Reminder of important clinical lesson

A wolf in sheep's clothing

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Carcinoid tumours are often difficult to diagnose because of non-specific symptoms, mimicking those of irritable bowel disease or Crohn's disease. The authors describe a 57-year-old patient with clinical and radiological features, suggestive of Crohn's disease, in whom the diagnosis was proved to be a carcinoid tumour involving the terminal ileum. This case report illustrates that ileal carcinoid should be considered in the differential diagnosis of Crohn's disease, particularly in older patients.

BACKGROUND

Carcinoid tumours, although rare, are the most common distal small bowel malignancy and generally arise in the appendix or terminal ileum.^{1 2} Although the majority are asymptomatic, they may cause symptoms due to

obstruction or intussusception.³ Hormonal manifestations, called carcinoid syndrome, are rare.² Clinical features and radiological findings may mimic Crohn's disease. Given this, the condition is often diagnosed late following disease progression, by which time prognosis is poor.⁴

Figure 1 Small bowel conventional barium study showing an irregularly narrowed long segment of the ileum (arrows).

Figure 2 Axial reconstructed multidetector CT images of the abdomen showing a solid mass in the mesentery (thick arrow). Fine tissue strands (desmoplastic reaction) in the periphery of the mass infiltrate the surrounding mesentery and the small bowel (thin arrows).

CASE PRESENTATION

A 57-year-old man presented with a 6-week history of lower abdominal pain with colicky attacks and transient watery diarrhoea associated with weight loss and malaise. His medical history included partial epilepsy and a cerebral vascular accident. Valproinic acid was the medication given. Physical examination revealed a palpable mass in the lower abdomen.

INVESTIGATIONS

Routine blood tests and ileocoloscopy were normal. A small bowel study (figure 1) and a CT scan (figures 2 and 3) were performed.

Small bowel study showed an irregularly narrowed long segment of the ileum (arrows). The distal ileum was free of disease. The CT images of the abdomen showed a solid spiculated mass in the mesentery.

DIFFERENTIAL DIAGNOSIS

Initial diagnosis was of Crohn's disease.

TREATMENT

Treatment was started with mesalamine and oral prednisolone, but with no effect.

OUTCOME AND FOLLOW-UP

Because the images were considered not completely typical for Crohn's disease, the patient underwent a diagnostic laparotomy that revealed an 8-cm mass involving the mesentery and retroperitoneum, with ischaemic small bowel loops. Histology confirmed a malignant carcinoid tumour. Postoperative urinary 5-hydroxyindol acetic acid levels and octreotide scintigraphy were normal. There were no signs of distal metastasis. A few weeks later, a radical resection of the carcinoid process was performed with resection of the ileum and a right hemicolectomy with jejunotransversotomy and partial resection of the right ureter. Five years later, he is still doing well.

DISCUSSION

Carcinoid tumours, although rare, are the most common distal small bowel malignancy and generally arise in the appendix or terminal ileum.^{1 2} They are well-differentiated neuroendocrine tumours. They are often dormant until late in the disease and are thus locally advanced at presentation, by which time prognosis is poor.

Although the majority are asymptomatic and are found coincidentally at appendectomy, they may cause symptoms due to obstruction or intussusception.³ Clinical features and radiological findings may mimic Crohn's disease.

Figure 3 Coronal CT images of the same mass shown in figure 2.

This can cause a delay in the diagnosis. It has been reported that approximately 2.3% of patients with ileal carcinoid are first diagnosed and treated for Crohn's disease.⁵

The presence of an enhancing mass in the mesentery of the ileum with an extensive desmoplastic reaction, which can be seen on CT or MRI images, is a good clue to the diagnosis. Other signs of Crohn's disease and not of carcinoids include fissures and ulcers, fistulae sinuses and inflammatory polyps.

Learning points

- ▶ Carcinoid tumours are often difficult to diagnose because of non-specific symptoms mimicking those of irritable bowel disease of Crohn's disease.
- ▶ Ileal carcinoid should be considered in the differential diagnosis of Crohn's disease, especially in older patients presenting with obstructive symptoms.
- ▶ The presentation of an enhancing mass in the mesentery of the ileum with an extensive desmoplastic reaction, which can be seen on CT or MRI images, can be a good clue to the diagnosis.
- ▶ Laparotomy should be considered early, especially when the diagnostic uncertainty remains.

Prognosis can be dramatically improved with early diagnosis and surgical resection can be curative in this stage. Therefore, it is imperative to keep this differential diagnosis in mind.

This case report illustrates that ileal carcinoid should be considered in the differential diagnosis of Crohn's disease, especially in older patients presenting with obstructive symptoms. Laparotomy should be considered early in the course, especially in case of diagnostic uncertainty.

Competing interests None.

Patient consent Obtained.

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