Video Journal and Encyclopedia of GI Endoscopy (2014) 1, 632–635

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SUPPLEMENTATION TO THE ENCYCLOPEDIA

Tumours in the Small Bowel ☆☆

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Received 3 April 2013; received in revised form 12 August 2013; accepted 20 August 2013

KEYWORDS
Small bowel tumours;
GIST;
NET;
Melanoma;
Lymphoma;
Sarcoma;
Video

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.

Video related to this article

Video related to this article can be found online at http://dx.doi.org/10.1016/j.vjgien.2013.08.001.

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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2212-0971
http://dx.doi.org/10.1016/j.vjgien.2013.08.001

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 adenocarcimomas 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)

4. Key learning points and tips and tricks

4.1. Key learning points

- Small bowel tumours are rare and originate from a wide variety of benign and malignant entities.
- Typical clinical presentation in earlier stage is overt or occult midgastrointestinal bleeding. This applies to all malignant entities [3].
- Adenocarcinomas are the most common primary malignant neoplastic lesions of the small bowel.
- Submucosal tumours like GIST or NET have to be differentiated from harmless bulges. Central umbilication, pathologic vessels or an ulceration of the overlying mucosa are diagnostic hints for tumours.
- Lymphomas show a wide range of endoscopic findings, from diminutive lesions to large masses.
- Melanomas are the most frequent metastases to the small bowel.

4.2. Tips and Tricks

- In patients with recurrent GI bleeding in spite of normal conventional and capsule endoscopy, imaging techniques and balloon enteroscopy should be considered in the search for tumour besides repeating upper and lower GI endoscopy.
- Patients with mid-GI bleeding should be informed about the low risk of capsule retention due to possibility of underlying tumour. However, even in the unlikely event of retention this has rather to be considered as diagnostic help. Hence, prior performance of Patency capsule test in the absence of other risk factors is not helpful.
- In case of incidental diagnosis of an ileal NET, investigation of the entire small bowel might be considered, as these tumours may be multifocal. However, the therapeutic impact of this additional examination has yet to be determined.
- Presence of mucosal discoloration, ulceration, umbilication, bridging folds, pathologic vessels on the surface, constant form and sharp margins may be helpful to distinguish submucosal tumours from harmless bulges at capsule endoscopy.
- Tumours of the small bowel with extramural growth and metastasis are preferably detected by imaging techniques as CT scan and MRI, while endoscopic procedures may detect small intraluminal lesions earlier. Therefore a combination of radiological and endoscopic procedures is recommended for diagnosis of small bowel tumours, depending on clinical features and proliferation of the tumour [3].

5. Complications and risk factors

- Small bowel tumours are a risk factor for capsule retention [17]. However, capsule retention due to small bowel tumour is still rare and rather considered a diagnostic aid than a complication.
- Suspected small bowel tumour as the indication for VCE has a significantly higher risk of retention, regardless of the endoscopic finding [18].
- Endoscopic polypectomy increases the risk of bleeding and perforation during device-assisted enteroscopy. Diagnosis of a tumour by tissue sampling and endoscopic resection of tumours or polyps may cause bleeding or perforation, DBE and SBE show overall complication rates from app. 1% for diagnostic to 4% for therapeutic procedures [19-21].
- Diagnostic double balloon enteroscopy in patients with diffuse small bowel lymphoma seams to bear a special risk of perforation, especially if carried out after chemotherapy [22].
- Cases of small bowel tumours detected by double balloon enteroscopy but missed by capsule endoscopy have been reported [3,23].

6. Scripted voiceover

Voiceover Text

Small bowel tumours are rare, but are represented by a wide range of different benign and malignant entities.

Adenocarcinomas are the most frequent primary malignant small bowel tumours. Partial obstruction of the lumen and infiltration of the mucosa are typical findings.

Small bowel tumours often present as submucosal tumours, as this gastrointestinal stromal tumour with ulceration of the overlying mucosa

This submucosal GIST shows a constant form and a clear boarder, differentiating it from a harmless bulge

The next capsule endoscopy shows a pedunculated and bleeding fibrolipoma of the jejunum. Histology cannot be predicted by capsule endoscopy in these submucosal tumours as ulceration of the overlying mucosa is also observed in benign mesenchymal tumours

Push enteroscopy in this patient additionally demonstrates the large ulceration of the mucosa.

Neuroendocrine tumors also typically present as submucosal mass. This lesion shows a central umbilication. Neuroendocrine tumors may appear multiple

In another case, infiltration of the mesentery by a neuroendocrine tumor causes red spots of the small bowel mucosa. Distribution to only one segment helps differentiating these spots from multiple usual angiectasias.

However, a diffusely infiltrating neuroendocrine carcinoma may resemble any other malignant tumour. Here a large ulcerated mass in the jejunum is seen during push enteroscopy

Malignant melanoma is the most frequent origin of metastases to the small bowel. Like in this capsule endoscopy, the dark pigmentation is a key diagnostic feature. However, it is not visible constantly. Other parts of the tumour show unspecific ulceration or reddish, swollen mucosa.

Voiceover Text

Amelanotic melanoma, however cannot be distinguished endoscopically from other tumours like in this case, where an ulcerated, infiltrating, and protruding mass of the small bowel is seen

Non-Hodgkin's lymphoma may involve the small bowel. The endoscopic appearance of small bowel lymphomas has a great variation. The next case of mantle cell lymphoma causes diffuse infiltration of the proximal small bowel with lymphangiectasia and erythema

In this patient with abdominal pain and mid GI bleeding capsule endoscopy diagnosed a solid ulcerated tumour. Stenosis lead to capsule retention as demonstrated by double balloon enteroscopy. Surgical resection proofed an isolated large B-cell lymphoma of the mid small bowel.

Sarcomas of the small bowel are rare tumours. Here a large protruding and ulcerated mass is found in the proximal small bowel of an elderly patient with overt bleeding. Endoscopic appearance does not allow differention from other tumours

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