Small bowel findings reveal tumor spectrum

June 03, 2006 | MRI [1]
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The spectrum of usual and unusual primary neoplasms involving the duodenum, jejunum, and ileum is extremely wide. Our own database of digestive pathology contains a range of benign small bowel neoplasms (adenoma, leiomyoma, lipoma, familial polyposis, hemangioma, lymphangioma, and fibroma), as well as examples of malignancy (adenocarcinoma, carcinoid tumor, lymphoma, leiomyosarcoma, direct extension from extraintestinal tumors, and metastasic lesions).

The spectrum of usual and unusual primary neoplasms involving the duodenum, jejunum, and ileum is extremely wide. Our own database of digestive pathology contains a range of benign small bowel neoplasms (adenoma, leiomyoma, lipoma, familial polyposis, hemangioma, lymphangioma, and fibroma), as well as examples of malignancy (adenocarcinoma, carcinoid tumor, lymphoma, leiomyosarcoma, direct extension from extraintestinal tumors, and metastasic lesions).

Many of these tumors have a characteristic appearance on barium studies, ultrasound, CT, and MRI. But diagnostic difficulties and pitfalls cannot be ruled out. This review of key imaging findings, coupled with a comparison of radiological and pathological information, illustrates the strengths of combining diagnostic approaches.

Fewer than 6% of all primary gastrointestinal tumors involve the small intestine. Most occur in the duodenum, where they prompt different symptoms, radiological features, and treatment compared with tumors in the jejunum and ileum. Duodenal neoplasms also require different treatment strategies than jejunal and ileal tumors.

Most small bowel tumors are asymptomatic until the late stages of disease. Symptoms-when they do occur-include nonspecific abdominal pain, weight loss, diarrhea, and constipation. Signs related to a complication (bleeding, obstruction, or perforation) may also be observed. It can take six months from a patient's initial presentation to reach a clinical diagnosis. Earlier diagnoses from radiology are needed to improve the poor prognosis of these tumors.

The ratio of benign to malignant tumors in the small intestine is estimated as ranging from 1:1 to 1:4, depending on the series. Tumors can develop in the epithelial cells, lymphoid tissue, muscle, vessels, and nerves. The most common benign tumors are leiomyomas, adenomas, and lipomas. Fibromas, hamartomas, neurogenic tumors, hemangiomas, and lymphangiomas are less frequent. The most common malignant tumors are adenocarcinomas, lymphomas, and carcinoid tumors. Metastatic tumors and leiomyosarcomas can also be found.

The majority of adenomas and adenocarcinomas occur in the duodenum and jejunum. Leiomyomas and leiomyosarcomas are more common in the jejunum. Lipomas, lymphomas, and carcinoid tumors appear more frequently in the ileum.[1]

Barium examination (preferably enteroclysis) remains the imaging modality of choice for evaluating small bowel tumors. This is especially true for intraluminal and mucosal neoplasms. CT, however, is superior for evaluating the intramural component and performing preoperative staging and therapeutic follow-up. Barium studies and CT should consequently be regarded as complementary techniques. Plain-film radiography may demonstrate air-fluid levels when obstructions and calcified lesions are present.

The great distention of the small bowel provided by enteroclysis is now being combined with cross-sectional imaging and multiplanar reformatting to produce the newer techniques of CT enteroclysis and MR enteroclysis. CT enteroclysis generally involves administration of positive or neutral enteral contrast without fluoroscopic monitoring. Positive contrast can also be used with fluoroscopic infusion, allowing dynamic bowel imaging prior to CT. MR enteroclysis can provide superb soft-tissue contrast and functional information without exposing patients to ionizing radiation. Although further investigations are needed to validate MR enteroclysis, it may become an alternative modality of choice for patients with suspected small intestinal neoplasms or obstructions.[2]

EXAMPLES OF MALIGNANCY

Adenocarcinoma (Figure 1) accounts for 40% of malignant small bowel neoplasms. Approximately 42% of small bowel adenocarcinomas are located in the duodenum, while 35% involve the proximal

jejunum. The most frequent gross specimen findings are ulceration and infiltration. Tumors involving the jejunum and ileum, however, usually appear as annular constriction. Clinical manifestations include early bowel obstruction and bleeding from ulceration.

Plain-film radiography can show a dilated proximal jejunum and air-fluid levels if obstruction is present. A barium examination will reveal annular narrowing or stricture formation, filling defects, polypoid and/or ulcerated masses, or a combination of these signs. The most typical radiological manifestation is a narrowed segment with features of mucosal destruction, also known as the "apple core" sign.

CT may show an eccentric focal mass or a circumferential irregular thickening of the small bowel wall. It can miss lesions smaller than 2 cm in diameter, though, possibly making barium examination a better option. CT is the most effective technique
for studying retroperitoneum tumor extent and assessing the existence of liver metastases, though it may miss mesenteric invasion.[3-5] MRI may help to demonstrate tumor extension and metastatic disease.

Small bowel involvement by primary or secondary lymphoma (Figure 2) occurs in 20% of lymphoma patients. Primary small bowel lymphoma accounts for just 5% of these cases. Most primary small bowel lymphomas are non-Hodgkin's and arise from B cells. Small bowel lymphomas that complicate celiac disease originate in T cells.

Primary and secondary gastrointestinal lymphomas generally involve the stomach. But small bowel involvement may still be a systemic manifestation of lymphoma. The ileocecal region is the most common location for primary small bowel lymphoma (children and young adults), whereas lymphoma is more often found in the proximal jejunum in patients with celiac disease. Multicentric lymphoma involving different segments of the gastrointestinal tract occurs in 10% to 50% of cases. Pathological findings of primary gastrointestinal tract lymphomas usually show lymph node extension limited to the area of lymphatic drainage of the involved segment. There is not usually any parenchymal or extra-abdominal nodal involvement. Small bowel lymphomas that are present in patients with Crohn's disease, celiac disease, or AIDS or those undergoing chemotherapy may be difficult to diagnose given the overlap in signs and symptoms. Early detection in such cases requires a high index of suspicion.

Radiography is unlikely to show any significant findings of small bowel lymphoma.[1] The wide variety of growth patterns will be reflected in a varying spectrum of results from barium studies, but lumen narrowing is the most common radiological finding. Cavitary forms may also be found. CT imaging may show an infiltrating, circumferential, cavitary, or mesenteric pattern, depending on the nature of the lymphoma.[6] MRI can provide complementary information on metastatic involvement, but this is not usually necessary.

The most common small bowel sarcoma arises from the muscularis propia and is known as leiomyosarcoma. It accounts for 10% to 16% of all small bowel tumors. Other sarcomas, such as fibrosarcoma and angiosarcoma, are rare. Small bowel leiomyosarcomas are most often found in the jejunum and ileum. The tumors are pathologically and radiologically indistinguishable from leiomyomas.

Occult bleeding caused by central mass ulceration and abdominal pain are the most common symptoms at clinical onset. Plain-film radiography is again unlikely to produce any relevant findings. Deformation of the bowel segment involved is the most common observation on barium studies. CT shows leiomyosarcoma as a mesenteric necrotic mass with low central attenuation. A fluid-fluid level is occasionally seen inside the mass.[3,4] When cystic liver metastases are present, MRI may be necessary to differentiate them from other benign cystic lesions.

Carcinoid tumors account for 20% to 40% of small bowel tumors. Most of these will be found at the duodenojejunal junction. Small bowel carcinoid tumors originate in the wall's submucosal layer, arising among the neurogenic cells that are spread throughout the bottom of the Lieberkuhn crypts. Typical clinical signs, which include cutaneous flushing, diarrhea, and abdominal pain, are concomitant with the existence of hepatic metastases. Lower grade obstruction can also be seen, produced by local fibrosis following the carcinoid's serotonin secretion.

Obstructions containing calcifications will occasionally be picked up on plain abdominal radiography. The desmoplastic fibrosis reaction may result in barium studies showing fixation of intestinal loops, lumen kinking, and angulation.[7] CT is very useful for determining the precise anatomic area involved. The tumor itself usually appears as a soft-tissue mass that may contain calcifications. Extension examinations can show lymphadenopathies as well as long distance metastases (usually in the liver).[4,8] We have not found any published reference to features from MRI that could help with differential diagnosis.

Gastrointestinal stromal tumors represent 9% of all malignant small bowel tumors. In the small intestine, they most commonly occur in the jejunum or ileum and are seen less frequently in the duodenum. Most arise pathologically within the intestinal wall's muscularis propria. Their defining feature is expression of the tyrosine kinase growth factor receptor KIT (CD117). Immunoreactivity for KIT distinguishes gastrointestinal stromal tumors from true leiomyomas, leiomyosarcomas, schwannomas, and neurofibromas. Involvement of the muscular layer results in a propensity for exophytic growth.

It is difficult to differentiate benign from malignant gastrointestinal stromal tumors with radiography unless obvious metastases are present. The most common clinical manifestation is gastrointestinal bleeding from mucosa ulceration. Other signs and symptoms include nausea, vomiting, abdominal pain, weight loss, abdominal distention, and intestinal obstruction.[9]

Radiography will show evidence of small intestinal dilatation or a soft-tissue mass if obstruction occurs. Barium examination should reveal an intraluminal or submucosal mass with typically sharply defined margins. Intraluminal, mural, and extraserosal components are well depicted on CT scanning. Metastases to the liver, omentum, and peritoneum may be seen if the gastrointestinal stromal tumor is malignant. MRI signal intensity will depend on the degree of hemorrhage and necrosis.

Metastatic tumors (Figure 3) can result from three different mechanisms.[1] Tumor spread may be hematogenous (lung, breast cancer, and malignant melanoma), invasion may be direct from contiguous tumors (renal, adrenal, pancreatic, and colonic), or there may be intraperitoneal seeding (mucinus tumors of the ovary, appendix, or colon). Clinical signs and symptoms include abdominal pain, vomiting, anemia, abdominal mass, weight loss, and motility disorders of the gastrointestinal tract. Other complications, including perforation, obstruction, stenosis, and intussusception, can also occur.

Radiography may find a mass, obstruction, and/or pneumoperitoneum. Barium examination can show evidence of intraperitoneal seeding in pelvic loops and in the ileocecal region. Hematogenous metastases appear as multiple nodules along the antimesenteric border, where the vasa recta branches out into a rich submucosal plexus. CT can be useful to show the extent of metatasic involvement and for primary tumor identification.[10] MRI may be considered as a complementary study to visualize extent.

BENIGN TUMORS

Leiomyoma (Figure 4) is widely considered to be the most common benign small bowel neoplasm, with an average incidence of 22% to 46%. It usually appears as a solitary mass before spreading throughout the jejunum and ileum. Most leiomyomas arise from the circular or longitudinal muscle layer, though a few rare exceptions start in the muscularis mucosae. Leiomyomas have more symptomatic clinical manifestations than any other benign small bowel tumors. These include occult gastrointestinal bleeding, anemia, and abdominal pain.

Plain-film radiography is of limited use for diagnosis. Barium examinations will frequently show the tumor growing intraluminally or submucosally. Extraluminal or subserosal, bidirectional (dumb-bell), and intramural growth patterns will be seen less often. There are no radiological differences between leiomyoma and leiomyosarcoma.[7] Features such as solitary mass, well-defined edges, absence of large ulceration areas, and lack of excavation or fistula are consequently considered benign. CT and MRI are both useful for completing the preoperative study and assessing the absence of metastases or mesenteric changes.

Adenomas are estimated to account for 15% to 20% of benign small bowel tumors. They are seen mainly in the duodenum, especially in the periampullary region, and are classified histologically as tubular, tubulovillous, and villous. Adenomas most often affect older people, though they can be found in younger patients when associated with adenomatous polyposis. Many authors consider small bowel adenoma to be a precursor to malignancy. The tumors are usually asymptomatic or associated with minimal, intermittent, and nonspecific symptoms, such as bleeding, anemia, and abdominal pain or discomfort.

Plain-film radiography is again of limited use for diagnosis, though it can demonstrate air-fluid levels where obstruction is present. Small intraluminal filling defects may be seen on barium studies. CT is useful for depicting larger lesions and will show a well-defined soft-tissue mass surrounded by a thin rim of contrast. Moderate enhancement is seen after intravenous contrast administration.[3,6] We have not found any specific features on MRI related to diagnosis of small bowel adenoma.

Lipoma is considered to be the second most common benign small bowel mass. This neoplasm arises from the submucosa and usually is encapsulated, homogeneously lobulated, and yellowish in color. Lipomas are usually asymptomatic, though obstruction can lead to clinical manifestation, and will on

average measure 3 to 4 cm.
Plain-film radiography may occasionally show calcifications throughout the abdomen. These calcifications can be moved by normal intestinal peristalsis. A lipoma will appear on barium examination as a smooth, sessile, small single filling defect that tries to adjust to the bowel's lumen shape. CT demonstrates a small, well-defined spherical mass with homogeneous density. Attenuation values are consistent to those of fat.[10] We have found MRI to be a helpful diagnostic tool owing to the common signal intensity between lipoma and fat.
Fibromas are a very rare group of tumors, accounting for 5% of all benign small bowel masses. They develop from the fibrous tissue of the subserosa and submucosa and exhibit extraluminal growth. It is not unusual to find a central calcification corresponding to cystic and mucous degeneration. Fibromas have a clinical manifestation similar to all other benign small bowel masses, though with a high frequency of intususspection. We have yet to find any typical imaging features that could help in differential diagnosis.
Peutz-Jeghers syndrome consists of mucocutaneous pigmentation in the hands, feet, and perioral region, with gastrointestinal polyposis (hamartomas). Hamartomas may be located anywhere along the gastrointestinal tract, though they are most common in the small bowel. The polypoid lesions found in Peutz-Jeghers syndrome cannot be identified definitively on plain-film radiography, barium examination, or CT.[1]
Hemangiomas are less common than glandular or lipomatous tumors. They can be found throughout the gastrointestinal tract, though they are most commonly located in the small bowel. It is unusual, however, to find hemangioma in the duodenum.
Hemangiomas account for almost two-thirds of all vascular lesions in the small bowel. The classic pathological description includes three different patterns: cavernous, capillary, and diffuse (less common). Vascular masses can be covered by the mucosal or serosal layer and sometimes occupy the small bowel muscular coats.[1,4] Most are asymptomatic, but a few may present as palpable masses and/or with slight gastrointestinal bleeding.
Neither plain-film radiography nor barium studies can offer relevant findings for identifying small bowel hemangioma. They are difficult to spot on routine CT scans as well. Large masses will exhibit the classical behavior of delayed, progressive, and nodular enhancement following contrast. Phleboliths may be seen. MRI demonstrates hemangiomas as large masses, presenting with very high signal on T2-weighted imaging and low signal for T1-weighted studies. They are generally heterogeneous and may include calcifications.
Lymphangiomas occur 50% less often than hemangiomas. Imaging findings cannot be specific for such a rare tumor.
Between 2% and 6% of benign neoplasms in the small intestine are neurogenic tumors. These lesions arise from the subserosal nerve sheaths or from cells of the plexi of Auerbach and Meissner. They appear as polypoid masses along the antimesenteric border. Most are subserosal. Few, if any, have malignant potential.
There are five different types of neurogenic tumor:
- neurilemmoma (schwannoma): solitary and encapsulated;
- neurofibroma: multiple in cases of neurofibromatosis (von Recklinghausen disease);
- ganglioneuroma;
- paraganglioma; and
- gangliocytic paraganglioma.

Barium examination and CT may show a smooth-surfaced, well-defined polypoid mass that is occasionally ulcerated. Tumors may sometimes appear as intraluminal protrusions or as a subserosal mass. Small intramural lesions (less than 2 cm) are usually found in neurofibromatosis.[5]
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Disclosures:

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