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Rare ileocaecal intussusception from small bowel adenocarcinoma

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Abstract

Intussusception in adults is uncommon, representing only 1% of bowel obstruction cases and often indicates an underlying pathological mass, frequently requiring surgical resection. This report presents a rare histological subtype of adenocarcinoma in an adult, complicated by extensive ileocolic intussusception. A female in her mid-fifties presented with acute abdominal pain, nausea, vomiting, and rectal bleeding. Imaging revealed a 20 cm ileocolic intussusception caused by a calcified mass in the ileum. A laparotomy revealed a grossly oedematous ileum and a palpable mass as the lead point within it. A small bowel wedge resection was performed. Histopathology showed a poorly differentiated adenocarcinoma with focal neuroendocrine differentiation (NED). The patient's recovery was prolonged, and subsequent CT studies have not demonstrated recurrence so far. Small bowel cancer, although rare, is on the rise. Adenocarcinomas are the most common histological subtype, and neuroendocrine differentiation in such tumours is extremely rare. This represents a rare instance of ileal adenocarcinoma with neuroendocrine differentiation in a patient with unaltered anatomy. The presence of neuroendocrine differentiation may confer a poorer prognosis, as seen in other gastrointestinal tumours, however the long-term behaviour and management of this rare subtype remains unclear. Further research is needed to understand the prognostic implications and to develop a standardised treatment regimen for this rare pathology.

Keywords Case report, Intussusception, Small bowel adenocarcinoma, Neuroendocrine differentiation, Histopathology

1 Introduction

Intussuception is defined as "the invagination of one segment of the bowel into an immediately adjacent segment". The intussusceptum refers to the proximal aspect that intussuscepts into the distal segment (intussuscipiens) [1]. Intussusception in adults is rare and accounts for just 1% of cases of bowel obstruction. The proximal bowel is drawn into the peristalsing distal bowel, which leads to oedema, eventually compromising the vascular flow and leading to necrosis and perforation if left untreated [1]. The management of intussusception in adults is different from children, as in adults it is frequently

associated with a pathological lesion acting as a lead point, especially in the ileocolic region. Therefore, treatment usually involves resection en bloc of the intussuscepted segment or causative mass, rather than reduction via insufflation, as can be often used in young children. Small bowel intussusceptions can be transient and asymptomatic and are sometimes incidentally captured on imaging in asymptomatic patients [1].

2 Patient information

A female in her mid-fifties presented to the emergency department (ED) with severe generalised abdominal pain, nausea, vomiting and bleeding per rectum. She had a background of oesophagitis, duodenitis, alcohol excess and Factor V Leiden heterozygote with previous pulmonary emboli and deep vein thrombosis, but was not currently anticoagulated. Of note, she had no past medical history of inflammatory bowel disease or any familial polyposis syndrome. She also reported experiencing daily abdominal pain for the past eight months, along with weight loss. She had previously visited the emergency department for these symptoms, where they were initially attributed to chronic pancreatitis.

3 Clinical findings

On examination, her abdomen was mildly distended, with marked generalised tenderness on palpation. She was focally peritonitic in the lower abdomen. Observations performed in ED demonstrated mild tachycardia but she was apyrexial.

4 Timeline

Figure 1 Outlines the timeline of events from presentation to discharge.

5 Diagnostic assessment

5.1 Laboratory findings

Her CRP was mildly raised at 29 mg/l but with a normal haemoglobin and white blood cell count. Differential diagnoses at the time included bowel obstruction, acute diverticulitis and perforation.

5.2 Imaging

A contrast enhanced CT (computed tomography) of the abdomen and pelvis was performed in the portal venous phase (Fig. 2). It showed a large ileocaecal intussusception measuring at least 20 cm in the lower abdomen, causing upstream small bowel obstruction. There was marked mural oedema throughout the affected bowel with small volume surrounding fluid, however there was no perforation, collection or features of established bowel ischaemia. A lead point was identified, consisting of a calcified mass in the ileum measuring 17 mm, which had intussuscepted into the caecum. The patient was referred to the general surgical team and booked urgently for theatre.

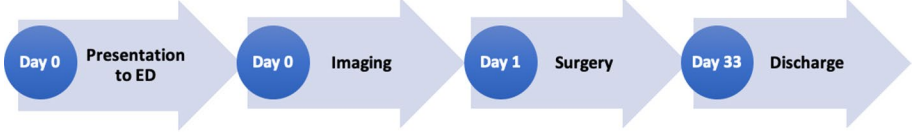

Fig. 1 outlines the timeline of events

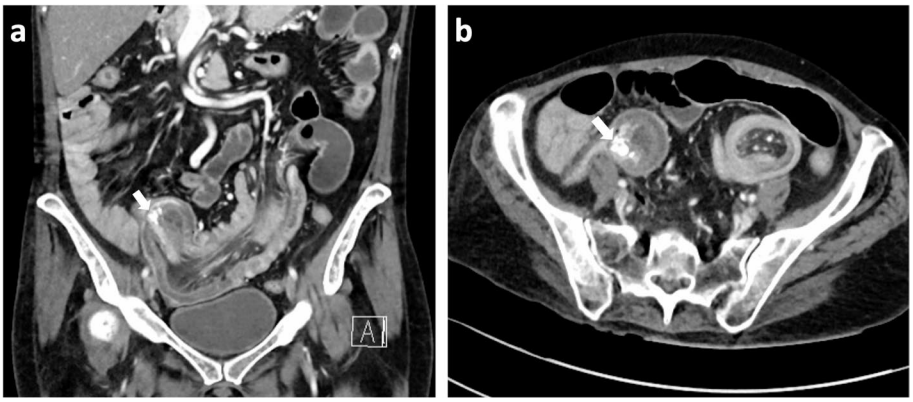

Fig. 2 Coronal (**a**) and axial (**b**) contrast-enhanced portal venous phase CT images showing an extremely long ileocolic intussusception with resultant small bowel obstruction. The lead point is a 17 mm calcified mass in the ileum (arrows)

6 Therapeutic intervention - surgery

Prior to theatre, laparoscopic approach was deemed unsuitable, as there was extensive small bowel obstruction due to the intussusception, and reduction of the intussusception would not be possible with laparoscopic instruments since the small bowel had progressed to the caecum. A midline laparotomy was performed and the bowel was exposed. The small bowel and colon were meticulously inspected and a lead point was identified. Clear reactive fluid was found in the abdominal cavity; there was no evidence of pus or enteric contents. The small bowel had intussuscepted and tracked distally all the way into the caecum (Fig. 3). The intussusception lead point was identified around a hard palpable mass 30–45 cm from the ileocaecal junction. The terminal ileum was grossly oedematous, but the bowel was deemed viable. The intussusception was reduced as much as possible, freeing up a 30 cm length of small bowel. Due to the bowel's gross oedema, serosal tears were noted over the lead point after reduction. A small bowel wedge resection was performed around the lead point and a small mesenteric lymph node dissected; these specimens were sent to histopathology for analysis. A side-to-side small bowel anastomosis was performed.

7 Pathology specimen

The specimen (Fig. 4) comprised a segment of small bowel with noticeable intussusception measuring 100 mm in length with a maximum diameter of 40 mm. There was evidence of partial serosal defect (tearing). Slicing through the specimen revealed a speckled calcified nodule within the lead point of the intussusception, measuring 15 mm in maximum diameter. It was 80 mm from the nearest margin.

8 Histology

Histological examination showed the tumour was comprised predominantly of poorly differentiated sheets of tumour cells with focal atypical glands, signet rings and focal areas with a rosette pattern (Fig. 5). There was a central focus of extracellular mucin. Immunohistochemistry showed the lesion was positive for BerEP4 (carcinoma marker). There was patchy positive staining for neuroendocrine markers chromogranin and synaptophysin. There was weak positive staining for CDX2 (marker for intestinal adenocarcinoma). The mesenteric lymph node was negative for tumour. The final diagnosis was a

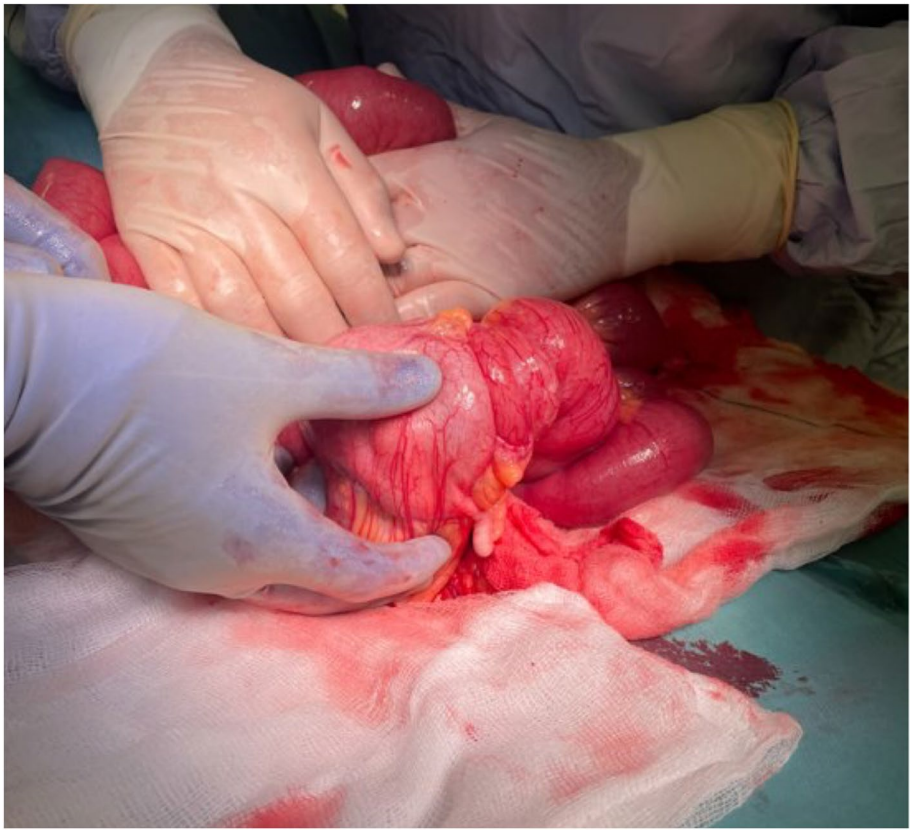

Fig. 3 Intra-operative photograph of the small bowel intussuscepting into the caecum

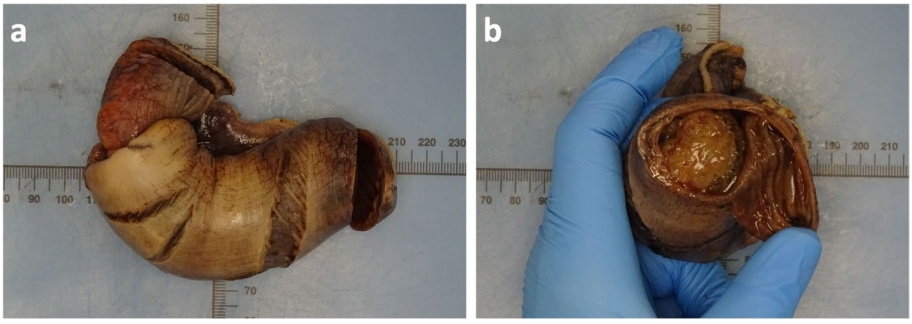

Fig. 4 Small bowel resection with serosal tearing (**a**). There was a speckled calcified nodule within the lead point of the intussusception measuring 15 mm in maximum diameter, comprising the tumour (**b**)

30 mm poorly differentiated adenocarcinoma with focal signet rings and focal neuroendocrine differentiation (pT3 pN0 pM0).

9 Follow-up and outcomes

Post-operative recovery was prolonged due to complex discharge planning needs, stemming from a combination of postoperative complications and package of care/social care problems. Specifically, the patient experienced wound dehiscence, and recurrent severe urinary tract infections. The package of care also took time to arrange, as the patient required significant support to recover at home, including social care, a health assessment, and the provision of necessary equipment. The patient was then discharged

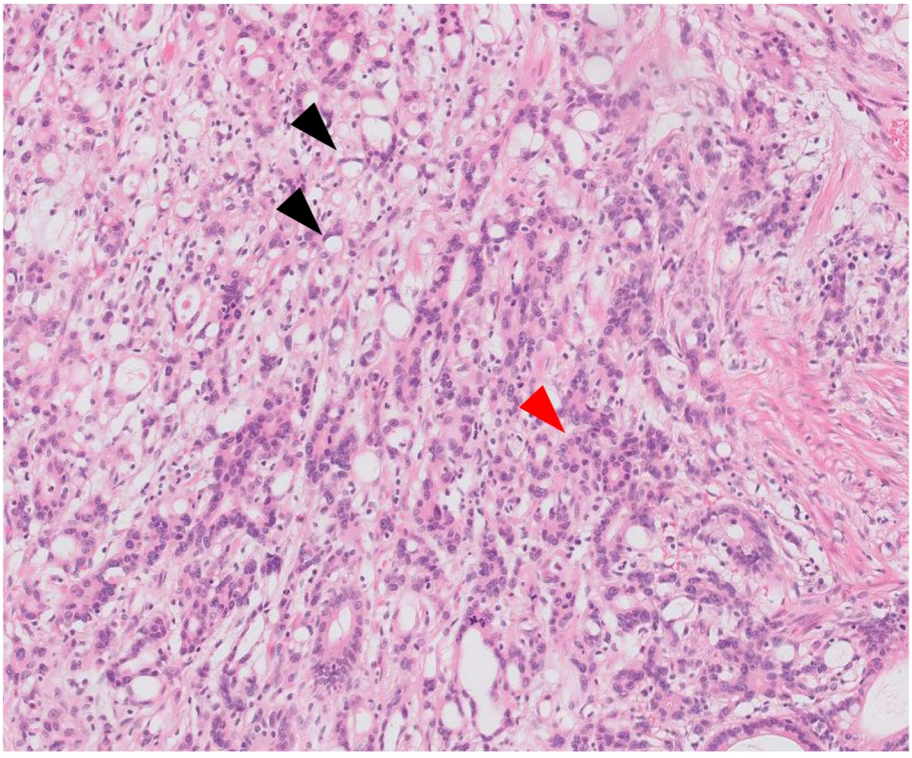

Fig. 5 Histological examination showed the tumour was comprised predominantly of poorly differentiated sheets of tumour cells with focal atypical glands, signet rings (black arrows) and focal areas with a rosette pattern (red arrow)

approximately one month after surgery. Oncology follow-up appointments were arranged, but the patient did not attend.

Nine months later, the patient re-presented with vomiting, abnormal liver function tests and severe electrolyte disturbances. Her condition had significantly deteriorated due to complications related to chronic pancreatitis, which was secondary to chronic alcohol excess, and was evidenced by multiple peripancreatic pseudocysts. The worsening liver function, ascites, and portal hypertension were directly attributable to the newly diagnosed liver cirrhosis. Additionally, the patient had a coeliac artery occlusion and a distal splenic artery pseudo-aneurysm, for which a failed interventional radiology (IR) procedure attempt had occurred, indicating complex vascular involvement contributing to their overall systemic decline. As the patient was not a candidate for liver transplantation or trans-jugular intrahepatic portosystemic shunt (TIPS), she will be considered for a clinical trial for severe, untreatable ascites. She has undergone CT studies since initial presentation which have not shown local or nodal recurrence or metastatic disease thus far.

10 Discussion

Small bowel cancer is rare, accounting for less than 1% of all cancer in the United Kingdom, however the incidence is rising, with a 176% increase in cases since the early 1990 s [2]. Median age of incidence is 60 years old [3]. Symptoms can be vague such as non-specific abdominal discomfort and diagnosis is usually made after an acute presentation such as small bowel obstruction or gastrointestinal haemorrhage, and is often diagnosed at stage IV disease. This carries a median prognosis of < 20 months, even in those who

receive chemotherapy or metastasectomy [4–6]. The main histological subtypes are adenocarcinoma (40%) and neuroendocrine tumours (40%), followed by the less common stromal tumours and lymphoma. The most commonly affected part of the small bowel for all cancers is the duodenum (50%), followed by the jejunum (30%) [3].

Risk factors for adenocarcinoma include Crohn's disease (conferring an 8-fold increase in risk, most commonly in the ileum [3, 7]) alcohol, smoking, coeliac disease and familial syndromes such as Lynch syndrome, familial adenomatous polyposis and Peutz-Jeghers syndrome [8]. There may be other contributing factors, such as low fibre diets and the overconsumption of red, smoked and processed meats [3].

Neuroendocrine cells comprise approximately 2% of cells in the gastrointestinal tract and have the ability to receive signals from the nervous system, as well as secrete hormones, peptides and monoamines [9]. Neuroendocrine differentiation has been identified in cancer specimens of non-neuroendocrine organs such as lung, breast and gastrointestinal tract. The differentiation can be found as single cells or small nests within the adenocarcinoma specimen. Specific histological immunohistochemical markers can help identify these, such as staining for chromogranin A and synaptophysin [10]' [9].

Gastrointestinal adenocarcinoma with neuroendocrine differentiation is histologically different from MiNEN (mixed neuroendocrine neoplasm), previously known as MANEC (mixed adeno-neuroendocrine carcinoma) [9], which requires at least 30% of neuroendocrine component in the tumour (plus another tumour component such as adenocarcinoma or squamous cell carcinoma) [11], and is also rare [12]. A study by Xu et al. found that in a database of patients with NETs and MiNENs, only 5.9% of cases were MiNENs and only 4.9% of MiNENs were in the small intestine as opposed to 42.2% of NETs [13]. Our case did not fit the histological criteria for MiNEN as the neuroendocrine component was less than 30% of the tumour.

Gastrointestinal adenocarcinoma with neuroendocrine differentiation is even rarer than MiNEN. It has been reported in the literature in the oesophagus, stomach, duodenum, jejunum, colon and rectum [14]. One case of this entity in the ileum the authors could identify in the literature was in a patient with an ileal conduit and renal transplantation [15], which could possibly relate to the carcinogenic properties of urine within bowel, the immunosuppressive agents necessary after renal transplantation [16] or due to patient demographic and lifestyle factors. Another reported case of small bowel adenocarcinoma with neuroendocrine differentiation was reported by Tsujii et al. in 2002 [17]. They described a 40 mm mesenteric tumour causing right-sided ureteric obstruction in a 62-year-old male, which was post-operatively found to be arising from within an ileal diverticulum. It is unclear if this could be related to chronic inflammation, as the mucosa lining the narrow neck of the diverticulum showed dysplastic change. The patient described in their case report received systemic adjuvant chemotherapy (agent unspecified), but unfortunately developed inguinal nodal, liver and brain metastases and died 18 months after his primary surgery. No other case reports of adenocarcinoma with focal neuroendocrine differentiation in the small bowel were identified by the authors.

The effect of neuroendocrine differentiation on the long term disease behaviour of small bowel adenocarcinomas such as our case is not known. Prognosis has been evaluated in adenocarcinoma with neuroendocrine differentiation in the colon, in a meta-analysis performed by Zeng et al. which evaluated eleven studies, and deemed focal

neuroendocrine differentiation to confer a lower 5 year survival compared to those without neuroendocrine differentiation [10]. Another study by Wang et al. found that gastric adenocarcinomas with neuroendocrine differentiation were associated with poorer survival and were more resistant to neoadjuvant chemotherapy [18]. It could be postulated that since neuroendocrine differentiation confers poorer survival elsewhere in the gastrointestinal tract, that this could apply to ileal/small bowel tumours, however there is no published evidence for this that the authors could identify.

There is no set treatment regimen for small bowel adenocarcinomas in general. If the tumour is localised, resection is the treatment of choice [3]. There is evidence that metastasectomy for oligometastatic disease improves survival; with a median overall survival of 34.5 months versus 17.1 months for those who get chemotherapy alone [4]. There is evidence of response to fluoropyrimidine + platinums (+ bevacizumab) and to immunotherapy for MMR (mismatch repair) deficient tumours [4]. There is varying literature regarding efficacy of chemotherapy and immunotherapy agents in the adjuvant setting, with some retrospective studies showing benefit and some showing no benefit [3]. The first global prospective study evaluating adjuvant chemotherapy, specifically fluoropyrimidine- based adjuvant therapy with or without oxaliplatin, the BALLAD study, is currently underway and the results are yet to be published [19].

Amongst this uncertainty, there is even less evidence on the benefit of oncological agents in small bowel adenocarcinoma with neuroendocrine differentiation, with no specific recommended chemotherapy regimen found in the literature, therefore if encountered, the specific treatment may vary depending on the centre and oncological expertise. In all cases, including those which have undergone resection, close surveillance would be prudent, given the rarity and possible unpredictable nature of this pathology.

11 Limitations

This case report is limited by incomplete follow-up due to the patient's non-attendance at oncology appointments, resulting in a lack of long-term data. There is uncertainty surrounding the biological behaviour of focal neuroendocrine differentiation, and the lack of consensus on appropriate adjuvant therapy. Additionally, molecular analysis was not available. As this is a single case, the findings may not be generalisable to the broader patient population.

12 Learning points

- Intussusception in adults can be due to a pathological lead point, and imaging should be scrutinised to assess for this.
- The ongoing BALLAD trial may offer guidance on optimal treatment regimens for small bowel adenocarcinoma, though its results are not expected to directly address the impact of neuroendocrine differentiation in these cases.
- The long term behaviour of small bowel adenocarcinoma with neuroendocrine differentiation is unknown, highlighting the need for close surveillance of these patients and further research.

13 Patient perspective

Patient perspective was obtained during a subsequent inpatient stay under hepatology for portal hypertension: "*I had been experiencing abdominal pain for several months, but it became much worse suddenly, and I was very worried when I started vomiting and seeing blood. It has been an ongoing issue, and things don't seem to be improving. I am glad that my case is being used for education, happy to help young doctors.*".

14 Conclusion

Small bowel adenocarcinoma is rare, and focal neuroendocrine differentiation is even rarer. This represents a rare instance case of adenocarcinoma with neuroendocrine differentiation in the ileum of a patient with non-altered anatomy, and therefore the clinical significance of this histological feature in small bowel tumours remains uncertain due to the lack of reported cases. The results of the BALLAD trial are pending and may provide some guidance on the recommended adjuvant treatment regimen for small bowel adenocarcinoma, but is not likely to address the implications and recommendations for the conundrum of adenocarcinoma with neuroendocrine differentiation.

Author contributions

S.M., M.A., G.R., A.D., K.H., M.P., R.A., A.G., L.C., S.L., S.K., N.P., D.T. and A.W. prepared the main manuscript text. G.R. prepared Figure 1 A.G. prepared Figure 2 A.D. prepared Figs. 3 and 4. All authors have reviewed and approved the final version of the manuscript and agree to its submission.

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Data availability

All relevant data are included in the article. Additional anonymised clinical or imaging data may be made available from the corresponding author upon reasonable request and with appropriate institutional approval.

Declarations

Ethics approval and consent to participate

Institutional review board at City St Georges University of London waived the requirement for formal ethical review. Not applicable as no intervention/research specific tests were performed.

Patient confidentiality all patient information has been anonymised to protect privacy

No identifiable details are included in this manuscript, in accordance with ethical standards

Consent for publication

Written informed consent was obtained from the patient for publication of this case review, including accompanying images.

Competing interests

The authors declare no competing interests.

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