Imaging of Small Bowel Tumors and Mimics

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Abstract

Small bowel tumors are rare with nonspecific and protean clinical presentation. Early diagnosis of small bowel tumors is desirable as they can be associated with significant morbidity. In malignant small bowel tumors, delayed diagnosis may result in dissemination and metastasis leading to poor clinical outcomes. Imaging evaluation of small bowel can be challenging due to unpredictable luminal distension, peristalsis, and motion. In addition, the lack of distinction between the intraluminal lesions and intraluminal contents can be difficult at times. Computed tomography (CT) and magnetic resonance (MR) enterography are the most common imaging techniques for the evaluation of small bowel tumors. While these techniques may not be able to detect small tumors, they provide comprehensive evaluation of lumen, wall, and extramural structures in tumors more than 2 cm. Acquaintance of imaging appearance of common benign and malignant small bowel tumors may allow improved detection during evaluation of CT and MR enterography studies. In this review, we discuss the imaging appearances, approach, and differential diagnosis of small bowel tumors on cross-sectional imaging studies.

Keywords
- small bowel
- tumors
- CT
- MRI

Introduction

Small bowel tumors are rare. They comprise 3 to 6% of gastrointestinal tract neoplasms.[1] The diagnosis of small bowel tumors is delayed by the nonspecific nature of the symptoms (abdominal pain, weight loss, gastrointestinal bleeding) and low clinical suspicion.[2] The small bowel neoplasms can present with complications like intussusception, obstruction, and perforation.[3] Thus, early diagnosis is desirable by accurate interpretation of radiologic findings.

The occurrence of small bowel tumors is more in the proximal small bowel in comparison to the distal small bowel.[4] The different segments of the small bowel have predilection for specific histologic subtypes tumors, for example, adenocarcinoma is more common in the duodenum and jejunum, and carcinoid tumor is more common in the ileum.[1] The risk factors for malignant small bowel tumors are alcohol, tobacco, chronic inflammatory diseases including celiac disease and Crohn's disease, human immunodeficiency virus infection, certain foods (e.g., red meat, smoked, salty, and fatty food), and inherited syndromes including Peutz-Jeghers syndrome (PJS), hereditary nonpolyposis colorectal cancer, and familial adenomatous polyposis (FAP).[5–9]

Benign small bowel tumors comprise 0.5 to 2% of all gastrointestinal neoplasms.[10] Benign small bowel tumors

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are generally solitary. Multiple tumors are seen in polyposis syndromes.[11] Primary malignant small bowel tumors are less common than benign neoplasms.[4] The malignant small bowel tumors generally have poor prognosis due to delayed presentation. As small bowel can tolerate mild obstruction, patients usually present late when at least two-thirds of the lumen is obstructed.[12]

At imaging, benign small bowel tumors usually appear round and well circumscribed with smooth margins. The malignant tumors have irregular margins with heterogeneous enhancement and may be associated with invasion of adjacent structures.

In general, the imaging detection of small bowel is challenging due to bowel peristalsis, mobility of abdominal structures, respiratory motion, nonuniform distension with luminal agents, and lack of contrast between normal bowel tissue and mass.[13] Plain radiograph has no role in patients with small bowel tumors except when they present in emergency with features of small bowel obstruction and perforation. Ultrasound also has limited role in detection of small bowel tumors. However, in patients at high risk of small bowel tumors like polyposis syndrome, celiac disease, or Crohn's disease, ultrasound evaluation of small bowel may be facilitated by luminal distension using protocols similar to computed tomography enterography (CTE). Barium meal follow through is not considered reliable for the detection of small bower tumors. CT is the widely used modality for imaging of small bowel tumors due to its availability and its speed of acquisition. Furthermore, CT is less susceptible to motion artifacts and provides excellent spatial resolution.[13] However, the ionizing radiation exposure and the need for intravenous iodinated contrast agent are the major limitations of CT. CTE protocol is the preferred technique for suspected small bowel tumors.[14,15] Patients should have at least 4 to 6 hour fasting status before the study.[16] Oral contrast is administered for adequate distention of the lumen. Neutral oral contrasts are preferred to positive oral contrast.[17] These provide better delineation of mucosal enhancement, mural thickness, and mesenteric vasculature.[18] A few studies have evaluated a novel CT technique employing carbon dioxide instillation (virtual CT endoscopy) for the evaluation of small bowel lesions including tumors.[19,20] MR enterography is an alternative to CTE. The key benefit of MR enterography is the absence of ionizing radiation. This makes MRI particularly attractive for imaging in children and for repeat examination. However, the limitations include long acquisition time, limited availability, and a relative higher cost.[21]

In comparison to cross-sectional imaging, endoscopy (or enteroscopy) and capsule endoscopy are better for the detection of small intraluminal tumors.[15] However, a study reported better sensitivity of CTE over capsule endoscopy for the detection of submucosal lesions.[22] Though with the current state of the art endoscopy techniques, the entire small bowel can be evaluated, endoscopic techniques do not provide information regarding the extramural extent of the diseases.[3,13] The various imaging techniques utilized in evaluation of small bowel tumors are listed in ►**Table 1**. The various benign and malignant small bowel tumors are mentioned in ►**Table 2**.

Table 1 Various diagnostic modalities for small bowel tumors

Endoluminal (invasive/ semi-invasive)	Radiological (noninvasive)	
	Fluoroscopic	Cross-sectional imaging
Upper GI endoscopy	SBFT	CT enterography
Colonoscopy	SBE	MR enterography
Push enteroscopy		PET-CT
Balloon enteroscopy		PET-CT
Balloon enteroscopy		Enterography
Endoscopic ultrasound		Virtual CT
Capsule endoscopy		Endoscopy

Abbreviations: CT, computed tomography; MR, magnetic resonance; GI, gastrointestinal; PET-CT, positron emission tomography-computed tomography; SBE, small bowel enteroclysis; SBFT, small bowel follow through.

Table 2 Various benign and malignant small bowel tumors

Benign	Malignant
Lipoma	Adenocarcinoma
Polyp	Lymphoma
Leiomyoma	Neuroendocrine tumor
Gastrointestinal stromal tumor (GIST)	Malignant GIST
	Metastasis
Inflammatory fibroid polyp	Gastrointestinal Neuroectodermal tumors
Hemangioma	

Benign Small Bowel Tumors

Lipoma

Despite being a rare tumor, small bowel lipoma is the most common benign lesion of the bowel causing intussusception in adults. Lipoma arises from submucosa and consists of mature adipose tissue surrounded by a thin capsule. Lipoma is usually sessile; however, it can be pedunculated. Almost 50% of lipomas are found in the ileum, and less than 50% of patients with small bowel lipoma are symptomatic.[23] Lesions more than 2 cm in diameter can be symptomatic and may cause bowel obstruction or gastrointestinal bleeding.[24] Small bowel lipoma has no malignant potential.

On gastrointestinal contrast studies, lipomas have characteristic appearance and demonstrate mobility. They produce a solitary smooth intraluminal filling defect. They demonstrate a pseudopedicle at their tip.[25] CT and MRI features are diagnostic. A well-defined homogeneous mass with fat attenuation (−40 to −120 HU) on CT is seen (►**Fig. 1**).[12] The MRI findings of small bowel lipoma include homogeneous signal intensity corresponding to macroscopic fat without contrast enhancement.[13]

Polyp

Polyps comprise one-fifth of the benign small bowel neoplasms.[26] Polyps generally are asymptomatic. However, polyps can lead to obstruction or intussusception when they grow large enough. Pathologic subtypes are hamartomatous, hyperplastic, adenomatous, and inflammatory.[13] Polyps can be solitary or multiple. Numerous polyps are seen in inherited syndromes (►**Fig. 2**). Polyps appear as small (<2 cm), homogenously enhancing masses which protrude into the bowel

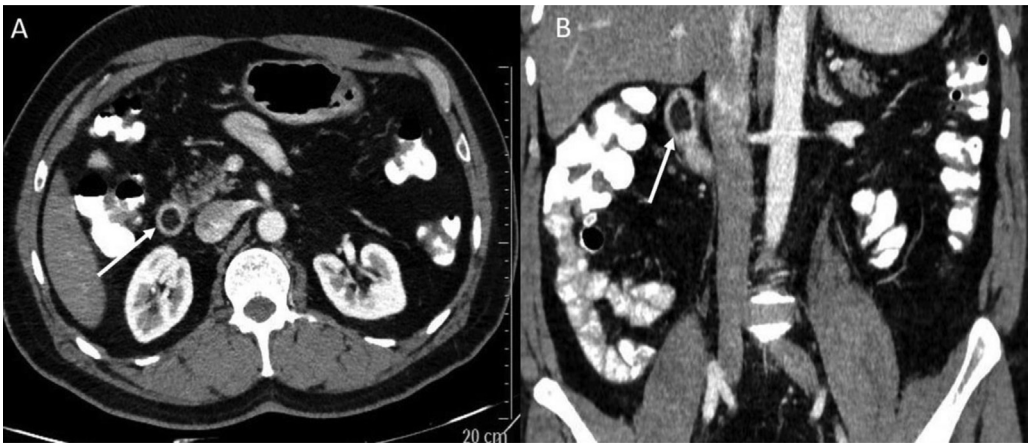

Fig. 1 Lipoma: Axial (**A**) and coronal sections (**B**) of contrast-enhanced computed tomography abdomen showing a well-defined fat attenuation lesion in second part of duodenum (white arrows).

lumen.[11] The adenomatous subtype has increased risk of malignant transformation and is associated with polyposis syndromes. Adenomatous polyps are seen in patients with FAP. Hamartomatous polyps are associated with juvenile polyposis syndrome, PJS, and Cowden syndrome.[27] Cronkhite–Canada syndrome is an acquired nonfamilial polyposis syndrome that is characterized by gastrointestinal polyposis, onycholysis, cutaneous pigmentation, and alopecia.[28] The risk of malignant transformation is higher for tumors more than 1 cm.[23] Size greater than 2 cm and extraserosal extension are highly suggestive of malignant degeneration within a polyp.[29]

Leiomyoma

Leiomyoma is a rare tumor. It is more common in the jejunum than the ileum.[30] Clinical presentation is due to tumor ulceration and bleeding, causing abdominal pain, gastrointestinal bleeding, and chronic anemia. At CT and MRI, it appears as a well-defined homogeneously enhancing mass. Calcification and ulceration may occur in larger tumors.[31] Imaging features may be indistinguishable from gastrointestinal stromal tumor (GIST). Larger tumors (>6 cm) with irregular margins and lymphadenopathy should cause suspicion for malignancy.[30]

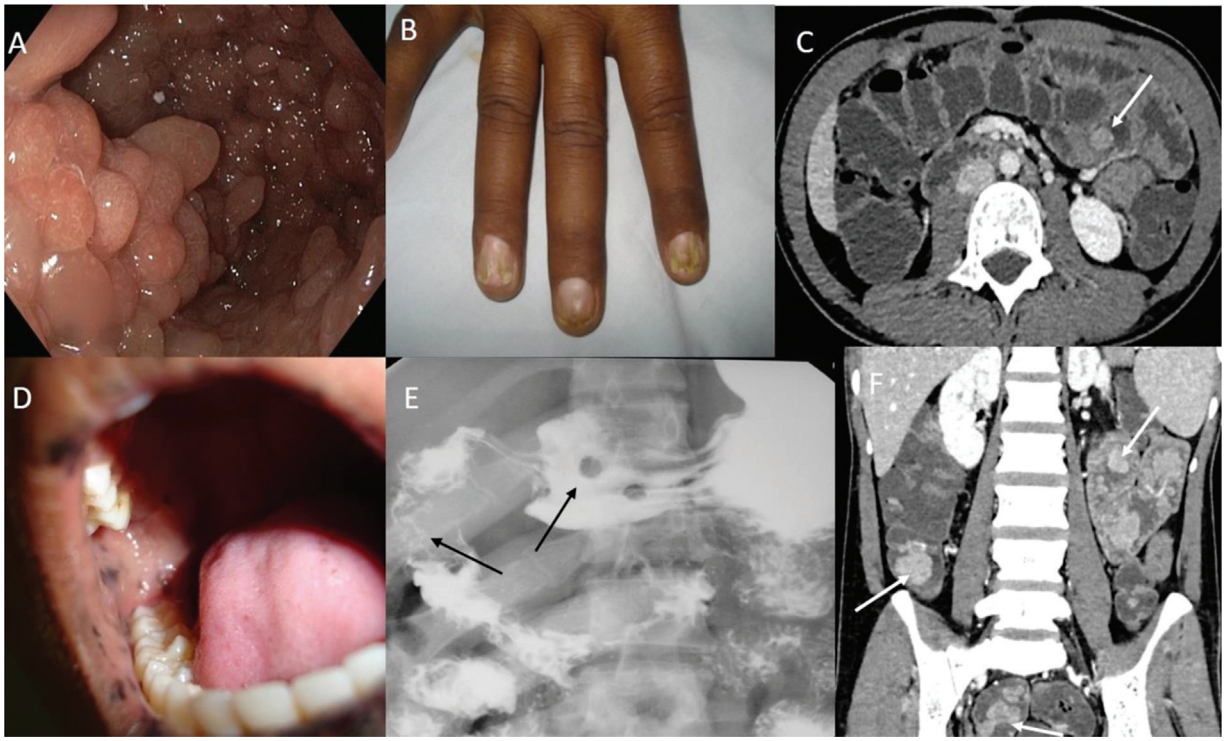

Fig. 2 Polyps: (**A**) Endoscopic image showing multiple polyps and (**B**) involvement of fingernails in Cronkhite-Canada syndrome; (**C**) Axial computed tomography (CT) enterography showing well-defined round homogenously enhancing polyp in the jejunal lumen (arrow); (**D**) Circumoral muco-cutaneous pigmentation; (**E**) Barium meal follow through showing round filling defects (consistent with polyps) in the antrum of stomach (arrows) and (**F**) Coronal CT enterography section showing multiple enhancing polyps within the small and large bowel (arrows).

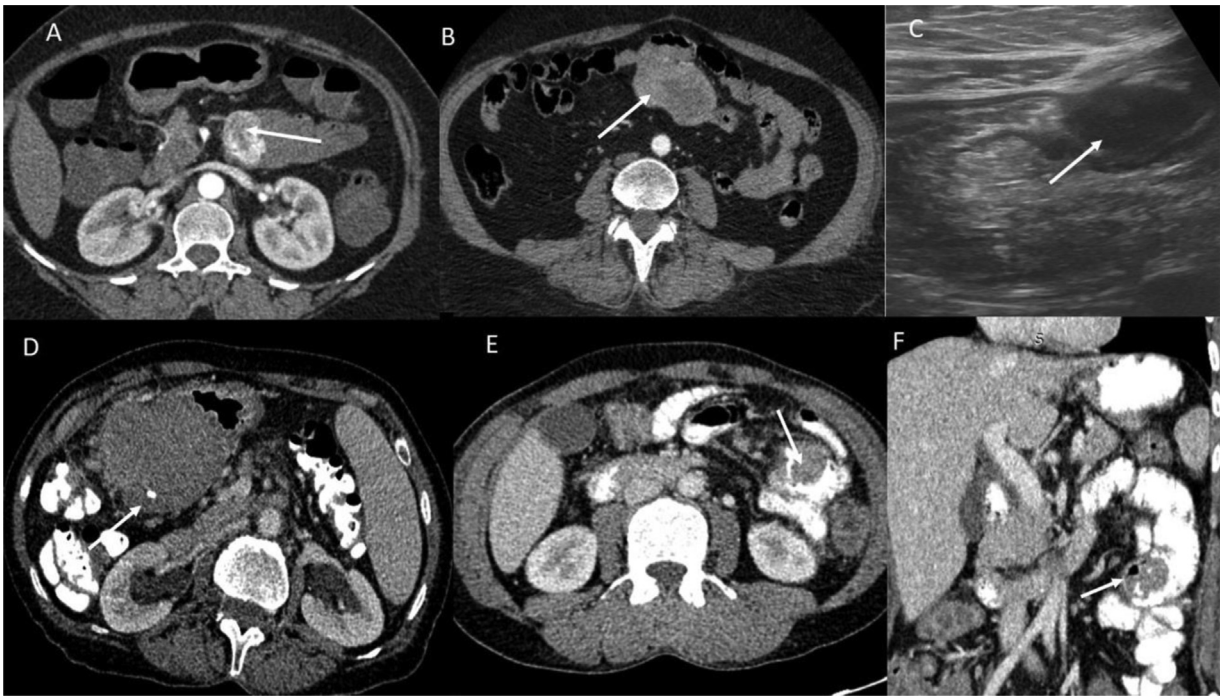

Fig. 3 Gastrointestinal stromal tumors: (**A**) Hypervascular intraluminal mass (arrow) in the D-J flexure/proximal jejunum; (**B**, **C**): necrotic tumor involving distal jejunum on computed tomography (CT; arrow in B) and ultrasound (arrow in C); (**D**) Axial contrast-enhanced CT abdomen showing hypoenhancing exophytic mass lesion arising from stomach with multiple foci of calcification; (**E** and **F**) Axial and coronal images showing endophytic jejunal gastrointestinal stromal tumor.

GIST

GISTs are the most frequent mesenchymal tumor arising from gastrointestinal tract.[32] More than one-third of the GISTs arise in the small bowel. They are more common in the proximal small bowel.[33] GISTs arise from the interstitial cells of Cajal and are characterized by c-*KIT* (CD117) expression.[32] Other mutations include platelet derived growth factor receptor α, succinate dehydrogenase, v-raf murine sarcoma viral oncogene homolog B1, and neurofibromatosis type 1.[33] They tend to have a wide spectrum of clinical behavior, ranging from benign tumors (which are incidentally discovered) to malignant lesions with considerable overlap in the imaging and microscopic features of both entities.[34] All GISTs are potentially malignant.[35,36]

At imaging, benign GISTs are indistinguishable from other mesenchymal tumors. They are well-circumscribed lesions with a variable enhancement pattern. They commonly extend exophytically from the bowel lumen. Calcification is a rare finding (►**Fig. 3**).[37] Smaller lesions (< 2cm) appear as hyper-enhancing lesions. With increasing size, there is development of necrosis in the tumor core. Larger lesions thus appear as heterogeneously enhancing cavitating lesions. Although imaging features may not be entirely reliable for distinguishing benign and malignant GISTs, larger masses with necrosis, local invasion, and hemorrhage suggest malignant behavior.[38] Metastases in such cases commonly occur to the liver, omentum, or peritoneum (►**Fig. 4**). Significant lymphadenopathy is not seen in these cases and favors other malignant neoplasms like lymphoma or metastatic disease. The prognosis of GIST depending upon CT features is mentioned in ►**Table 3**.

Treatment for resectable GISTs is wide local excision. Systemic treatment in the form of chemotherapy is often administered. Imaging features of GISTs change post-imatinib therapy and include intralesional hemorrhage, cystic degeneration of tumor, and development of ascites.[39] The radiologist must thus be aware of these changes and always acquire a multiphasic CT following a noncontrast scan in follow-up patients with GIST on chemotherapy.[39]

Hemangioma

These rare submucosal tumors occur more commonly in the jejunum. They may be sessile or pedunculated.[40] On CT, hemangioma appears as enhancing, intraluminal polypoid mass. On MR, hemangiomas show marked T2-weighted hyperintensity with avid nodular enhancement in the arterial phase.[41] The enhancement is retained in the delayed phase.

Malignant Small Bowel Neoplasms

Adenocarcinoma

Adenocarcinoma accounts for 25 to 40% of primary malignant tumors of the small bowel.[42,43] Proximal jejunum or distal duodenum is the most involved sits.[44] Presentation may be nonspecific or related to malignancy-induced gastrointestinal bleeding or obstruction.

CT appearances include circumferential annular (apple core) mural thickening or eccentric and irregular mass with luminal narrowing (►**Fig. 5**).[45] There may be extension into adjacent fat, vascular invasion, lymphadenopathy, peritoneal and distant metastases (most often to the liver).

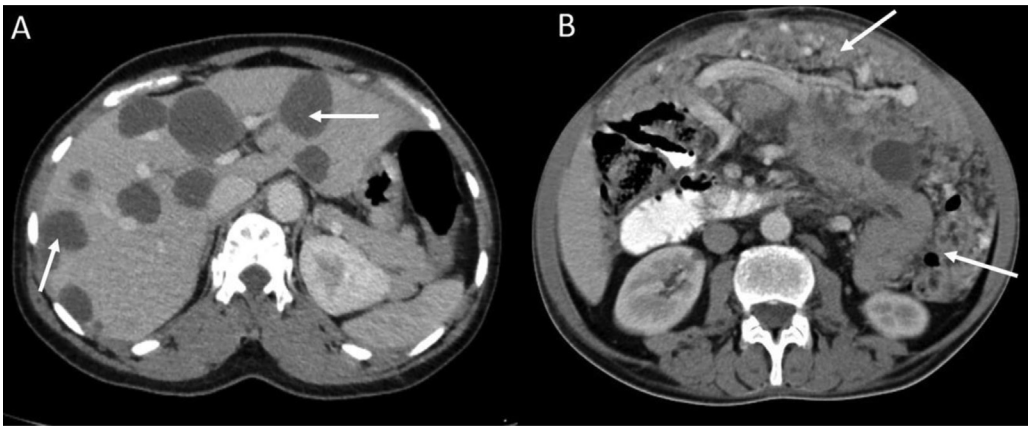

Fig. 4 Malignant gastrointestinal stromal tumor: (**A**) Multiple liver metastases are seen (arrows). The cystic appearance of these lesions is due to imatinib therapy. (**B**) Marked peritoneal thickening and nodularity (arrows) secondary to peritoneal metastases is seen.

Neuroendocrine Tumor

Gastrointestinal neuroendocrine tumors (GNET) originate from the enterochromaffin cells within the gastro-entero-pancreatic system.[34] These are the second most common malignant tumors of the small bowel (20–25% of malignant tumors).[46] One-third of GNETs originate from the small bowel.[47] Ileum is the most common site of involvement. The characteristic appearance of small bowel GNET is a well-defined solitary avidly enhancing mural mass (►**Fig. 6**). Smaller lesion may be missed on CT. MRI may allow detection of some of these lesions due to better soft tissue resolution. Multiple GNETs occur in one-fourth of the patients (►**Fig. 6**).[48] Carcinoid tumors may cause asymmetric or nodular mural thickening. The key to correct diagnosis in these cases is the identification of spiculated (due to desmoplastic reaction caused by secretion of serotonin) calcified mesenteric lesion with or without liver metastases.[49,50] Mesenteric involvement by direct extension or via lymphatics occurs in approximately 40 to 80% of the cases. Mesenteric metastases calcify in 70% of the cases.[49] Differential diagnoses include treated lymphoma or retractile mesenteritis. Somatostatin-analog imaging exams have an important role both in diagnosis and staging (►**Fig. 6**). In-pentetreotide imaging (Octreoscan) has now been replaced by newer analogue agents such as 18F-FDOPA and 68Ga-DOTATATE.[51]

Table 3 Prognostic features of GIST on imaging

CT features	Prognosis
Site Stomach Duodenum Ileum/jejunum	Favorable Intermediate Unfavorable
Size <5cm 5–10 cm >10 cm	Favorable Intermediate Unfavorable
Margin Regular Irregular	Favorable Unfavorable
Homogeneous enhancement	Favorable
Hemorrhage	Unfavorable
Necrosis/cystic degeneration	Unfavorable
Air	Unfavorable

Abbreviations: CT, computed tomography; GIST, gastrointestinal stromal tumor.

Lymphoma

Primary gastrointestinal lymphoma is the most common form of extranodal lymphoma.[52] Diagnosis can be ascertained by the lack of peripheral or mediastinal lymphadenopathy, normal white blood cell count, and differential leucocyte count without the involvement of liver or spleen.[53] Ileum is the most common site of involvement due to abundant lymphoid tissue (►**Fig. 7**). Lymphoma of the small bowel is categorized into five forms: pseudoaneurysmal, polypoid, endoexoenteric, stenosing, and mesenteric (►**Fig. 7**).[54] The most common type of small bowel lymphoma is the polypoidal form with single or multiple polypoidal lesions protruding into the lumen.[55] This type of lymphoma may act as a lead point for intussusception. The pseudoaneurysmal form involves the submucosa and muscularis layers causing mass-like mural thickening. The lack of obstruction is due to complete replacement of the muscle layer with lymphoid tissue.[54] The endoexoenteric form or the cavitatory form produces a large soft-tissue mass communicating with the bowel lumen producing a characteristic air-contrast contrast material level. Malignant GIST may produce a similar appearance.[12] The stenosing form of lymphoma is uncommon. It is usually encountered in patients with celiac disease. This form occurs most commonly in the distal duodenum.[12] In the mesenteric form, tumor extends into the mesentery from the bowel wall.

Immunophenotypically, lymphomas are of two types: B-cell and T-cell lymphoma. B-cell lymphoma (diffuse large B-cell lymphoma) is the most common type and is usually present is ileum, whereas T-cell lymphoma is usually associated with celiac disease and is present commonly in jejunum

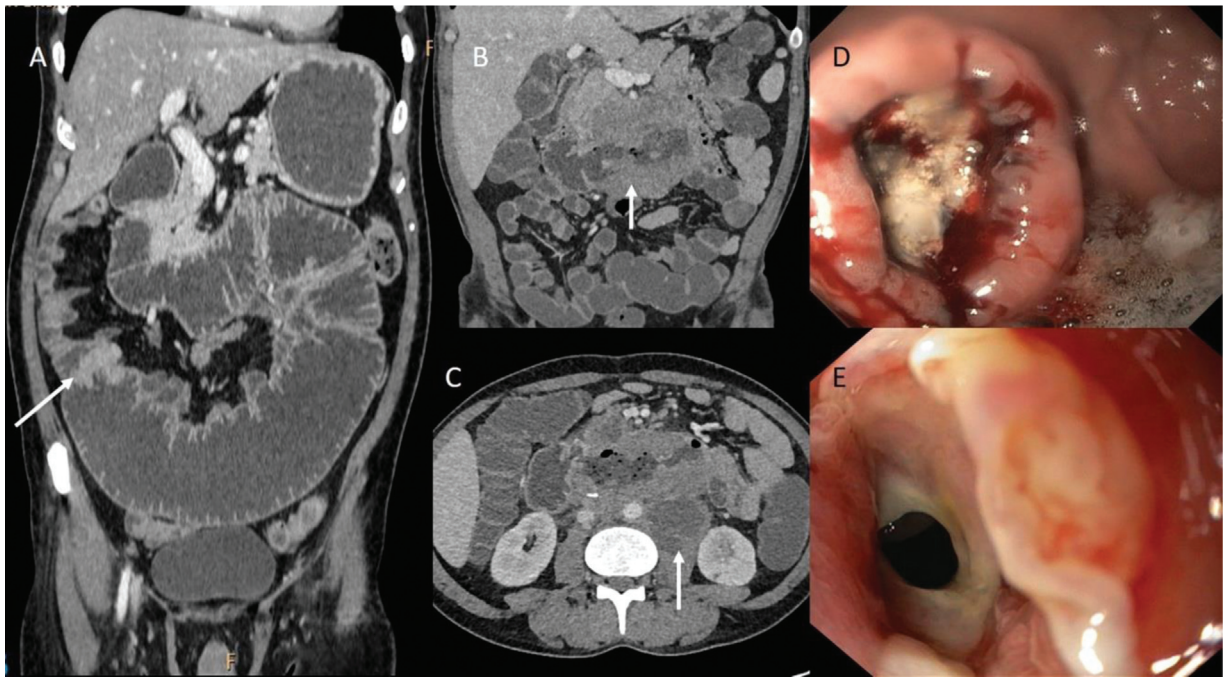

Fig. 5 Adenocarcinoma: (**A**) Coronal computed tomography (CT) enterography image showing asymmetric enhancing mural thickening (arow) involving distal jejunum causing intestinal obstruction; (**B**, **C**): Coronal and axial CT enterography image showing asymmetric mural thickening involving the duodenum with adjacent invasion (arrow in B) and necrotic retroperitoneal lymph nodes (arrow in C); (**D**, **E**): Enteroscopic images showing intraluminal polypoidal (**D**) and annular growth patterns (**E**).

and proximal ileum.[55] T-cell lymphoma is generally multifocal and may be associated with complication like perforation.

On barium meal follow through, there is irregular wall thickening, fold effacement, aneurysmal bowel dilatation, single, or multiple filling defects due to polypoid masses. At CT and MRI, the imaging appearance of small bowel lymphoma parallels the morphological forms described above. Extensive regional and distant adenopathy help in confirming the diagnosis and from other neoplasms.

Differentiating lymphoma from primary adenocarcinoma can be challenging. The features favoring lymphoma are distal site of involvement (ileum), marked homogeneous wall thickening usually greater than 2 cm, multifocal involvement, and extensive lymphadenopathy.[12]

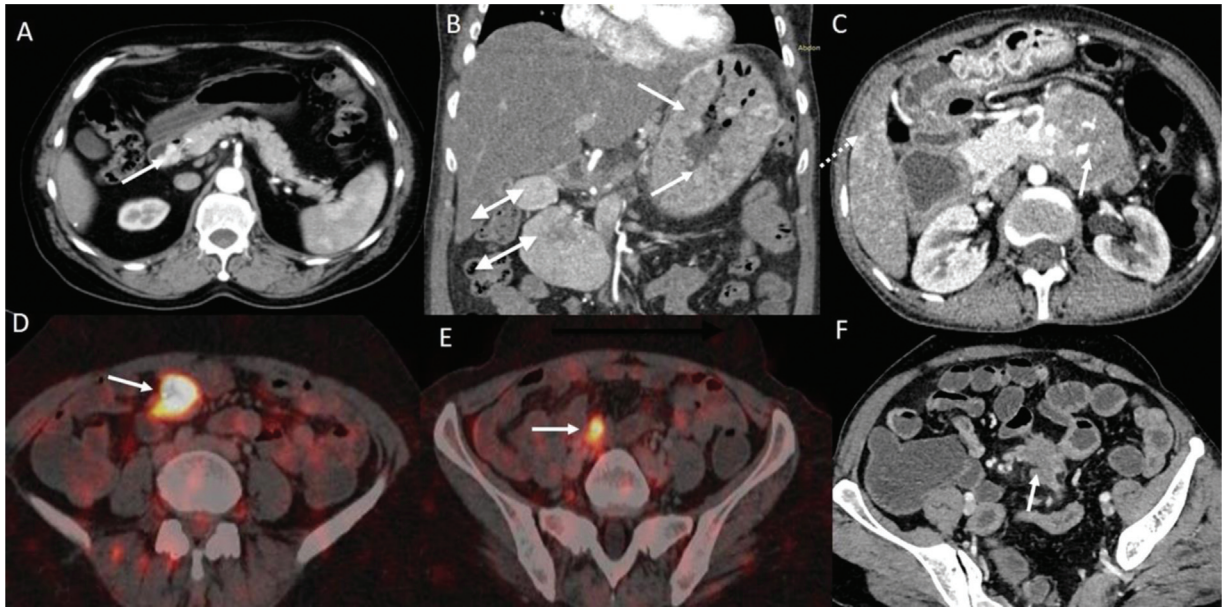

Fig. 6 Neuroendocrine tumor: (**A**) Solitary hypervascular lesion within the duodenal lumen (arrow); (**B**) Coronal image showing multiple neuroendocrine tumors (NETs) in the stomach (white arrows) and hypervascular lymph nodes (double headed white arrows), (**C**) Hypervascular NET in the duodenum with calcifications (arrow) with liver metastasis (dashed arrow); (**D**, **E**): Somatostatin analogue positron emission tomography images showing avid lesions in the mesentery (arrow); (**F**) Computed tomography enterography image showing spiculated hypoenhancing mesenteric mass lesion (arrow).

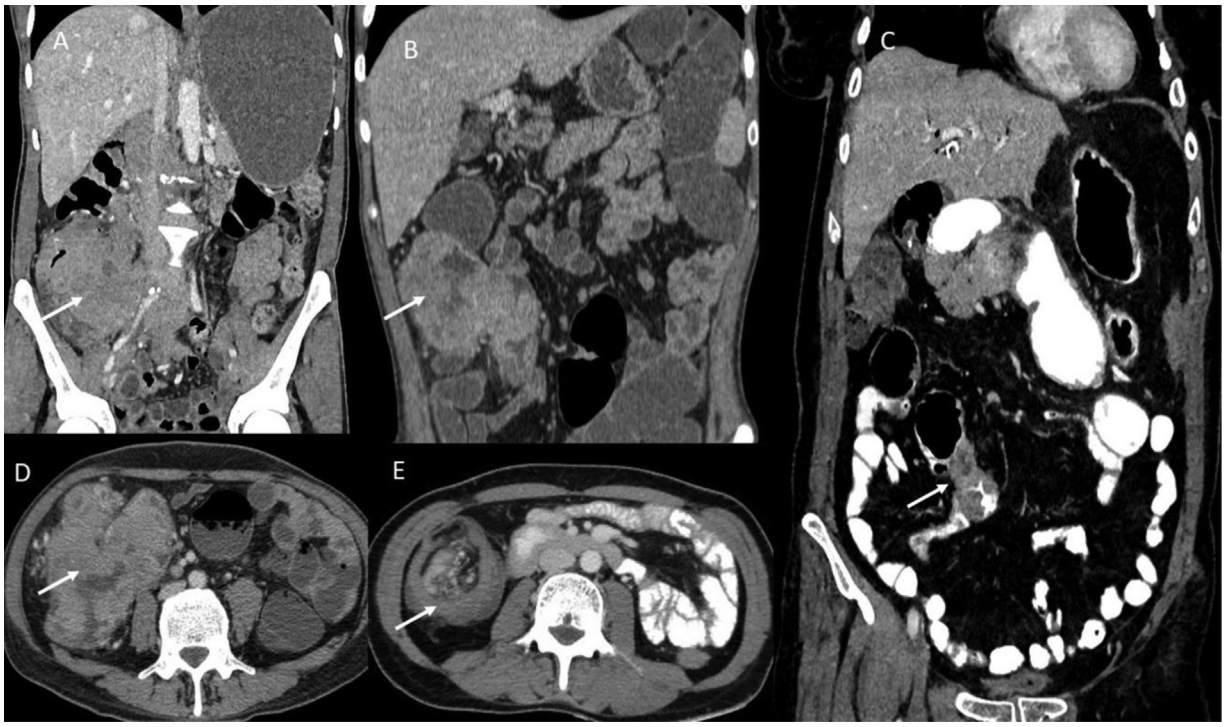

Fig. 7 Lymphoma: (**A**–**C**): Coronal computed tomography enterography images showing heterogeneously enhancing lesion in relation to terminal ileum (arrow). **B**: Asymmetric wall thickening involving terminal ileum (arrow); **C**: Circumferential mural thickening causing luminal stenosis in proximal ileum (arrow); **D**: Asymmetric mural thickening with exophytic component in terminal ileum (arrow); **E**: Ileo-colic intussusception secondary to lymphoma (arrow).

Metastasis

Metastasis to the small bowel is rare. Melanoma, lung cancer, and breast cancer are the tumors that may show small bowel spread.[29] Small bowel metastases may be solitary or multiple with a variety of appearances. At one end of the spectrum, they can mimic benign lesions with discrete, smoothly marginated nodules showing homogenous enhancement. At the other end, there are large mass with cavitation, invasion of adjacent structures, and intraperitoneal spread (►**Fig. 8**).[56,57] Metastases must be suspected when a solid small bowel mass is seen in a patient with malignancy known to metastasize to bowel.

Mass-Like Small Bowel Lesions: Small Bowel Tumor Mimics (►Fig. 9)

Small Bowel Diverticulitis

Other than duodenal or Meckel's diverticulitis, small bowel diverticulitis is rare. Jejunum is more commonly involved.[58] On CT, there is a thick-walled well-defined mass-like

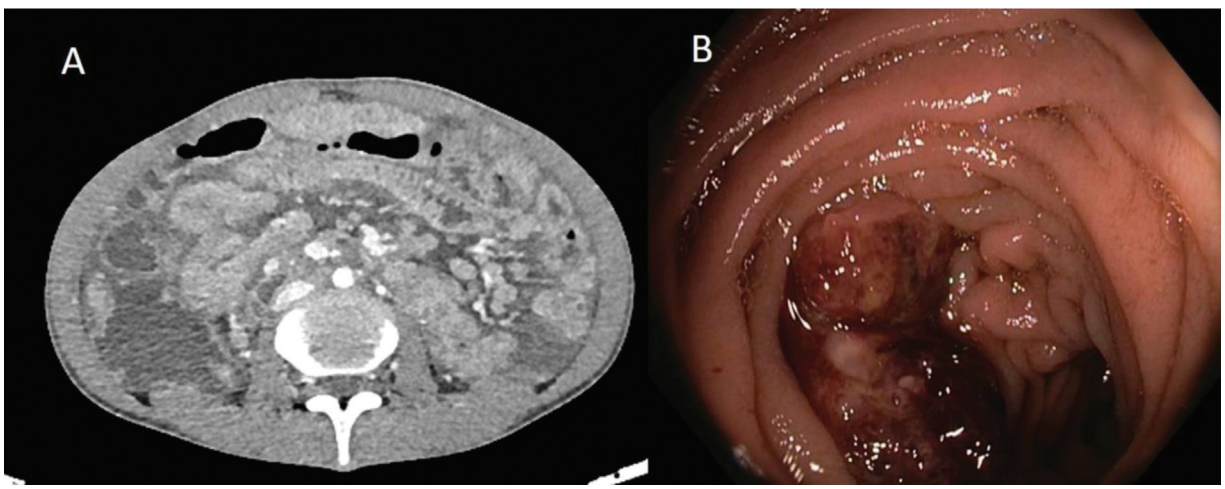

Fig. 8 Small bowel metastases: (**A**) Multiple enhancing serosal deposits in a case of ovarian cancer; (**B**) Enteroscopic image showing intraluminal polypoidal hemorrhagic soft tissue mass in jejunum in a patient with choriocarcinoma.

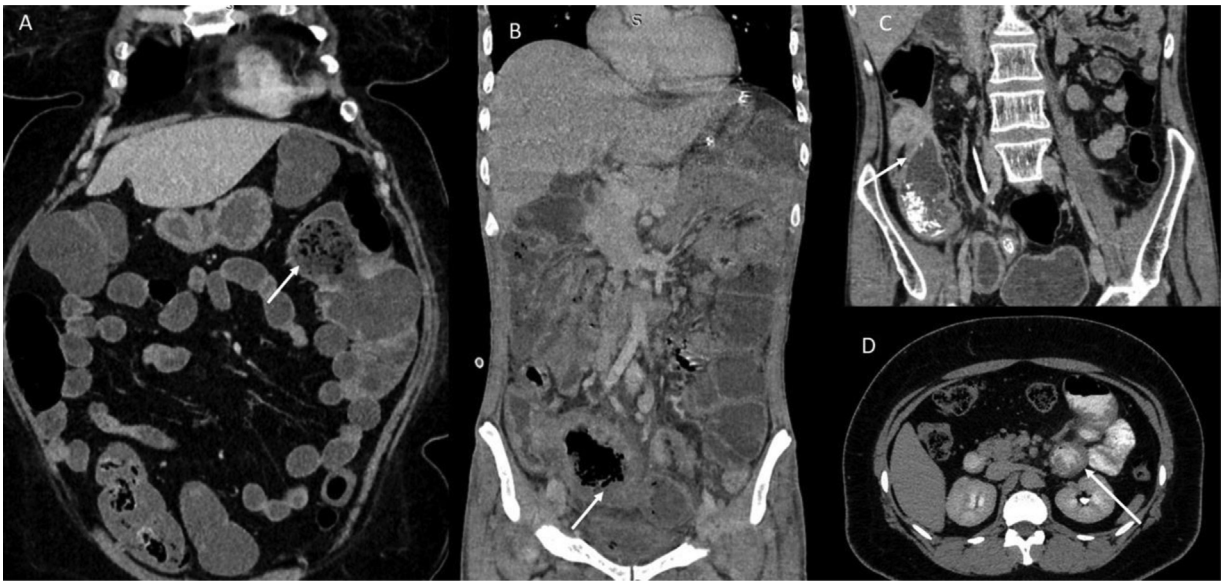

Fig. 9 Neoplasms mimics: (**A**) Jejunal diverticulitis (arrow); (**B**, **C**): Dilated terminal ileum with mural thickening (B, arrow) and mural thickening involving cecum with dilated terminal ileum (C, arrow) in a patient with ileocecal tuberculosis; (**D**) Mural thickening involving jejunal loops in a patient with Henoch–Schonlein purpura.

Table 4 Differential diagnosis for small bowel tumor

Intramural hematoma	Anticoagulation, coagulopathy, trauma, vasculitis, mass-like, mural hyperattenuation with luminal narrowing; spontaneous resolution
Small bowel diverticulitis	Elderly patients, jejunum, ovoid, mass-like structure containing air, fluid, lesion with adjacent fat standing
Meckel's diverticulitis	Mass-like structure in continuity with small bowel
Eosinophilic gastroenteritis	Nodular/Irregular focal, segmental or diffuse thickening vs. lymphoma
Giardiasis/Whipple's disease	Fold thickening in duodenum and proximal jejunum
Localized lymphangiectasia	Low attenuation wall thickening of jejunum
Sclerosing mesenteritis	versus carcinoid

structure containing intestinal contents (debris, fecal material, and gas). There is associated bowel wall thickening and mesenteric fat stranding. In some patients, additional diverticulitis may also be seen at other sites.[59,60]

Meckel's Diverticulum

Meckel's diverticulum is associated with several complications including diverticulitis, perforation, enterolith formation, bowel obstruction, bleeding from ectopic gastric mucosa, and neoplasm.[61] On CT, Meckel's diverticulum may be confused for a bowel origin mass, but evaluation of serial thin sections reveals a visualized as a blind-ending, fluid, or debris-filled, dilated mass-like structure in continuity with the ileum. CT enterography has higher sensitivity in evaluating Meckel's diverticulum.[62]

Small Bowel Hematoma

Small bowel hematoma occurs in the setting of anticoagulation, coagulopathies, vasculitis, trauma, and malignancy.[63,64] On CT, the thickened bowel shows mural hyperattenuation, and luminal narrowing.[12] The mural hyperattenuation in acute phase helps in the differentiation of small bowel hematoma from other causes of bowel wall thickening.[65] With aging of the hematoma, the hyperdensity of the bowel may disappear. Complete resolution of hematoma occurs over a few weeks.

The other differential diagnoses of small bowel tumors are mentioned in ►**Table 4**.

Conclusion

Imaging plays an important role in the detection and characterization of small bowel tumors. It is important to be aware of lesions that can mimic small bowel tumors as they have entirely different management.

Conflict of Interest
None declared.

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