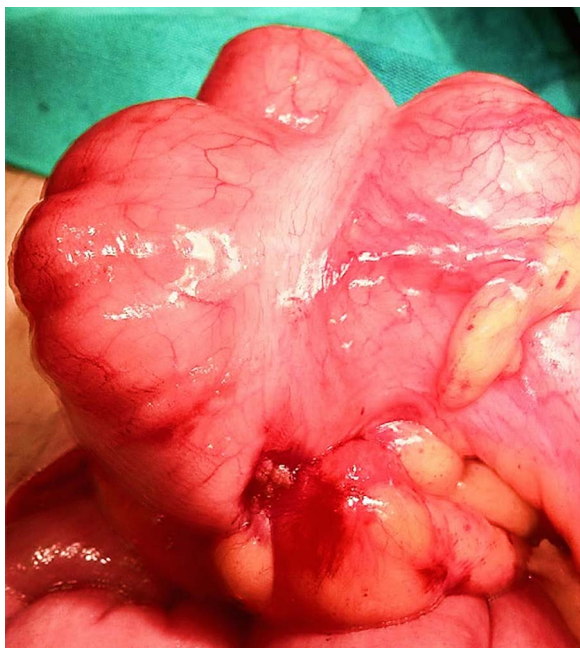
Endometrial tissue is most often found in the serosa and muscular layer of the intestinal tract. Serosal endometriosis may result in fibrosis and adhesions, while the involvement of the muscular layer causes smooth muscle hyperplasia, resulting in a reduction of the intestinal lumen and causing a mass that is propelled through the intestine by peristalsis. If the endometrial deposit is located on the appendix, appendicular inversion and its intussusception into the caecum can result.<sup>6</sup>

We found 194 cases published in English in PubMed from 1925 to 2012, using the keywords 'appendix intussusception', 'appendiceal intussusception', 'intussuscepted AND appendix' and 'appendiceal AND intussuscepted'.

The clinical presentation of appendiceal intussusception includes a broad spectrum of symptoms, which can be classified into four types: the first mimics the symptoms of acute appendicitis; the second includes the typical symptoms of intussusception (abdominal pain and vomiting for several days, occasionally with diarrhoea, vomiting or melaena); the third is characterised by pain in the lower right quadrant of the abdomen, associated with recurrent vomiting and melaena for several weeks or months; in the fourth type, patients are completely asymptomatic.<sup>1 8-11</sup> A chronic presentation occurs in 63% of cases and the most frequently reported symptoms are abdominal pain (78%), vomiting (26%) and rectal bleeding (23%). There is a palpable mass in the right lower quadrant in 37% of paediatric patients and in 13% of adult patients.<sup>2</sup>

Appendicular intussusception can be classified into five types marked by differing patterns of invagination of the appendix and caecum (box 1),<sup>8 12</sup> with this case being type 5.

The diagnosis of appendicular intussusception is often difficult and the entity frequently mistaken for a caecal malignancy.<sup>4 12 13</sup> Abdominal ultrasound may identify a doughnut/target sign (ie, a concentric ring) as part of the appendicular topography, which



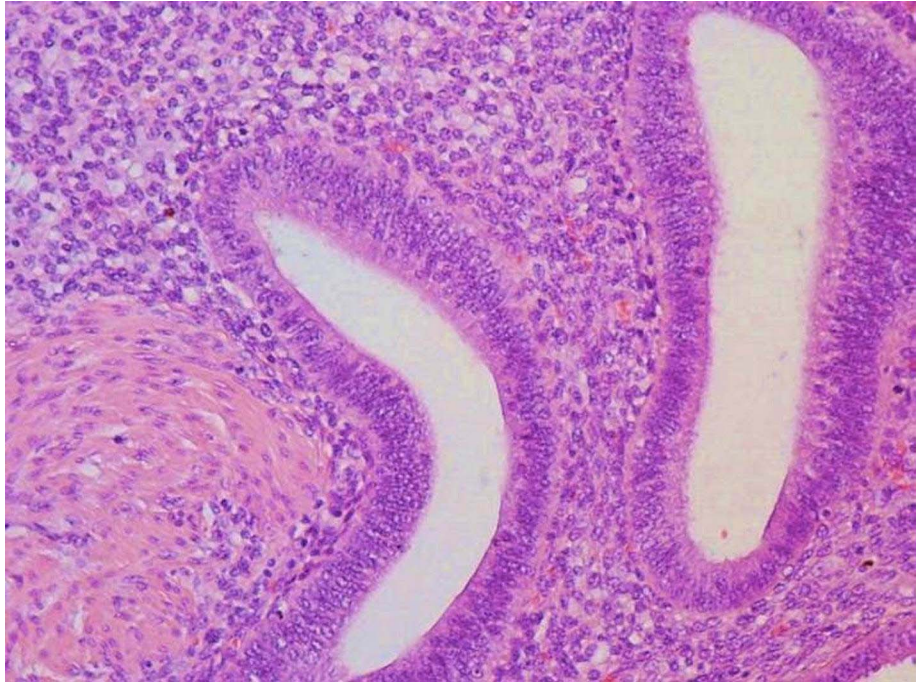
**Figure 3** Caecum, with the presumed origin of the inverted appendix.



**Figure 4** Internal aspect of the caecum, showing the ileocaecal valve on the right and the inverted appendix on the left.



**Figure 5** Endometrial glands and the involving stroma (H&E,  $\times 100$ ).



practically confirms a diagnosis of intussusception. Abdominal CT also identifies a target lesion in the affected bowel. CT with three-dimensional reconstruction improves anatomical resolution and helps to support the diagnosis.<sup>2</sup>

Colonoscopy may also assist as it allows direct visualisation of the lesion and in this examination the invaginated appendix is seen as a polypoid lesion with a central depression at the appendiceal orifice.<sup>2,4</sup>

In centres with appropriate expertise endoscopic ultrasonography is considered the best procedure for visualisation of submucosal tumours to determine their origin and has been used to demonstrate appendiceal intussusception.<sup>4</sup>

Histopathological examination is mandatory for the definitive diagnosis of an intussusception due to endometriosis, which is the most common cause of appendiceal intussusception in adults (33%).

Treatment is surgical since invagination of the appendix increases the risk of secondary intussusception of other bowel segments.<sup>1</sup> Depending on the pathology that triggers the intussusception, treatment may vary from appendicectomy to right hemicolectomy<sup>1</sup> but as most of the precipitating lesions are benign, appendicectomy is often definitive.<sup>2</sup> Previous reports

indicate 49% of patients underwent partial colectomy, a similar proportion open appendicectomy and in 2% of patients a laparoscopic approach to appendicectomy was used.<sup>2</sup> Colonoscopic removal of an intussuscepted appendix was also attempted by some authors, closing the inverted stump with an endoloop device and/or a polypectomy snare. This procedure does carry the risk of perforation of the appendiceal stump or the caecum.<sup>2</sup>

In this case, we undertook an ileocaecal resection, as the anatomy of the intussusception caused technical difficulties for performing appendectomy.

In patients with confirmed endometriosis, the use of Danatrol or luteinising hormone-releasing-hormone analogues (eg, leuprorelin) preoperatively can decrease inflammation and vascularisation thus facilitating the surgical procedure,<sup>7</sup> however, such an approach was not feasible in this case as the preoperative diagnostic imaging suggested an underlying malignancy.

#### Box 1 Types of invagination (McSwain, 1971)

1. Involves the distal portion of the appendix, with its invagination into the proximal portion
2. Involves the middle portion of the appendix, with its invagination into the proximal portion
3. Involves the base of the appendix, with its invagination into the caecum
4. The proximal portion of the appendix is invaginated into the distal portion
5. Complete inversion of the appendix into the caecum

#### Learning points

- Appendicular intussusception is a rare phenomenon that is difficult to diagnose, with a non-specific clinical presentation that can simulate acute appendicitis and raise the suspicion of colonic malignancy.
- US and CT can identify the lesion, but have limited diagnostic value.
- Surgical treatment is always the first option in these cases, and the type of resection is determined by the degree of suspicion for malignancy.
- Endometriosis is a rare cause of intussusception and diagnosis can only be confirmed by histological examination.

**Competing interests** None.

**Patient consent** Obtained.

**Provenance and peer review** Not commissioned; externally peer reviewed.

## REFERENCES

- 1 Liang H, Huang M, Wei P, *et al.* Endometriosis-induced appendiceal intussusceptions. *Am J Surg* 2009;197:e66–8.
- 2 Chaar CI, Wexelman B, Zuckerman K, *et al.* Intussusception of the appendix: comprehensive review of the literature. *Am J Surg* 2009; 198:122–8.
- 3 Offodile A, Hodgins JB, Arnell T. Asymptomatic intussusception of the appendix secondary to endometriosis. *Am Surgeon* 2007;73:299–300.
- 4 Tseng P, Lee Y, Chiu H, *et al.* Appendiceal intussusception diagnosed with endoscopic sonography. *J Clin Ultrasound* 2006;34:348–50.
- 5 Nycum LR, Moss H, Adams JQ, *et al.* Asymptomatic intussusception of the appendix due to endometriosis. *Southern Med J* 1999;92:524.
- 6 Moradi P, Barakate M, Gill A, *et al.* Intussusception of the vermiform appendix due to endometriosis presenting as acute appendicitis. *ANZ J Surg* 2007;77:758–60.
- 7 Verspyck E, Lefranc JP, Guyard B, *et al.* Treatment of bowel endometriosis: a report of six cases of colorectal endometriosis and a survey of the literature. *Eur J Obst Gynec Rep Bio* 1997;71:81–4.
- 8 Honmyo U, Shimada S, Yamamoto S, *et al.* Reversible protruded lesion regarded as idiopathic appendiceal intussusceptions. *Digest Endos* 2001;13:90–3.
- 9 Franco LL, Rolim CF, Garcia RA, *et al.* Qual é o seu diagnóstico? *Radiol Bras* 2003;36:V–VII.
- 10 Marín MR, Baños PA, Valverde FM, *et al.* Appendiceal intussusception resulting from endometriosis presenting as acute appendicitis. *Am Surgeon* 2010;76:906–8.
- 11 Emre A, Akbulut S, Yilmaz M, *et al.* An unusual cause of acute appendicitis: appendiceal endometriosis. *Int J Surg Case Rep* 2013;4:54–7.
- 12 Mehmood S, Phair A, Sahely S, *et al.* Appendiceal intussusception caused by endometriosis. *Lancet* 2012;380:1202.
- 13 Hanly AM, Ryan EM, McNamara DA. Appendiceal intussusception and endometriosis mimicking colorectal cancer. *Colorectal Dis* 2010;13:225–6.

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