Small intestine

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Incidence Midgut carcinoids are the most common NETs with a peak age of diagnosis at 60–70 years. They are the second most common small bowel malignancy . Pathology They arise from the enterochroma ffi n cells and secrete sero tonin and substance P . They are either solitary or multiple (30%) (Figure 57.17) and the primary tumour can be small (5–20 /uni00A0 mm). They are often indolent but often discovered at an advanced stage when regional (lymph nodes; 36%) and distant (liver; 48%) disease is pr esent. From the latter, the patient may develop carcinoid syndrome. Clinical presentation Chronic abdominal pain is the most frequent initial symptom. The carcinoid syndrome is seen in 20–30% of patients with liver metastases. Small bowel ischaemia producing pain and diarrhoea arises from involved lymph node metastases, which constrict the mesenteric vessels. However, up to 20% of patients are diagnosed during the investigation of liver metastases or). they may be incidentally found during surgery for another reason (laparotomy or appendicectomy). Diagnosis - Chromogranin A is often elevated (non-specific) and elevated 24-hour urinary 5-HIAA is specific. Cross-sectional CT or MRI often shows the mesenteric lymph nodes with a char - acteristic spiralling of vessels trapped in the desmoplastic reaction (Figure 57.18) and will demonstrate if liver metas - tases are present. Biopsy is diagnostic and Ki-67 grading is an important prognostic factor and is mandatory for reporting. This is often via an ultrasound-guided liver biopsy . In the search for a primary tumour, CT and/or MRI is follow ed by 68 Ga-DOTATOC PET , fused with CT . Patients must be evaluated for the presence of Hedinger syndrome (right heart valve fibrosis). A referral is made to the cardiology department, where transthoracic echocardiography will be carried out.

Figure 57.17 Multiple neuroendocrine tumours in the small bowel. Figure 57.18 Contrast-enhanced (bowel) cross-sectional computed tomography scan with mesenteric lymph node metastasis drawing in the adjacent mesenteric vessels in the desmoplastic reaction (white circle).

Stratified treatment of NETs based on stage /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF Treatment ENETS (European Neuroendocrine Tumor Society) has developed an algorithm for the treatment of small intestine NETs (Summary box 57.3). Surgical Patients without distant metastases (stages I–III) are all poten tial candidates for curative surgery of the primary tumour and regional nodal metastases. All patients should be discussed in a multidisciplinary team meeting. Surgery is only undertaken if it is thought that an R0 resection can be achieved. The limiting factor is often lymph node involvement around the superior mesenteric artery , especially in the presence of severe desmoplastic reaction. Concomitant cholecystectomy should be considered owing to the risk of gallstone formation second ary to SSAs. In the presence of stage IV (metastatic disease) surgery is contemplated when the patient has obstructive symptoms (pal liative) or if an R0 resection (curative) can be achieved with concomitant liver metastasectomy(ies). In this setting, patients should ha ve peroperative protection with intravenous SSAs to avoid a carcinoid crisis. When curative intent is not possib le, there are a

number of locoregional therapies to the liver. Patients are always treated with SSAs to prevent a crisis. Therapeutic options include radiofr equency ablation, embolisation and chemoemboli sation. Liver transplantation is only considered in a highly selected population (see also Chapters 69 and 74). Medical SSAs are an e ff ective treatment for syndrome control for functional NETs. SSAs may also be used for antiproliferative purposes in stable or progressive disease in tumours up to a Ki-67 index of 10% (grades I and II). Loperamide or other similar agents may be useful in the control of diarrhoea. When SSA treatment fails, a second-line option is PRRT . Currently yttrium- or lutetium-labelled SSAs are most frequently used. For grade III tumours (NEC), cisplatin-based systemic chemo therapy is the treatment of choice. Paul Wermer , 1898–1975, physician, The Presbyterian Hospital, New Y ork, NY , USA, described this condition in 1954. /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF

Disease Localised Regional Distant status Stage I/II III (N1) IV (M1) Surgical Radical resection Radical – curative intent approach Resection of Resection of primary primary nodes (along mesentery) nodes (along mesentery) liver metastases Aim R0 R0 Palliative resection No resection Resection of Due to primary irresectable nodes (along mesentery) comorbidities liver metastases To avoid obstructive complications (R1)

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