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ARTICLE

Diagnosis and staging of small bowel tumours

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Abstract

Small bowel neoplasms comprise only 1% of gastrointestinal neoplasms. Despite their rarity, it is important to diagnose small bowel tumours early to maximize patient survival.

Keywords: *Intestinal neoplasms; adenocarcinoma; lymphoma; gastrointestinal stromal tumours; carcinoid; intestinal CT.*

Introduction

Although the small bowel constitutes over 75% of the length and 90% of the mucosal surface of the alimentary tract, it is the site of only 1% of gastrointestinal carcinomas. The American Cancer Society estimates that 5100 persons in the United States will develop cancer of the small bowel in 2006, with 1340 deaths.

Early diagnosis of small bowel tumours is a diagnostic challenge for both clinicians and radiologists. Patients present with non-specific symptoms such as abdominal pain, weight loss and gastrointestinal bleeding.

Adenocarcinoma

Adenocarcinoma is the most frequent primary small bowel tumour, accounting for 40% of these neoplasms[1]. More than half of small bowel adenocarcinomas arise in the duodenum, even though this organ comprises only 4% of the entire length of the small bowel. Most arise in the region of the ampulla of Vater. A smaller percentage of tumours arise in the jejunum, particularly in the first 30 cm distal to the ligament of Treitz. Ileal carcinomas are the least common, except in patients with Crohn's disease[1].

On barium studies, advanced small bowel adenocarcinomas appear as "apple core" lesions similar to those found elsewhere in the gastrointestinal (GI) tract. These annular lesions are short, circumferentially narrowed segments with mucosal ulceration and overhanging proximal and distal borders. These malignant strictures are usually rigid, with a fixed, unchanging appearance during compression.

On computed tomography (CT), small bowel adenocarcinomas may manifest as a discrete tumour mass, annular narrowing (Fig. 1) with abrupt concentric or irregular "overhanging edges", or as an ulcerative lesion. Duodenal adenocarcinomas tend to be papillary or polypoid, while more distal lesions tend to be annular. Usually only a short segment of the bowel is involved and gradual narrowing of the lumen leads to partial or complete small bowel obstruction. The mass itself usually shows moderate enhancement after the intravenous administration of contrast material[2–4].

Carcinoid

These well differentiated endocrine tumours arise from the enterochromografin cells at the base of the crypts of Lieberkuhn. Approximately 70% of GI tract carcinoid tumours arise in the appendix and are virtually always benign. The small bowel is the second most frequent location for carcinoid tumours and they tend to congregate in the distal ileum, with nearly 40% located within 60 cm of the ileocaecal valve[1].

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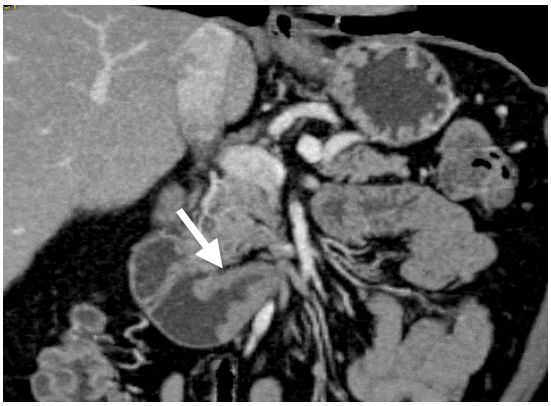

Figure 1 **Duodenal adenocarcinoma produces annular narrowing of the third portion of the duodenum on this coronal, reformatted image.**

Small bowel carcinoids begin as small polypoid masses but have a tendency to invade into the submucosa and eventually infiltrate the muscularis propria, serosa, and then invade mesenteric lymph nodes[1].

The majority of GI carcinoids are found within 60–90 cm of the ileocaecal valve: 50% in the appendix and 33% in the small bowel. Carcinoids account for approximately 50% of all appendiceal tumours and are found in 0.03%–0.69% of appendectomy specimens. Over 90% are less than 1.5 cm in size and there is a 99% 5-year survival rate for appendiceal carcinoids. Small bowel carcinoids that come to surgery are usually small, 1–2 cm, submucosal-intramural tumours that bulge slightly into the lumen. As they enlarge, they can become polypoid and cause intussusception or obstruction[1].

Tumours less than 2 cm in size are usually discovered incidentally. They appear as smooth, rounded, 1–2 cm elevations in the distal ileum. The tumours may be mucosal or submucosal in appearance, without desmoplastic changes, and be indistinguishable from other tumours such as gastrointestinal stromal tumours (GISTs), lipomas, adenomas, submucosal metastases and lymphoma. When submucosal carcinoids ulcerate, they produce a "target" or "bull's-eye" appearance that can be seen with metastatic melanoma or breast cancer, lymphoma, and Kaposi's sarcoma[5].

Mural thickening and thickening of the valvulae conniventes is initially seen on barium studies. As carcinoid grows into the mesentery, specific radiographic features can be detected: crowding of folds at the edge of the tumours, kinking of the bowel wall, narrowing of the lumen, tethering, fixation, and angulation of the involved small bowel loops, and occasionally an annular constricting lesion. Exocentric masses cause separation and displacement of adjacent bowel loops. Retraction of the small bowel loops towards the root of the mesentery can also be identified. Segmental small bowel ischemia producing regularly thickened valvulae conniventes may result if the mesenteric vessels are encased, resulting in bowel ischemia[5].

When confined to the bowel wall, small carcinoid tumours are difficult to detect on routine CT scans. The introduction of multidetector CT (MDCT) permits visualization of even small carcinoids as robustly enhancing submucosal lesions. Visualization of these lesions is improved when water or other neutral contrast is given as an intraluminal contrast agent and if multiplanar reconstructions are obtained[6].

MDCT is superb in delineating mesenteric extension of carcinoid tumours and liver metastases. CT shows an ill-defined mesenteric mass that contains calcification in up to 70% of cases. This mass has a spiculated, stellate, and infiltrative appearance (Fig. 2). This spiculation does not usually indicate tumour invasion but is related to the desmoplastic changes that occur in the mesentery in response to secretion of serotonin and tryptophan. The mesenteric vessels may be involved as a direct result of tumour encasement and narrowing, or indirectly as a result of the vasoactive hormones that affect the vessel wall[6].

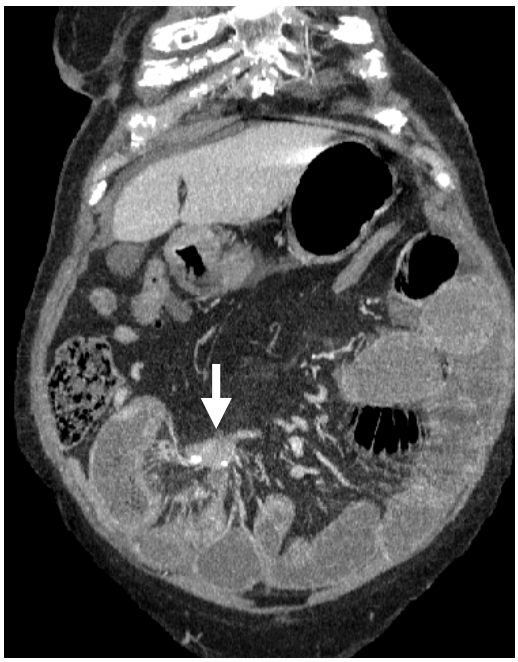

Figure 2 **This coronal reformatted image shows a spiculated, calcified carcinoid tumour (arrow) causing distal small bowel obstruction.**

Three-dimensional CT angiography is very useful in depicting the full extent of the mesenteric mass and its relationship to the blood vessels, important factors in preoperative planning[6].

On magnetic resonance, carcinoid tumours cause focal, asymmetric bowel wall thickening. On unenhanced scans, these lesions are isointense to muscle on T1-weighted images and iso or mildly hyperintense to muscle on T2-weighted images. The primary lesions will show contrast enhancement. Mesenteric masses range between 2 and 4 cm and are typically isointense to muscle on

T1- and T2-weighted images. Desmoplastic stranding manifests as hypointense strands[7].

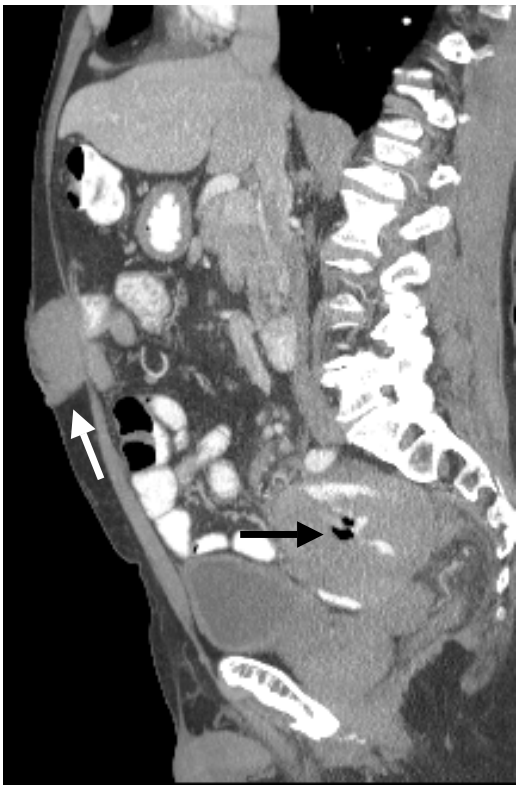

Figure 3 **Sagittal reformatted image shows a large, homogeneous, non-obstructive, ulcerating ileal lymphoma (black arrow). Note the enlarged periumbilical lymph node (white arrow).**

Lymphoma

Reflecting the diverse morphology of the disease, small bowel lymphoma has a great variety of radiologic appearances. The principle features are an infiltrating form, a polypoid form, multiple nodular defects, and an endoenteric–exoenteric with ulceration and fistula formation[1].

Circumferentially infiltrating lymphoma involves a variable length of small bowel with thickening and later effacement of folds. The lumen is more often widened than narrowed, so called "aneurysmal dilatation". This implies a localized, non-obstructive dilatation of the bowel caused by destruction of the muscle wall and myenteric plexus by infiltrating tumour[1].

On CT, lymphoma of the small bowel typically presents as a homogenous density mass causing mural thickening and separation of adjacent small bowel loops. These lesions tend to be more homogenous and show less contrast enhancement than other malignant tumours[8–10].

Deep ulceration within a large lymphomatous mass arising from the small bowel but with exocentric growth, constitutes the cavitary endoenteric–exoenteric lesion (Fig. 3). Non-Hodgkin's lymphoma of the small bowel does not evoke a desmoplastic reaction so that bowel obstruction is rare even though the associated tumour mass may be large compared to adenocarcinoma[8–10].

Lymphomatous proliferation that is predominantly submucosal produces a nodular pattern, creating multiple filling defects on barium studies. The nodules may be small and diffuse but more commonly are variable in size and number. A solitary polypoid lymphoma may serve as the lead point for an intussusception.

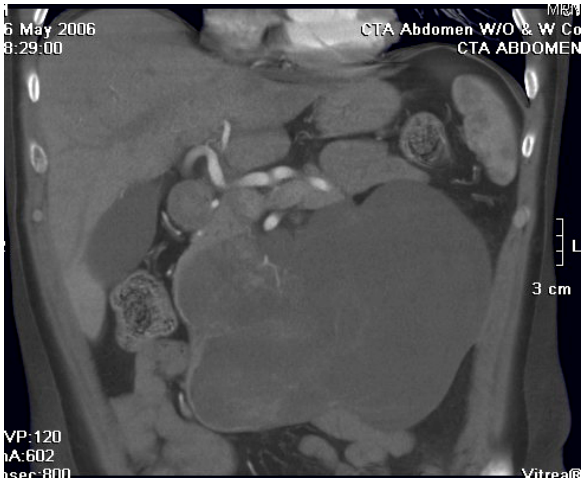

Figure 4 **Large, predominantly cystic GIST tumour causing mass effect but no obstruction in the left upper quadrant.**

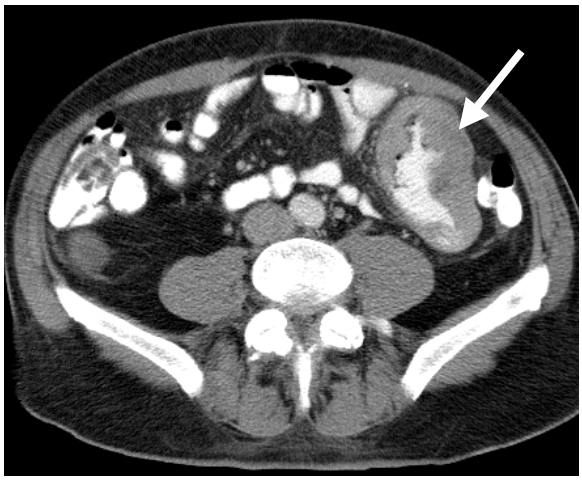

Figure 5 **Metastatic melanoma to the distal jejunum causing a large, ulcerating mass (arrow).**

Gastrointestinal stromal tumours (GISTs)

GISTs are usually solitary tumours that have a wide spectrum of histologic features. They can occur anywhere in the GI tract and any portion of the gut wall. Most are gastric in origin and centred within the submucosa or

muscularis mucosa. The small bowel is the second most common site of GISTSs; nearly one third arise in this location. Although these tumours can develop anywhere in the small bowel, a disproportionate number arise in the duodenum. Unlike gastric GISTs, those that occur in the small bowel are composed of spindle cells and as a group, a higher percentage are malignant[1].

On barium studies, submucosal GISTs appear as smooth, round filling defects that may or may not ulcerate. Subserosal GISTs are extrinsic or exocentric masses that displace adjacent bowel loops. Intraluminal GISTs may be hypervascular and cause haemorrhage and ulceration.

On CT, GISTs present as a mass that can be smooth, irregular or lobulated in appearance (Fig. 4). The tumour may have regions of low attenuation, central liquifactive necrosis with or without air–fluid levels, ulceration, calcification or direct extension into adjacent organs and vascular encasement[11–13].

The malignant potential of these lesions cannot be stated with certainty on cross sectional imaging. Some believe that all of these tumours should be considered to be of at least low-grade malignancy.

Metastases

Metastases account for approximately 50% of all small bowel neoplasms. Indeed in a patient with a known neoplasm, a small bowel neoplasm is most likely a metastasis. The most common metastases are from primary malignancies of the colon, stomach, pancreas, melanoma, breast and lung. Metastases develop through four major pathways: direct extension, intraperitoneal seeding, lymphatic spread, and haematogenous metastases (Fig 5)[14].

Summary

Small bowel neoplasms are rare. They often have overlapping imaging features. The differential diagnosis can be focused when typical imaging features are seen in specific locations. A polypoid or constricting annular mass in the duodenum suggests adenocarcinoma. A mesenteric mass with calcifications and spiculations suggests carcinoid tumour. A large, homogeneous mass with mural thickening and lumen dilation in the ileum suggests lymphoma. Lymphoma, metastases and GISTs can cause large masses.

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