Small Bowel Tumors: The big five. Key imaging features using CT and MR.

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Learning objectives

To learn to differentiate the five most common small bowel tumors and its mimickers using CT and MR imaging features.

To get acquainted with risk factors and clinical syndromes associated with small bowel lesions.
Background

Small bowel tumors are rare, accounting for 3-6% of all gastrointestinal neoplasms. In most cases clinical presentation is not specific; major symptoms can include gastrointestinal bleeding, anaemia, abdominal pain and small bowel obstruction. The radiologist has an important role in identifying the disease at an early stage.

To evaluate small bowel abnormalities enteric contrast is needed to distend the lumen. Luminal distension should be at least 2 cm, as collapsed small bowel loops can be easily misinterpreted as wall thickening or abnormal enhancement. Bowel wall thickness more than 3 mm is considered abnormal.

For MR enterography and enteroclysis enteric contrast media with low signal on T1 weighted images and high signal on T2 weighted images are chosen.

In general, a circumferential mass with prestenotic dilatation, mesenteric fat infiltration and lymphadenopathy are features of small bowel neoplasms. More specific findings are mentioned below per entity.
Findings and procedure details

**Adenocarcinoma**

Adenocarcinoma is the second most common primary malignant tumor of the small bowel, representing 25-40% of all small bowel neoplasms (colon carcinoma is 50 times more common). Adenocarcinoma occurs especially in the duodenum (50% of cases), while the jejunum is the second most prevalent site. Occurrence in the ileum is related to Crohn's disease.

The typical imaging representation of small bowel adenocarcinomas is a focal unilocular, circumferential mass with shouldering of the margins (Fig 1). However, early adenocarcinomas can present as an intraluminal polypoid mass, which may lead to intussusception. Adenocarcinomas usually show moderate enhancement and may present with partial or complete obstruction. Ulceration is a quite common feature, mimicking a small bowel lymphoma in case of a large aggressive ulcerating adenocarcinoma (Fig 2). Features that favour adenocarcinoma are mesenteric fat infiltration and less bulky lymphoid metastases than can be seen in small bowel lymphoma. Metastases to lymph nodes, liver or peritoneum occur quite frequently (Fig 3).

Risk factors for adenocarcinoma of the small bowel are: HNPCC (hereditary nonpolyposis coli colon cancer), familial adenomatous polyposis, Peutz-Jeghers and celiac disease.

**Lymphoma**

Lymphomas make up about 20% of all small bowel tumors. The (distal) ileum is the most common site, owing to the largest amount of lymphoid tissue that is normally present. Many different histologic types of lymphomas exist, which cannot be differentiated based on imaging features. Most prevalent small bowel lymphomas are non Hodgkin lymphomas.

Fig 4 shows imaging features of an enteropathy associated T-cell lymphoma (EATL) that can be suggested based on patient's history of celiac disease and the specific imaging characteristics as shown in this case (Fig 4).

The most common presentation is a thick walled infiltrating mass with aneurysmal (nonobstructive) dilatation (Fig 5). Aneurysmal dilatation is based upon destruction of the bowel wall and myenteric plexus. It can also appear as an intraluminal polypoid mass or a large exocentric mass with extension into the surrounding soft tissues with possible
ulceration and formation of fistulas (Fig 6). A large adenocarcinoma and lymphoma can have a similar imaging appearance in some cases (Fig 2 & Fig 5). Bulky mesenteric or retroperitoneal lymphadenopathy and splenomegaly are findings that can support the diagnosis of lymphoma.

Risk factors include celiac disease, Crohn's disease, SLE, immunocompromised individuals and a history of chemotherapy or extra-intestinal lymphoma.

Carcinoid tumor

Gastrointestinal carcinoids are well-differentiated endocrine tumors accounting for 44% of all small bowel tumors. They are more common than carcinoids in the tracheo-bronchial system. Most common presenting gastrointestinal site is the distal ileum, followed by the appendix, rectum and stomach. In about one third of the cases patients present with multiple tumors. There is an association with multiple endocrine neoplasia type I (MEN I).

A minority of patients (< 10 %), especially those with hepatic metastases will develop a carcinoid syndrome, due to intratumoral serotonin production. Symptoms can include flushing, sweating, bronchospasm, right-sided cardiac valvular fibrosis, abdominal pain and diarrhea.

Imaging features range from small submucosal lesions with arterial enhancement to lesions that infiltrate the bowel wall and mesentery (Fig 7 & Fig 8). Small polypoid lesions may lead to an intussusception. In case of transmural tumor extension the typical picture is a calcified, spiculated mesenteric mass with a desmoplastic reaction and adjacent thickened small bowel loops (Fig 9).

Small bowel loops can retract towards the root of the mesentery, leading to kinking of the bowel wall and narrowing of the lumen. In Some cases segmental small bowel ischemia may occur due to occluded mesenteric vessels as a result of fibrosis. Liver metastasis are usually hypervascular with or without central necrosis (Fig 10). Most of the lymph node metastases show calcifications, similar to the primary tumor.

GIST

Gastrointestinal stromal tumors (GISTs) represent 9% of all small bowel tumors. These tumors most frequently occur in the stomach, followed by jejunum and ileum. Occurrence in colon, rectum, esophagus and appendix is rare.
GISTs are mesenchymal tumors and histologically they can be classified as benign, borderline or malignant. About 20-30% of GISTs are malignant at presentation. In the small bowel they are more often malignant than in the stomach. At imaging tumor size is the most predictive factor of metastatic potential. Tumors smaller than 2 cm are usually benign, whereas masses larger than 5 cm are often malignant. Malignant GISTs predominantly grow extraluminally and can show necrosis, haemorrhage, calcification (post therapy) or fistula formation.

Typically a GIST is a well defined, exophytic mass with heterogeneous enhancement and a clear delineation from the mesenterium (Fig 11). An intraluminal mass is less common.

Obstruction is rare because GISTs do not involve the circumferential bowel wall, in contrast to adenocarcinoma. Unlike carcinoid tumors, the primary lesion in GIST is large and represents the predominant finding.

Liver metastases are usually hypervascular. In general, lymph node metastases are not seen. So, if lymphadenopathy is seen another diagnosis should be considered. Mesenteric or (less common) omental metastases are more common in recurrent disease than at presentation. This is thought to be due to spill during surgery; despite radical surgical resection 40-90% of patients have recurrence of disease in the liver or mesentery. These metastases can be easily missed, as they often have a low-density center. Gleevec can be given in case of metastatic disease. After chemotherapy liver and mesenteric metastases become hypovascular or even cystic.

**Metastases**

Metastases to the small bowel are more common than primary small bowel tumors. The spread can be intraperitoneal, hematogenous, lymphatic or by direct extension.

Intraperitoneal seeding is the most common pathway (50%), especially in ovarian, appendix and colon carcinoma. Metastatic cells implant on the mesenteric border of the bowel. Hematogenous metastases usually occur in breast carcinoma, melanoma and renal cell carcinoma (Fig 12). They can be polypoid and can cause intussusceptions (Fig 13).

**Differential diagnosis**

**Sclerosing or fibrosoing mesenteritis**
Mass-like mesenteric process that can mimic a malignant tumor like a carcinoid. They can be differentiated by the 'fat ring sign', which is a halo of fat surrounding the mesenteric mass.

**Peutz Jeghers** (Fig 14)

Large polyps in patients with Peutz Jeghers syndrome can mimic primary small bowel neoplasms and can also be malignant. The majority of the intestinal polyps in Peutz Jeghers syndrome are located in the jejunum and ileum.

**Inflammatory Bowel Disease (IBD)**

Wall thickening in inflammatory or infectious small bowel disease should be differentiated from malignant wall thickening. Distinguishing features of inflammation (Crohn's disease) are ulcerations, increased mesenteric vessels (comb sign) and increased surrounding fat (creeping fat) (Fig 15). An association between small bowel carcinoma and Crohn's disease is well-established.

This diagnosis is often hard to make pre-operatively, due to lack of typical imaging features. Although a target appearance of the bowel wall, being result of submucosal edema surrounded by enhancing mucosa and serosa, is a sign of a benign disease, including inflammation, ischemia and radiation enteritis. An indicator of malignancy is a small bowel obstruction that is refractory to medical therapy.

**Hemangioma** (Fig 16)

Most intestinal hemangiomas are located in the jejunum. Imaging features are the same as for hemangiomas in the liver: high signal on T2, nodular arterial enhancement and homogeneous enhancement in the delayed phase.

**Leiomyoma**

Leiomyomas are rare mesenchymal benign tumors that can mimic a GIST. The origin may be intraluminal, submucosal or extraluminal. Benign imaging features include sharp margins, homogeneous aspect and homogeneous enhancement.

**Lipomas** (Fig 17)

These are well-circumscribed intraluminal masses with fat attenuation. Liposarcoma of the small bowel is extremely rare.

**Desmoid** (Fig 18)
Rare, benign, locally aggressive mass composed of fibrous tissue. It can be a mimicker of a malignant bowel or mesenteric neoplasm. Often a history of previous abdominal surgery is present. Desmoid tumors can grow rapidly (mainly in Gardner syndrome) and tend to recur. Because these tumors can be very hard, percutaneous biopsy can be challenging.

**Adenomas**

Adenomas are pre-cancerous lesions that can present as polypoid pedunculated masses, a sessile mass or a mural based nodule within the mucosa. Lesions show homogeneous enhancement and are usually nonobstructive. Extrarerosal extension is suggestive of malignant degeneration.
Fig. 1: Axial T2 image. Unilocular mass with shouldering margins in the duodenum.

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**Fig. 2:** Axial CT. Thick walled jejunal mass with dilated lumen, PA: adenocarcinoma.

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**Fig. 3:** Perifocal mesenterial lymphadenopathy in small bowel adenocarcinoma

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**Fig. 4:** Coronal T2 images. Note extensive mesenterial lymphadenopathy on left image. Middle image shows ileal-ileal intussusception in the right lower quadrant. Right image shows multiple sites of wall thickening in the ileum in lower abdomen. All imaging characteristics consistent with multifocal EATL.

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**Fig. 5:** Axial CT. Typical image of thick walled mass with aneurysmal dilatation in small bowel lymphoma.

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Fig. 6: Coronal CT. Thick walled mass in terminal ileum, representing small bowel lymphoma, with fistula to bladder.

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Fig. 7: Post contrast coronal CT in arterial scan phase shows small enhancing luminal mass marked by red arrow.

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Fig. 8: Coronal CT shows large mesenterial mass in patient with carcinoid. Note also retraction of adjacent small bowel loops.

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**Fig. 9:** Axial CT. Spiculated calcified mesenterial mass and dilated small bowel loops, consistent with small bowel carcinoid.

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**Fig. 10:** Axial MR, T1 fatsat post contrast; arterial scan phase. Enhancing metastasis in right liver lobe, marked by red arrow.

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Fig. 11: Axial CT. Heterogeneously enhancing mass arising from ileum. Note clear delineation from mesenterium and absence of signs of obstruction.

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Fig. 12: Axial nonenhanced CT images and fused FDG-PET images. Two sites of wall thickening in the small bowel with marked FDG uptake, in a patient with metastasized melanoma

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Fig. 13: Axial and coronal CT of intussuscepting luminal mass of small bowel on right side in patient with melanoma. Also note extensive liver metastasis.

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Fig. 14: Axial T2 images. Multifocal small bowel intussusceptions in patient with Peutz Jeghers.
**Fig. 15:** Axial and coronal T2 images. Marked wall thickening of the terminal ileum with surrounding "creeping fat" and subtle comb sign of mesenterial vessels, in patient with Crohn's disease.
**Fig. 16:** Coronal T1 fatsat and coronal T2 shows enhancing clearly delineated luminal mass in the right upper quadrant, PA proven small bowel hemangioma.

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**Fig. 17:** Coronal and axial CT showing low density clearly delineated mass in duodenum/jejunum. Low intensity of the mass on MR T1 fatsat. Macroscopic image of lipoma.

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**Fig. 18:** Axial CT shows large clearly delineated mesenterial mass compressing the IVC, PA proven desmoid tumor.

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Conclusion

Small bowel neoplasms are rare tumors.

Clinical symptoms are often non-specific.

General features of the five most common small bowel tumors are summarized in Fig 19.
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<th>Location</th>
<th>Aspect</th>
<th>General</th>
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<td>hypervascular nodule with mesenterial</td>
<td>10% carcinoid syndrome</td>
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<tr>
<td></td>
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<tr>
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<td>Demarcated mass with exofytic extension</td>
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<td>Metastasis</td>
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**Fig. 19**

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