Unusual association of diseases/symptoms

Metastatic carcinoid tumour mimicking cholecystitis, and a rare case of intussusception

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This report describes an acute presentation of obstructive jaundice, with a clinical picture of cholecystitis. A primary carcinoid tumour in the terminal ileum with hepatic secondaries was found to be the cause. Additionally, in the terminal ileum was a closely associated lipoma leading to an ileo-caecal intussusception. There are few such cases in the literature, particularly in the absence of any changes in bowel habit or lower abdominal pain. The majority of cases of intussusception in clinical practice occur in the paediatric population. Of the small numbers (<5%) that occur in adulthood, the underlying aetiology is most commonly a primary adenocarcinoma, with a far smaller number being attributable to lipoma, lymphoma and polyps.

BACKGROUND

This was a rare presentation of a carcinoid tumour, and many other conditions were suspected before the diagnosis was made. The clinical picture at the outset was far more akin to cholecystitis, in the context of examination findings and biochemical tests. It highlights the importance of including carcinoid tumours among the differentials for all patients presenting with a mass in the small bowel, and to consider it in the context of liver metastases. Additionally, this case was significant as the patient had the added pathology of an adult-onset intussusception due to a lipoma in the terminal ileum, which accounts for less than 5% of all cases.

CASE PRESENTATION

A female patient in her late 60s presented to the emergency department with sudden onset on epigastric pain. She had also noticed yellow sclerae and darker urine over the past week. Her only past medical history of note was an episode of paroxysmal atrial fibrillation 20 years previously.

On examination, she was comfortable at rest but clinically jaundiced. She was afebrile, blood pressure was 190/110 and heart rate was 72 bpm (regular rate and character). Respiratory and cardiovascular examinations were unremarkable. There was hepatomegaly to 3 cm below the costal margin and right upper quadrant tenderness. Murphy's sign was positive.

INVESTIGATIONS

Initial blood investigations revealed deranged LFTs consistent with a mixed obstructive/hepatic picture, with markedly raised bilirubin, alkaline phosphatase and transaminases. Inflammatory markers were also significantly elevated.

An abdominal ultrasound scan performed on the day of admission demonstrated a large echogenic focus in the right lobe of the liver, and no gallstones (figure 1). An urgent CT scan demonstrated a loculated mass in the right lobe of the liver and a smaller mass in the left lobe. These were associated intrahepatic duct dilatation and an incidental finding of a mass in the terminal ileum, surrounded by

Figure 1 Abdominal ultrasound images of (left) loculated, necrotic mass in right lobe of the liver, measuring 11.92 cm in long axis; (right) smaller, 3.93 cm mass in left lobe.

Figure 2 Axial images from contrast-enhanced computerised tomography of the abdomen and pelvis. (A) A tri-loculated, hypodense mass in the right lobe of the liver as seen on the previous ultrasound scan, (B) peri-ileal lymph node mass (circled), (C) a tumorous mass in the terminal ileum and (D) the mass in the terminal ileum leading to an intussusception of the terminal ileum into the caecum.

Figure 3 Resected specimen of terminal ileum and proximal caecum. A lipoma causing an intussusciens is noted at the bottom right of the specimen. There are two carcinoid tumours, one in the centre of the image and another surrounded by an area of necrosis to the left. Multiple tubulovillous adenomata are noted.

multiple lymph nodes, which was intussuscepting into the caecum (figure 2).

DIFFERENTIAL DIAGNOSIS

Initially the differential diagnosis centred on cholelithiasis and included cholecystitis, cholangitis and pancreatitis. Following the investigations and imaging, the differential diagnosis included colonic carcinoma with hepatic metastases, and metastatic carcinoid tumour. An ultrasound-guided biopsy of the liver mass was performed but histology was inconclusive. Twenty-four-hour urinary 5-hydroxyindoleacetic acid (5-HIAA) was raised significantly at 3818 u/ml. This made the likelihood of a primary carcinoid tumour far more likely.

TREATMENT

Following discussion at the local multidisciplinary team meeting, a laparoscopic right hemicolectomy with resection of the terminal ileum was performed. This confirmed the diagnosis of metastatic carcinoid tumour. The cause of the ileo-caecal intussusception was a lipoma in the terminal ileum (figure 3).

OUTCOME AND FOLLOW-UP

Postoperative echocardiography demonstrated valvular regurgitation and an ejection fraction of <30%, consistent with carcinoid heart disease. The patient was referred urgently to a tertiary centre for further management but unfortunately was deemed unfit for systemic treatment and died a few months later.

DISCUSSION

Carcinoid tumours are a relatively rare, diverse group of tumours derived from neural crest cells.¹ These tumours do not fit the classical definition of cancer, although it is well-known that they can have considerable metastatic potential and may be resistant to systemic treatment.² They can be either 'non-functional' or 'functional' tumours; the latter by definition are capable of producing 5-hydroxytryptamine (5HT) among various other peptides. This is consequently metabolised to 5-HIAA, which is excreted in the urine and can be used as a marker for disease.³ They are the commonest neuroendocrine tumours of the gastrointestinal tract, most often found in the appendix and terminal ileum. In the gut, they arise from enterochromaffin cells found in the crypts of Lieberkühn throughout the small bowel. They commonly present in the sixth or seventh decade of life, and tend to be heralded by symptoms of small bowel obstruction.

Development of the carcinoid syndrome in these patients, a constellation of systemic features attributable to the marked rise in circulating humoral factors, is rare, occurring in around 5–7%. Carcinoid heart disease however can occur in up to 70% of patients and usually comprises pulmonary stenosis or tricuspid incompetence.^{4 5}

Intussusception is the projection of a proximal segment of hollow viscus into the lumen of the adjacent segment. Its incidence in adults on the whole is rare, and unlike in the paediatric population is usually secondary to an organic cause (>95%) rather than purely idiopathic.⁶ They can clinically be categorised by either anatomical location (eg, ileo-caecal, as in this case) or by aetiology (benign, malignant, idiopathic).^{7 8} Of the commoner, 'secondary' intussusceptions, up to 30% are due to a malignant cause, that is, adenocarcinoma of the small/large bowel,⁹ with lipomas, polyps and Meckel's diverticulum being among the commonest benign causes. The classical presentation in childhood (severe abdominal pains, bloody stool and a palpable mass) is extremely rare in adulthood, with the symptoms more akin to a chronic subclinical obstruction.⁸ This case was unusual in the sense that the degree of intussusception was quite marked, yet was not causing any symptoms.

Learning points

- ▶ Consider carcinoid tumours among differentials in patients with small bowel tumours.
- ▶ 24-h urinary 5-HIAA is a useful and simple test to perform and will assist in the diagnosis.
- ▶ Carcinoid disease can have primary tumours in many locations, and can spread to many organs.
- ▶ The carcinoid syndrome is a diffuse, multisystem disease.
- ▶ Intussusceptions are uncommon in adults.

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