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Small Intestinal Carcinoma

1. GROSS DESCRIPTION

Specimen

— endoscopic/laparoscopic or open biopsy/resection. Whipple's pancreaticoduodenectomy, segmental bowel resection, right hemicolectomy: depending on the tumour site in the proximal/mid-/distal small bowel, respectively. Needle core biopsy of a small intestinal or mesenteric mass is usually avoided due to the risk of capsule rupture and tumour seeding jeopardizing complete primary resection, suitability for which is assessed by CT scan.
— weight (g) and size/length (cm), number of fragments.

Tumour

Site

— duodenum (particularly periampullary) 70%: see Chapter 3.
— jejunum/ileum 30%.
— mucous membrane/muscularis/extramural.
— serosal/mesenteric/nodal/single/multifocal.
— mesenteric/anti-mesenteric border.
— Meckel's diverticulum.

Size

— length × width × depth (cm) or maximum dimension (cm).

Appearance

— polypoid/sessile/ulcerated/diffusely infiltrative/fleshy/pigmented/yellow/stricture/intussusception ± secondary ischaemic necrosis of the tumour tip/intussusceptum or receiving segment (intussuscipiens).

Duodenal carcinomas tend to be papillary or polypoid, distal carcinoma ulcerated and annular with constriction of the bowel wall (napkin ring-like). Presentation can be non-specific, e.g. anaemia or weight loss, with poorly defined central abdominal pain or signs of subacute obstruction. There may be a detectable mass either on abdominal examination or CT scan. Carcinoid tumour is nodular, yellow, uni-/multifocal causing

bowel obstruction due to fibrosis or acting as the apex of an intussusception. Malignant lymphoma can be subtle in the edge of a perforated jejunitis or a fungating, fleshy mural or mesenteric mass. There may be a preceding history of coeliac disease. Metastases are often serosal seedlings, nodules or plaques. GISTs are mural lesions which can be dumb bell-shaped with luminal and extramural components. They can also be separate from the bowel and mesenteric in location.

Edge
— circumscribed/irregular.

2. HISTOLOGICAL TYPE

Adenocarcinoma
— enteric pattern, well or moderately differentiated: usual type.
— anaplastic (poorly differentiated) forms also occur more frequently than in colorectal cancer.
— mucinous carcinoma: >50% of the tumour area.
— signet ring cell carcinoma: >50 % of the tumour cells.

Diagnosis of primary small intestinal adenocarcinoma is by exclusion of spread from more common sites, e.g. colorectum and stomach. Similar to the large intestine there is some evidence for a dysplasia (adenoma)–carcinoma sequence in the adjacent mucosa. Prognosis is poor due to late presentation and advanced stage.

Carcinoid tumour
— yellow/nodular/±multifocal.
— chromogranin/synaptophysin/CD56 positive.
— typically insular pattern of uniform cells in a dense fibrous stroma with vascular thickening.
— 20% have carcinoid syndrome implying liver metastases: facial flushing/asthma/thickening of cardiac valves due to release of the vasoactive peptides (e.g. serotonin) into the systemic circulation.
— low-grade malignancy: any functioning well-differentiated tumour; any tumour with angioinvasion; non-functioning tumour ≥2 cm or with invasion beyond the submucosa.
— high-grade malignancy: tumour with a high mitotic rate, cellular atypia or necrosis and poorly differentiated tumours/small cell carcinomas.

Carcinoid tumour has an overall 5-year survival rate of 50–65%. It is better for small lesions [metastatic rate: <1 cm (2%), 1–2 cm (50%), >2 cm (80%)] confined to the wall (85%) than those invading the serosa or beyond (5%). Metastases are to regional nodes and liver (multiple, solid/cystic): also bone, skin and thyroid. The above comments relate mostly to classical EC cell jejuno-ileal carcinoids. Duodenal carcinoids have a better prognosis, occur mainly in D1/D2 and include non-functioning G cell tumours, gastrinomas (Zollinger Ellison/MEN syndromes), somatostatinoma and gangliocytic paraganglioma.