Benign

Benign

The majority of small bowel neoplasms are benign, comprising adenomas, lipomas, haemangiomas and neurogenic tumours. They are frequently asymptomatic and identified incidentally , James Israel , 1848–1926, first found sulphur granules in pus from a discharging sinus in a man’s neck. Later, he became a famous Berlin urologist. John Law Augustine Peutz , 1886–1968, Chief Specialist for Internal Medicine, St. John’s Hospital, The Hague, The Netherlands. Harold Joseph Jeghers , 1904–1990, Professor of Internal Medicine, The New Jersey College of Medicine and Dentistry , Jersey City , NJ, USA.). - but can present with intussusception, small bowel obstruction and bleeding that may cause anaemia or may even be overt. Where these lesions do cause anaemia, the cause can be di ffi cult to diagnose, as CT or small bowel contrast studies do not show them easily . Capsule endoscopy or small bowel endoscopy have been used successfully where the facilities exist. Symptomatic lesions can be treated by small bowel resection. Peutz–Jeghers syndrome This is an autosomal dominant condition characterised by melanosis of the mouth and lips, with multiple hamartoma - tous (benign tumour-like malformations resulting from faulty development in an organ) polyps in the small bowel and colon (Figure 74.3). Melanin spots can also occur on the digits and perianal skin. Mutation of the STK11 gene on chromosome 19 has been found in a proportion of patients. Long-term follow-up of the original family described by Peutz has shown reduced survival as a consequence of complications of bowel obstruction and the development of a range of cancers. Regular colonic surveillance should be performed and female patients should attend breast and cervical screening. Despite the increased risk of malignancy in general, malignant change in the polyps themselves is uncommon. Resection may be indi - cated, however, for heavy and persistent or recurrent bleeding or intussusception. Polyps may be removed by enterotomy or, at laparotomy , snared via a colonoscope introduced via an enterotomy . Heavily involved segments of small intestine may occasionally be resected.

Figure 74.3 Melanin spots on the lips of a patient with Peutz–Jeghers syndrome (courtesy of Major PCM Manta, Indian Medical Service).

Small bowel malignancy is rare, classically presents late and is most often diagnosed after surgery for small bowel obstruction. Four types will be considered, which account for over 99% of small bowel malignancies: adenocarcinoma, neuroendocrine tumours (NETs), lymphomas and gastrointestinal stromal tumours (GISTs). Adenocarcinoma Small bowel adenocarcinoma is more often found in the jeju num than in the ileum and, although the aetiology is unknown, it is more common in patients with CD, coeliac disease, familial adenomatous polyposis, hereditary non-polyposis colon cancer and Peutz–Jeghers syndrome. The tumours present with anaemia, overt g astrointestinal bleeding, intussusception or obstruction. Prognosis is poor, particularly in patients with CD, in whom these tumours often present late because the symptoms are commonly mistaken for those of CD and treated conservatively . When diagnosed, surgical treatment is a resec tion of 5 /uni00A0 cm of non-involved bowel either side of the lesion and the a ff ected mesentery (Figure

74.4). A right hemicolectomy is likely to be required for tumours of the distal ileum. Neuroendocrine tumours NETs (previously known as carcinoid tumours) occur through out the gastrointestinal tract, most commonly in the appendix, ileum and rectum in decreasing order of frequency . They arise from Kulchitsky cells at the base of intestinal crypts (of Lieberkuhn). The primary is usually small, although significant lymph node metastases can occur. In up to one-thir d of cases of small bowel NETs, the tumours are multiple. They may produce dense fibrosis in the surrounding tissues, resulting in distortion and scarring of the bowel and associated mesentery , giving them a characteristic radiological appearance. NETs can produce a number of vasoactive peptides, most commonly 5-hydroxytryptamine (serotonin), but also histamine, prosta glandins and kallikrein. When they metastasise to the liver, the ‘carcinoid syndrome’ can become evident because the vasoactive substances escape the filtering actions of the liver. The clinical syndrome itself consists of reddish-blue cyanosis, flushing attacks, diar rhoea, borborygmi, asthmatic attacks and, eventually , pulmonary and tricuspid stenosis (Summary box 74.3). Classically , the flushing attacks are induced by alcohol. Surgical resection is usually su ffi cient for patients with pri mary disease, but the incidence of recurrence is significant. Summary box 74.3 Carcinoid syndrome /uni25CF /uni25CF /uni25CF /uni25CF /uni25CF Nikolai Kulchitsky , 1856–1925, Professor of Histology , Kharkov , Ukraine, who left Russia after the Revolution of 1917 and later worked at University College, London, UK. He described these cells in 1897. Johann Nathaniel Lieberkühn , 1711–1756, physician and anatomist, Berlin, Germany , described these glands in 1745. Thomas Hodgkin , 1798–1866, lecturer in morbid anatomy and curator of the museum, Guy’s Hospital, London, UK, described lymphadenoma in 1832. Denis Parsons Burkitt , 1911–1993, Irish-born surgeon who worked in Kampala, Uganda. - The extent of disease can be assessed preoperatively using oct - reotide scanning (somatostatin receptor scintigraphy), which - may detect otherwise clinically unapparent primary and sec - ondary tumours. Plasma markers of tumour bulk, such as chromogranin A concentrations, may be useful mar kers of disease recurrence, as well as of prognostic value. tic resection can be carried out in patients with meta - Hepa static disease. The treatment has been transformed by the use - of octreotide (a somatostatin analogue), which reduces both flushing and diarrhoea, and octreotide cover is usually used in tients with a carcinoid syndrome who have surgery to prevent pa a carcinoid crisis resulting from liberation of vasoactive sub - stances following handling of the tumour. NETs generally grow more slowly than most metastatic malignancies and patients may live with metastatic disease for many years. The tumour is not usually sensitive to chemo- or radiotherapy (see Chapter 69). Lymphoma Small bowel lymphoma may be primary or, more commonly , - secondary to systemic lymphoma. The incidence of small bowel lymphoma is increased in patients with CD and immunodeficiency syndromes. The classification of lymphoma is beyond the scope of this chapter but a number of points are worth noting briefly . It is rare for Hodgkin’s lymphoma to a ff ect the small bowel and most Western-type lymphomas are non-Hodgkin’s B-cell lymphomas. They usually present with anaemia, bleeding, perforation, anorexia and weight loss. - Small bowel T-cell lymphoma can develop in patients with coeliac disease. It usually presents with worsening of local the patient’s diarrhoea, pyrexia of unknown origin and obstructive symptoms. Mediterranean lymphoma is found mostly in North Africa and the Middle East and is often wide - spread at diagnosis. Burkitt’s lymphoma can aggressively a ff ect the ileocaecal region, particularly in children. The mainstay of treatment for these conditions is chemotherapy; however, sur - equired for obstruction, perforation or bleeding. gery may be r

Diarrhoea Palpitations Bronchospasm Tricuspid regurgitation Facial/upper chest /f_l ushing Figure 74.4 Small bowel adenocarcinoma.

GISTs are mesenchymal tumours and the distinction between benign and malignant types is di ffi cult even on histological examination. Increased size and high levels of c-kit (C D 1 1 7) staining are associated with malignant potential. GISTs are found most commonly in the stomach but can be found in other parts of the gut. They occur most commonly in the 50- to 70-year age group. Although the cause is unknown, patients with neurofibromatosis have an increased risk of developing these types of tumour. Patients may be asymptomatic and the tumour may present as an incidental mass on a CT scan. Symptoms include lethargy , pain, nausea, haematemesis or melaena. Surgery is the most e ff ective way of treating GISTs as the tumour is radioresistant and is not sensitive to conventional chemotherapy. Imatinib is a tyrosine kinase inhibitor that has been shown to be e ff ective in advanced cases and may also have a role in adjuvant treatment. It may be used preoperatively to reduce tumour size; however, the involuting tumour may perfo rate, precipitating a surgical crisis (see Chapters 11 and /uni00A0 67

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