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SUPPLEMENTATION TO THE ENCYCLOPEDIA

Tumours in the Small Bowel ☆ ☆



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Abstract

Small bowel tumours are rare and originate from a wide variety of benign and malignant entities. Adenocarcinomas are the most frequent primary malignant small bowel tumours. Submucosal tumours like gastrointestinal stromal tumours (GIST) or neuroendocrine tumours (NET) may show a central umbilication, pathologic vessels, bridging folds or an ulceration of the overlying mucosa. These signs help to differentiate them from harmless bulges caused by impression from outside, e.g. from other intestinal loops. Sarcomas of the small bowel are rare neoplasias with mesenchymal origin, sometimes presenting as protruding masses. Benign tumours like lipoma, fibrolipoma, fibroma, myoma, and heterotopias typically present as submucosal masses. They cannot be differentiated endoscopically from those with malignant potential as GIST or NET. Neuroendocrine carcinomas may present with diffuse infiltration, which may resemble other malignant tumours. The endoscopic appearance of small bowel lymphomas has a great variation from mass lesions to diffuse infiltrative changes. Melanoma metastases are the most frequent metastases to the small bowel. They may be hard to distinguish from other tumours when originating from an amelanotic melanoma.

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Video related to this article

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1. Background

- Neoplastic lesions of the small bowel occur rarely with predomination of benign tumours. Only 1-2% of all gastrointestinal malignant tumours concern the small bowel. Because of missing or unspecific early symptoms

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small bowel tumours are often diagnosed in advanced stage of disease.

- Capsule endoscopy can be helpful to detect small bowel tumours earlier and affect the therapeutic course [1,2].
- However, about 10-20% of small bowel tumours are missed by capsule endoscopy. A combination with computed tomography increases the diagnostic yield significantly [3].
- Typical clinical presentation in earlier stages is overt or occult midgastrointestinal bleeding. This applies to all malignant entities, while lymphomas have the lowest and gastrointestinal stromal tumours (GIST) the highest incidence of bleeding. Benign tumours have an almost equal bleeding potential with the exception of adenomas and aberrant pancreatic tissue, who rarely present with gastrointestinal bleeding [3].
- Further unspecific symptoms are iron deficiency anaemia of unknown origin, abdominal pain, weight loss and partial or total intestinal obstruction.
- In these cases VCE may lead to the correct diagnosis [4,5]. Balloon enteroscopy additionally enables biopsy, haemostasis, palliative dilation or stenting.
- Submucosal tumours have to be differentiated from harmless bulges due to impression from outside. Constant size, discoloration, central umbilication, or bridging folds are diagnostic hints for small bowel masses [6]. They can be benign mesenchymal tumours or have malignant potential like GIST and neuroendocrine tumours.
- Pathologic vessels may be a hint for malignancy, although systematic data are missing. Ulceration of the overlying mucosa may lead to midgastrointestinal bleeding [7].
- Adenocarcinomas are the most common primary malignant lesions of the small bowel. Approximately 50% of all adenocarcinomas arise in the duodenum and can be detected by upper endoscopy. They may appear as an infiltrating or exophytic lesion with ulcerations or stenosis [8].
- Gastrointestinal stromal tumours (GIST) are the most frequent malignant mesenchymal neoplasias of the small bowel. Size and degree of proliferation are predictive factors for malignancy. Endoscopic appearance is usually a submucosal mass, sometimes with ulceration of the mucosa. However, GISTs often show an eccentric extraluminal growth, leading to underestimation of the size by endoscopy.
- Sarcomas are rare malignant mesenchymal tumours, sometimes causing obstruction of the small bowel [9]. They may present as protruding masses, biopsy samples lead to the diagnosis [10].
- Lipomas are common benign submucosal tumours with a typical soft, deformable consistence and shallow yellow colour underneath normal mucosa. This colour is not as striking as in more flat, superficial lymphatic cysts. Fibrolipomas are a rare histological variant of lipomas resulting in a harder tissue. Ulcerations may cause bleeding.
- Neuroendocrine tumours (NET), which arise from enterochromaffine cells, are often highly differentiated and found incidentally. However, sometimes they may present with bleeding or obstruction [3]. Mid-gut NETs are preferably located in the appendix and ileum.
- Patients presenting with a carcinoid syndrome may suffer from a NET of the small bowel with hepatic metastasis

[11]. A complete small bowel examination via capsule endoscopy should be considered in search of the primary lesion.

- NET often present as submucosal tumours or multiple submucosal nodules. It is not possible to clearly distinguish them from other entities as benign mesenchymal neoplasias (e.g. myoma, fibroma) heterotopias (e.g. ectopic gastric or pancreatic mucosa), and potentially malignant tumours as GIST.
- NET may occur as multifocal submucosal tumours [12]. When affecting the mesentery only, the tumour itself may not be visible for capsule endoscopy. Red spots in a short intestinal segment can be indirect signs. CT scan may be helpful in these cases.
- Infiltrating neuroendocrine carcinomas cannot be differentiated endoscopically from other malignant tumours. Tissue sampling is required to clarify the subtype.
- Non-Hodgkin's lymphomas of the small bowel show a great endoscopic variation, sometimes presenting with ulcerative, nodular or infiltrative changes, but sometimes only with diminutive mucosal changes like erythema or lymphangiectasias [13]. A common endoscopic appearance is a firm, motionless mucosa. Perforation, obstruction and bleeding have been described as well [14].
- B-cell derived tumours account for the majority of small bowel lymphomas, while enteropathy associated T-cell lymphomas may develop as a complication of coeliac disease.
- While the majority of small bowel malignancies are primary lesions, some of them are of metastatic origin.
- Melanoma metastases are the most frequent metastatic tumours found in the small bowel [15]. Their dark pigmentation is diagnostic, while amelanotic types cannot be distinguished endoscopically from other tumours [16].
- Metastases from cancer of lung, breast, kidney, and other malignancies have been described as well.

2. Materials

- Video Capsule Endoscopes (PillCam SB1, PillCam SB2) (Given Imaging, Yokneam, Israel; EndoCapsule 1 (Olympus, Tokyo, Japan))
- Gastroscopes GIF Q160 (Olympus Tokyo, Japan)
- Double-balloon Enteroscope EN 450-T5 (Fujinon, Tokyo, Japan)
- Single-balloon Enteroscope SIF Q180 (Olympus, Tokyo, Japan)

3. Endoscopic procedure

- Small bowel Video Capsule Endoscopy (VCE)
- Gastroscopy
- Push Enteroscopy (PE)
- Single-balloon Enteroscopy (SBE)
- Double-balloon Enteroscopy (DBE)

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