CASE REPORT

Intestinal invagination secondary to intestinal adenocarcinoma in coeliac disease

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Accepted 28 March 2015

SUMMARY

Invagination is defined as a medical condition in which a part of the gastrointestinal tract has entered into another section. Intestinal invagination is a rare clinical entity among adults and there is an underlying structural lesion in most of the cases. Coeliac disease is considered as a risk factor for intestinal invagination, because of the associated inflammatory processes and motility disorders as well as the increased risk for secondary malignancies. We report a case of intestinal invagination secondary to intestinal adenocarcinoma in a woman with coeliac disease, whose adherence to a gluten-free diet was poor.

BACKGROUND

Invagination is defined as a medical condition in which a part of the gastrointestinal tract has entered into another section. It constitutes 1–3% of intestinal obstructions that necessitate surgery and 0.003–0.02% of all hospital admissions.¹ Although it is a common abdominal emergency during infancy, 5% of all invagination cases can be seen in adults. This condition is generally idiopathic among children, whereas there may be various structural problems in 80–90% of adult invaginations.² “While tumours are responsible for these structural problems in almost half of the cases, non-tumoural structural problems such as cystic fibrosis may also lead to intussusception”.^{3,4} However, inflammatory or idiopathic causes may also play a role in a small proportion of invagination in adult patients. Coeliac disease is considered a risk factor for adult invagination without a ‘lead point’; however, it may also cause invagination with a ‘lead point’ in the adult population by precipitating secondary malignancies, such as intestinal lymphoma or adenocarcinoma.²

We report a case of intestinal invagination secondary to intestinal adenocarcinoma in a 58-year-old woman with coeliac disease.

CASE PRESENTATION

A 58-year-old woman with the diagnosis of coeliac disease was admitted to the emergency ward. She had abdominal pain, distention, nausea and bilious vomiting for 2 days. Her history revealed that her adherence to a gluten-free diet was poor and she had been followed up for iron deficiency anaemia for a year. She had tenderness in the upper quadrants of the abdomen during physical examination. Her white cell count and haemoglobin levels were $8.9 \times 10^3/\mu\text{L}$ ($4000\text{--}10\,000 \times 10^3/\mu\text{L}$) and 8.4 g/dL ($12.5\text{--}14.0 \text{ g/dL}$), respectively. Her blood biochemistry testing revealed no abnormality, whereas direct abdominal

Figure 1 Plain abdominal X-ray of the patient.

X-ray demonstrated dilation of the bowel segments (figure 1). Abdominal ultrasonography showed thickening in the left lateral abdominal wall, reaching to 40 mm. Moreover, an enlarged mass with a secondary intestinal segment was detected via ultrasonography, suggesting intestinal invagination. The proximal part of this intestinal segment was dilated. Sequential axial images obtained via abdominal CT revealed invagination of proximal jejunal segments, which involved surrounding vascular structures and mesenteric fat tissue in the upper left abdominal quadrant (figure 2). The patient underwent emergency surgery with the diagnosis of ileus due to intestinal invagination.

During the operation, a jejunal invagination was detected 20 cm away from the ligament of Treitz. There was a 4 cm, ulcerovegetant, grey coloured hard mass lesion at the invaginated segment (figure 3). The invaginated segment of approximately 20 cm was resected. The patient did not have any problems during the early postoperative period and started her diet on the fourth postoperative day. She was discharged on the seventh postoperative day. The histopathological examination of the mass was reported as moderately differentiated adenocarcinoma. The mass invaded lamina propria (pT2); however, no vascular or perineural invasion was detected. The surgical margins and all of the four resected lymph nodes were tumour free. The patient was instructed to adhere to the gluten-free diet and was started on a surveillance programme with the diagnosis of intestinal invagination secondary to intestinal adenocarcinoma, which was developed on the basis of coeliac disease.

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To cite: Sahin C, Ozseker B, Sagiroglu T, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-208703

Figure 2 The axial CT images of the case. Invagination of proximal jejunal segments involving the surrounding vascular structures and mesenteric fat tissue in the upper left abdomen can be seen.

DISCUSSION

The association between the coeliac disease and intestinal invagination could not be entirely elucidated. The intestinal invagination seen in patients with coeliac disease is generally non-obstructive, transient and recurrent. Although the exact mechanisms are not known, inflammation and decreased intestinal motility are suggested to be the main reasons of the invagination seen in these patients when no other structural lesions are detected.⁵ The second important cause of the invagination in coeliac disease is the structural lesions developed due to secondary malignancies. It is known that untreated or poorly controlled coeliac disease is an important risk factor for intestinal malignancies.⁶ Adenocarcinoma and T-cell lymphoma associated with enteropathy are the most common tumours detected in coeliac disease patients. The intestinal adenocarcinoma risk of patients with coeliac disease is 10 times greater than in the general population.⁷

Figure 3 Photograph of the invagination taken intraoperatively.

Although the actual carcinogenesis process in coeliac disease is not entirely known, several possible mechanisms such as genetic factors, environmental carcinogens, chronic inflammation, chronic antigenic stimulation, release of proinflammatory cytokines and nutritional disorders have been suggested to play roles. Various mutations are also shown to be involved in the carcinogenesis process in coeliac disease patients. Several studies demonstrated the association between abnormal CpG island methylation/microsatellite pathway and coeliac disease.⁸ Moreover, the protective potential of the adherence to a gluten-free diet against the development of intestinal cancers has also been established.⁹ Besides the coeliac disease, inflammatory diseases such as Crohn's disease, familial adenomatous polyposis, hereditary non-polyposis colorectal cancer and Peutz-Jegher's syndrome, may also increase the risk of malignancy. Moreover, several environmental factors such as smoking, alcohol consumption, smoked foods and high meat intake may precipitate the development of intestinal malignancies.

Most invaginations in adults are enteroenteric or iliocolic. Enteroenteric invaginations are detected in most non-neoplastic cases whereas malignancies are the aetiological factor for 50–60% of colocolonic invagination cases.

The most common symptom of invagination in coeliac disease is abdominal pain.² Obstructive symptoms, nausea, vomiting and palpation of a smooth abdominal mass are other important findings of invagination. Intestinal adenocarcinoma generally presents with non-specific signs and symptoms such as anaemia, abdominal pain, weight loss, loss of appetite, gastrointestinal bleeding, nausea and vomiting. It may also present with obstruction symptoms in advanced cases. Therefore, physicians who's patients face these problems must consider invagination and intestinal adenocarcinoma in the differential diagnosis.

None of the diagnostic tests have been proven to be superior to the others in the diagnosis of intestinal pathologies.

Generally, multiple diagnostic modalities are used in tandem. Abdominal ultrasonography is commonly preferred for the diagnosis of invagination in infants. However, CT and MRI studies are the diagnostic methods utilised for the imaging of intestinal segments in the adult population. A 'target' or 'sausage' sign detected in CT is typical for invagination. These methods are non-invasive and provide additional information regarding the extra-luminal pathologies. Video capsule endoscopy (VCE) can also be used for the diagnosis of intestinal adenocarcinoma by directly visualising the intestinal lumen in a minimally invasive manner. Numerous studies have proved that VCE has better sensitivity compared to other radiological methods.¹⁰ However, its inability to localise the tumour accurately and to take biopsies from the tumour tissue are considered the most important drawbacks of VCE. The capsule may get stuck inside the narrowed intestinal lumen and this is another limitation of this diagnostic procedure.

Owing to the risk of intestinal ischaemia and malignancy, surgery is recommended for the treatment of invagination in adult cases.¹¹ The type and timing of the surgery may vary depending on the location, aetiology and obstruction grade of the invagination. The present case underwent emergency surgery because of the ileus symptoms, and no conservative treatment

was administered. Adjuvant chemotherapy is recommended in the presence of regional lymph node involvement or distant metastasis in intestinal adenocarcinoma cases. Most colorectal cancer chemotherapy regimens may also be used in different combinations for the treatment of intestinal adenocarcinoma. Since most intestinal adenocarcinomas are diagnosed in the advanced stages of the disease (74% are diagnosed at stage III/IV), its prognosis is poor. Overall, 5-year survival is around 15–42%.¹² As histopathological examination did not reveal any regional lymph node involvement in the present case, post-operative adjuvant chemotherapy was not administered.

Contributors CS took part in design of the study and writing of the manuscript. BO took part in supervision and design of the study. TS took part in literature research. NC took part in literature research and writing of the manuscript.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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Learning points

- Although invagination is rarely seen among patients with coeliac disease, it must be considered in patients with abdominal pain, especially among those whose adherence to a gluten-free diet is poor.
- In these cases, underlying malignancies such as adenocarcinomas must be excluded.
- The duration of the developmental stage of intestinal malignancies in coeliac disease is not exactly known; therefore, intestinal cancers must be kept in mind especially among patients whose adherence to a gluten-free diet is low.
- Adult intestinal invagination cases must be treated with surgery because of the risk of intestinal ischaemia and malignancy.

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