

Duodenal tumors on cross-sectional imaging with emphasis on multidetector computed tomography: a pictorial review

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ABSTRACT

Duodenal tumors are uncommon, but they can cause significant morbidity and mortality. As stomach and colon are more common sites of gastrointestinal malignancies, radiologists sometimes may neglect the duodenum. Multidetector computed tomography (MDCT) and magnetic resonance imaging (MRI) can accurately locate and characterize mass-forming duodenal lesions, making them invaluable for the differential diagnosis and determining management strategies such as biopsy or surgery. Although conventional endoscopy continues to play an important role in the diagnosis of duodenal tumors, MDCT and MRI are very useful for evaluating the duodenal wall, extraduodenal space, and surrounding viscera, as well as the intraluminal content seen on endoscopy. This pictorial review aims to illustrate the most common benign and malignant mass-forming duodenal lesions and to focus on the imaging features that are most helpful in reaching the correct diagnosis.

The duodenum has both intraperitoneal and extraperitoneal segments (1). Unlike conventional endoscopy, cross-sectional imaging techniques—multidetector computed tomography (MDCT) and magnetic resonance imaging (MRI)—can analyze the duodenal wall, extraduodenal space, and surrounding viscera as well as the intraluminal content (2). Small duodenal lesions may be difficult or impossible to detect. Large tumors can manifest in different ways, such as a soft-tissue mass compressing surrounding viscera or a polypoid or thickening mural lesion, often with necrotic changes. In other cases, however, due to the lack of specificity of imaging features, it is essential to interpret the findings in the patient's clinical context, and the definitive diagnosis sometimes requires additional studies and histologic examination. This pictorial review illustrates the cross-sectional imaging features of the most common mass-forming lesions of the duodenum.

Benign tumors

Lipomas

The duodenum is the third most frequent site for lipomas in the gastrointestinal tract, after the colon and ileum (2). Duodenal lipomas are very rare, being most common in elderly men. They are often asymptomatic, but tumors greater than 4 cm in diameter can cause symptoms such as abdominal pain, obstruction, or gastrointestinal bleeding due to pressure, intussusception, or ulceration (3). CT is reliable in its diagnosis, showing a lesion with smooth margins and typical negative Hounsfield values (Fig. 1a). On MRI, signal loss on fat-suppressed sequences or a chemical-shift artifact surrounding the lesion on out-of-phase T1-weighted images is pathognomonic (Fig. 1b, 1c) (1, 2).

Duodenal polyps

Duodenal polyps smaller than 2 cm are benign and asymptomatic (4). Except in polyposis syndrome (Fig. 2), polyps tend to be solitary and rarely produce duodenal obstruction. The most common epithelial polyps in the duodenum are adenomatous, which tend to appear in the sixth decade of life (5). According to their growth pattern, they are classified as tubular, tubulovillous, or villous. Villous adenomas have malignant potential, so resection is indicated (1).

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Other adenomas are resected when symptomatic. CT has poor sensitivity for ampullary adenomas, detecting less than 50% (5). On contrast-enhanced CT, ampullary adenomas appear as smooth, frondlike, flat, or lobulated filling defects in the duodenal lumen, as an enhancing mass with smooth margins, or as a bulging and enhancing papilla. The definitive diagnosis of adenomas requires histopathologic study; CT's role is to detect malignant features (ulceration, vascular invasion, lymph nodes, or metastases). Another rare benign tumor, Brunner's gland hamartoma (>5 cm), mainly occurs in the proximal duodenum and usually appears as one or more submucosal or sessile polyps.

Gastrointestinal leiomyomas

Gastrointestinal leiomyomas are one of the causes of gastrointestinal bleeding. While they are the most common mesenchymal tumors in the esophagus, they are less likely to occur in the duodenum (6). Typically, they present as a rounded submucosal well-circumscribed mass with homogeneous enhancement in the absence of other lesions or metastases (Fig 3). Calcification and ulceration are more often seen with larger tumors. Radiologic findings of leiomyoma are similar to other submucosal mesenchymal tumors like GIST or schwannoma, being virtually impossible to differentiate a leiomyoma from GIST based on cross-sectional im-

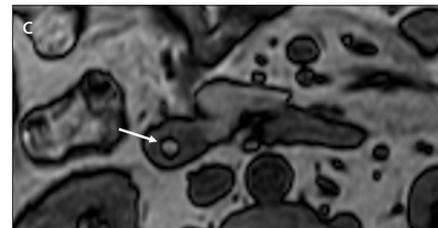


Figure 1. a–c. Duodenal lipomas in two different patients. Sagittal reconstructed contrast-enhanced CT image (a) shows a submucosal lesion (arrow) in duodenal bulb with negative Hounsfield values (-94 UH). On MRI, axial T1-weighted image (b) shows a hyperintense lesion (arrow), which demonstrates a surrounding chemical-shift artifact (arrow) in the out-of-phase T1-weighted image (c).

aging alone, particularly if necrosis and ulceration are present (6).

Malignant tumors

More than 60% of small bowel tumors arise in the duodenum and they have worse prognosis compared with other gastrointestinal malignancies (7, 8). The most common duodenal malignant tumors are discussed in this review: gastrointestinal stromal tumors, neuroendocrine tumors, adenocarcinoma, lymphoma, and metastases. The clinical and radiologic features of these tumors are shown in the Table. Some non-neoplastic entities, groove pancreatitis being the most important, may mimic a duodenal malignant tumor and histologic study may be required to differentiate between them.

Gastrointestinal stromal tumors (GISTs)

GISTs are the most common mesenchymal tumors of the gastrointestinal tract. GISTs can be classified as probably benign when they are smaller than 5 cm and the mitotic rate is less than 5 mitoses per 50 high-power fields. Less than 5% of GISTs affect the duodenum (9) nearly always in the second through the fourth parts. Duodenal GISTs seem to have a better prognosis than GISTs occurring in the stomach or other small-bowel regions (10). GISTs arise from the interstitial cells of Cajal, which are pacemaker cells for peristalsis located within the myenteric plexus. GISTs express the CD117 (c-kit) protein, a tyrosine kinase growth factor receptor, and CD117 expression distinguishes GISTs from less common

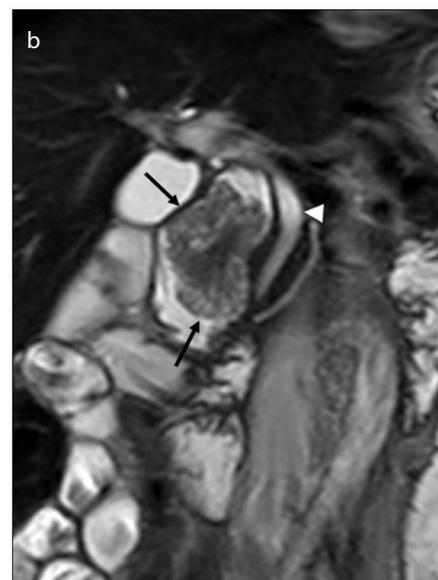
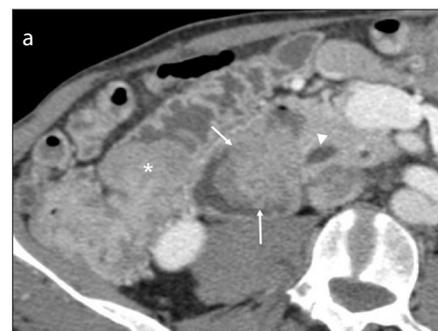


Figure 2. a, b. A 65-year-old man with juvenile polyposis syndrome (mutation in DPC4 - SMAD4) who had undergone multiple surgical procedures for recurrent subocclusive episodes. Curved-axial reconstructed contrast-enhanced CT image (a) and coronal-T2 weighted image (b) show an intraluminal duodenal polyp (arrows) causing moderate bile duct dilation (arrowhead). Note the colonic polyp (asterisk) in CT image (a).

Main points

- Duodenal neoplasms, including adenocarcinoma, carcinoid tumor, and gastrointestinal stromal tumors, represent a small percentage of gastrointestinal cancers.
- MDCT and MRI are noninvasive imaging modalities that can help to distinguish between malignant and benign lesions. Suspicious features of a lesion include irregular shape, ill-defined margins, heterogeneous enhancement, ulceration, and necrosis or cystic degeneration.
- Biliary and duodenal obstruction are rare in GISTs because they are usually soft. Instead, intratumoral hemorrhage, cystic and necrotic changes are common.
- Periampullary tumors show different growth patterns but “double duct” sign is typically associated with them as a result of upstream biliary and pancreatic ductal dilatation.
- The most common tumors metastasizing to the duodenum are melanomas, renal cell carcinomas and squamous cell carcinomas.

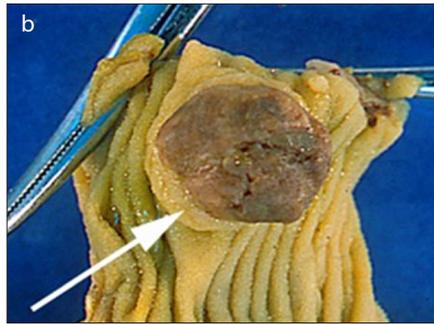
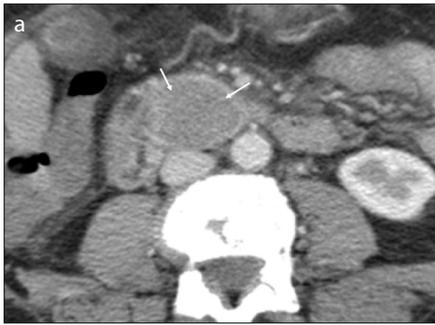


Figure 3. a, b. Duodenal leiomyoma diagnosed in a patient with obscure gastrointestinal bleeding. Axial contrast-enhanced CT image (a) shows a well-defined homogeneous duodenal mass (arrows), with a subtle enhancement after intravenous administration of iodinated contrast material. Histopathologic analysis of the lesion (b) confirmed the diagnosis of leiomyoma.



Figure 4. a, b. Duodenal gastrointestinal stromal tumors (GISTs) in two different patients. Axial contrast-enhanced CT images show two enhancing lesions (arrows) arising from the third portion of the duodenum (a) and the inferior duodenal flexure (b). Also note the central areas of low attenuation, suggesting necrosis.

mesenchymal neoplasms (leiomyomas, schwannomas, or neurofibromas). The predominant symptoms are gastrointestinal bleeding or anemia and abdominal pain or discomfort. Although they originate within the bowel wall, 30% to 40% of GISTs grow in an exophytic pattern, 29% to 44% in an intramural pattern, 18% to 22% in an endoluminal pattern, and 16% to 20% in a mixed pattern (3). Cross-sectional imaging techniques show smooth, well-circumscribed, hypervascularized tumors with varying degrees of intratumoral hemorrhage, necrosis, or cystic changes (Figs. 4 and 5). Calcification sometimes occurs similar to leiomyoma. Lymphatic involvement, biliary obstruction, and duodenal obstruction are uncommon (9). Malignant GISTs represent 10% of all malignant duodenal tumors (4) and are seen as a large exophytic tumor (Fig. 6). GISTs can be cured by surgical resection as long as there is no intraoperative tumor rupture. Chemotherapy with tyrosine kinase inhibitors is often administered, especially in patients with unresectable or

metastatic disease. The liver is the most common site of metastases, followed by the peritoneum (10).

Neuroendocrine tumors (NETs)

NETs account for 2% to 8% of all tumors of the gastrointestinal tract (11–13). About 90% of duodenal NETs are located in the first and second parts (2). G-cell tumors are the most common type of duodenal NET (65%), and one-third are functional (gastrinomas) (12). The next most common type is D-cell somatostatinomas, which occur exclusively in and around the papilla of Vater (1, 2). Carcinoid tumors are a subset of NETs that most often affect the gastrointestinal tract. Duodenal carcinoids represent less than 3% of all carcinoid tumors; most are sporadic, although they can also be associated with clinical syndromes such as multiple endocrine neoplasia type 1 (MEN-1) and neurofibromatosis type 1 (NF-1) (Fig. 7). Most carcinoid tumors are nonfunctional, so they present as bowel obstruction, abdominal pain, upper gastrointestinal bleeding, or when periampullary,

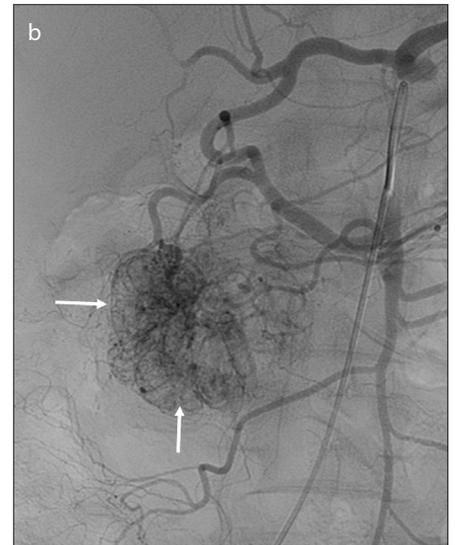


Figure 5. a, b. A 45-year-old woman with a duodenal GIST. Axial contrast-enhanced CT image (a) shows a very hypervascular soft-tissue mass (arrows) concentrated in the third portion of the duodenum. Arteriogram (b) shows that the main branches of the mass were coming from the inferior pancreaticoduodenal arch (arrows).

as jaundice. Unlike carcinoids in the midgut, duodenal NETs rarely manifest with classic carcinoid syndrome (12, 13).

On CT, NETs often appear as a focal polypoid mass that enhances during the arterial phase and may or may not wash out during the venous phase (Fig. 8). They can also present as an area of wall thickening or as an intramural mass. Early arterial-phase enhancement is an important criterion for distinguishing NETs from other duodenal masses. Between 10% and 20% of these tumors also have cystic features, which can be best evaluated on T2-weighted MRI. Nodal (10%) and distant (9%) metastases are uncommon (12). Enhanced regional adenopathy without an obvious primary lesion should raise strong suspicion of carcinoid tumor, and even more if a periduodenal desmoplastic reaction is identified (Fig. 9). Ampullary carci-

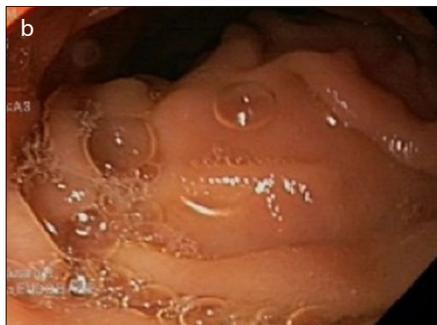
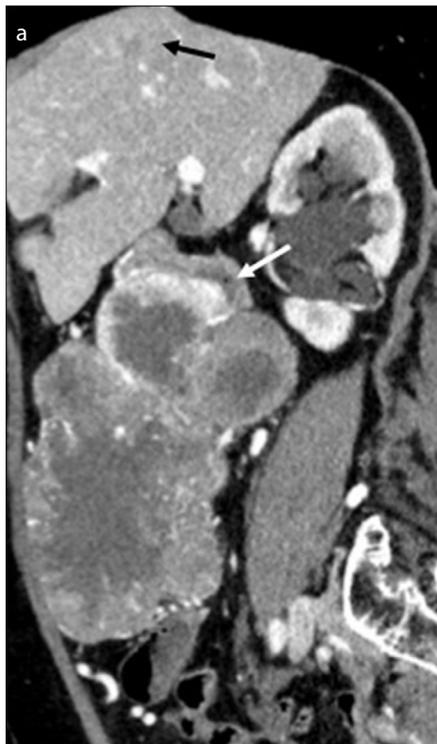


Figure 6. a, b. A 71-year-old woman with jaundice and abdominal pain. Sagittal reconstructed contrast-enhanced CT image (a) shows an enhancing mass (*white arrow*) arising from the second portion of the duodenum, associated with a large exophytic component with cystic and necrotic areas inside. Liver metastases are present (*black arrow*). Upper endoscopy (b) shows a soft, irregular mass protruding from the duodenum wall. The histopathological diagnosis was compatible with a malignant duodenal GIST.

noid tumors are usually more aggressive and can present with metastatic disease regardless of tumor size. The most common site of metastases is the liver, where they are usually seen as hypervascular lesions similar to the primary tumor.

Adenocarcinomas

Adenocarcinomas represent up to 80% to 90% of all primary malignant duodenal tumors (2). They usually present between the ages of 50 and 70 years and are slightly

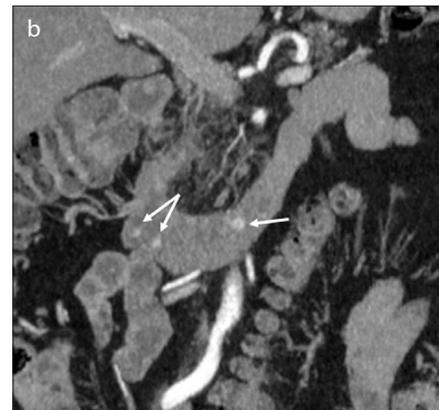


Figure 7. a, b. Multiple well-differentiated tiny enhancing duodenal carcinoid tumors (*arrows*) on axial (a) and coronal reconstructed (b) maximum-intensity CT images in a 56-year-old man with NF-1. An enlarged periduodenal lymph node is also seen (a, *short arrow*).

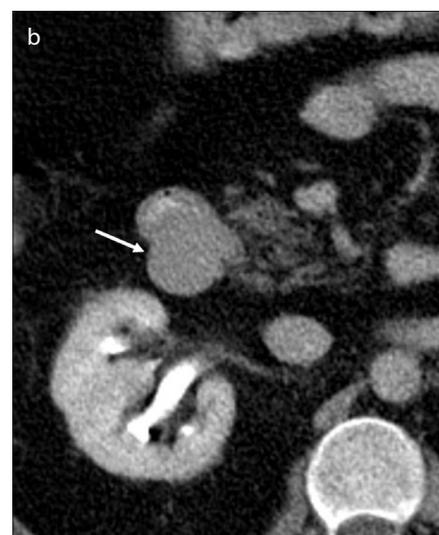
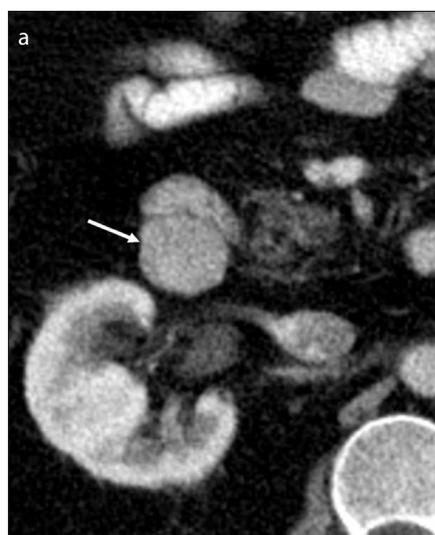


Figure 8. a, b. A 51-year-old man with MEN type-1. Axial contrast-enhanced CT image (a) reveals a well-defined, enhancing, exophytic mass arising from the descending duodenum (*arrow*). Axial excretory-phase contrast-enhanced CT image (b) demonstrates washout in the same mass (*arrow*). These findings are highly suggestive of a neuroendocrine tumor, and it was histologically confirmed.

more common in men (14). The risk of adenocarcinoma is increased in hereditary non-polyposis colorectal cancer, Peutz-Jeghers syndrome, and familial adenomatous polyposis. Nonspecific symptoms of duodenal adenocarcinomas include abdominal pain, weight loss, nausea, vomiting, occult gastrointestinal bleeding, pruritus, jaundice, and gastric outlet obstruction. On CT, the tumor can be seen as a concentric or asymmetric irregular thickening of a short segment of the bowel wall or a polypoid or fungating mass with or without luminal narrowing, necrosis, or ulceration (Fig. 10). Since they are predominantly fibrous, adenocarcinomas generally show mild and heterogeneous late enhancement. Adding diffusion-weighted images to MRI greatly increases the diagnostic accuracy, with high

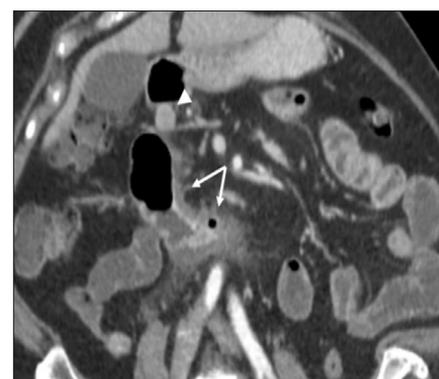


Figure 9. A 76-year-old woman with abdominal pain. Coronal-curved reconstructed contrast-enhanced CT image shows radiating bands emanating from the second duodenal portion due to desmoplastic reaction (*arrows*). A periduodenal hypervascular adenopathy (*arrowhead*) is associated. Histological study of the adenopathy confirmed a neuroendocrine tumor.

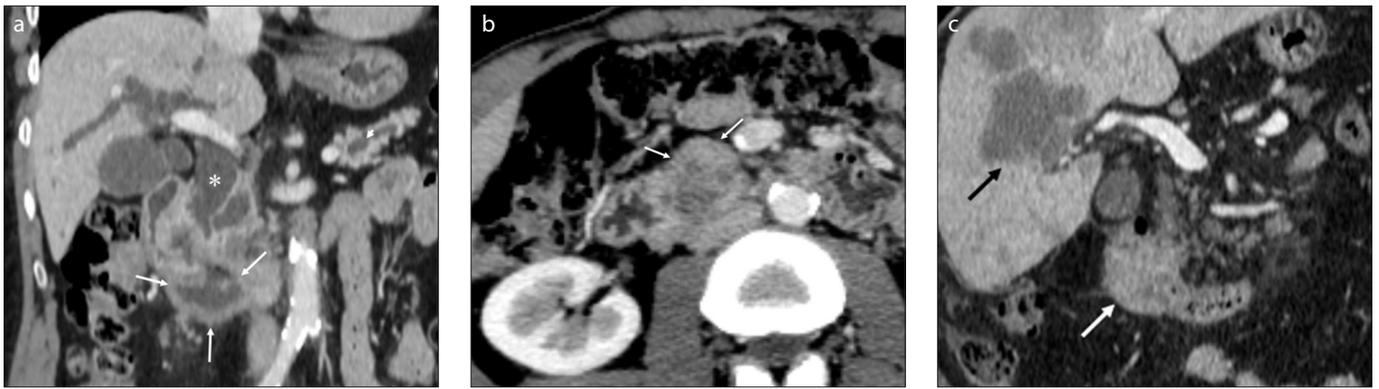


Figure 10. a–c. Duodenal adenocarcinomas in different patients. (a), an 82-year-old man with a periampullary tumor. Coronal reconstructed contrast-enhanced CT image shows severe dilation of the common bile duct (*asterisk*) and pancreatic duct (*short arrow*) secondary to an ulcerated duodenal mass (*arrows*). Histological study confirmed an intestinal-type adenocarcinoma; (b), a 51-year-old man with abdominal pain. Axial contrast-enhanced CT image reveals a heterogeneous exophytic endoluminal mass arising from the third part of the duodenum (*arrows*); (c), an 87-year-old man with jaundice and poor performance status. Coronal reconstructed contrast-enhanced CT image shows a homogeneous mass arising from the second portion of the duodenum, corresponding to a duodenal adenocarcinoma (*white arrow*); liver metastases are also seen (*black arrow*).

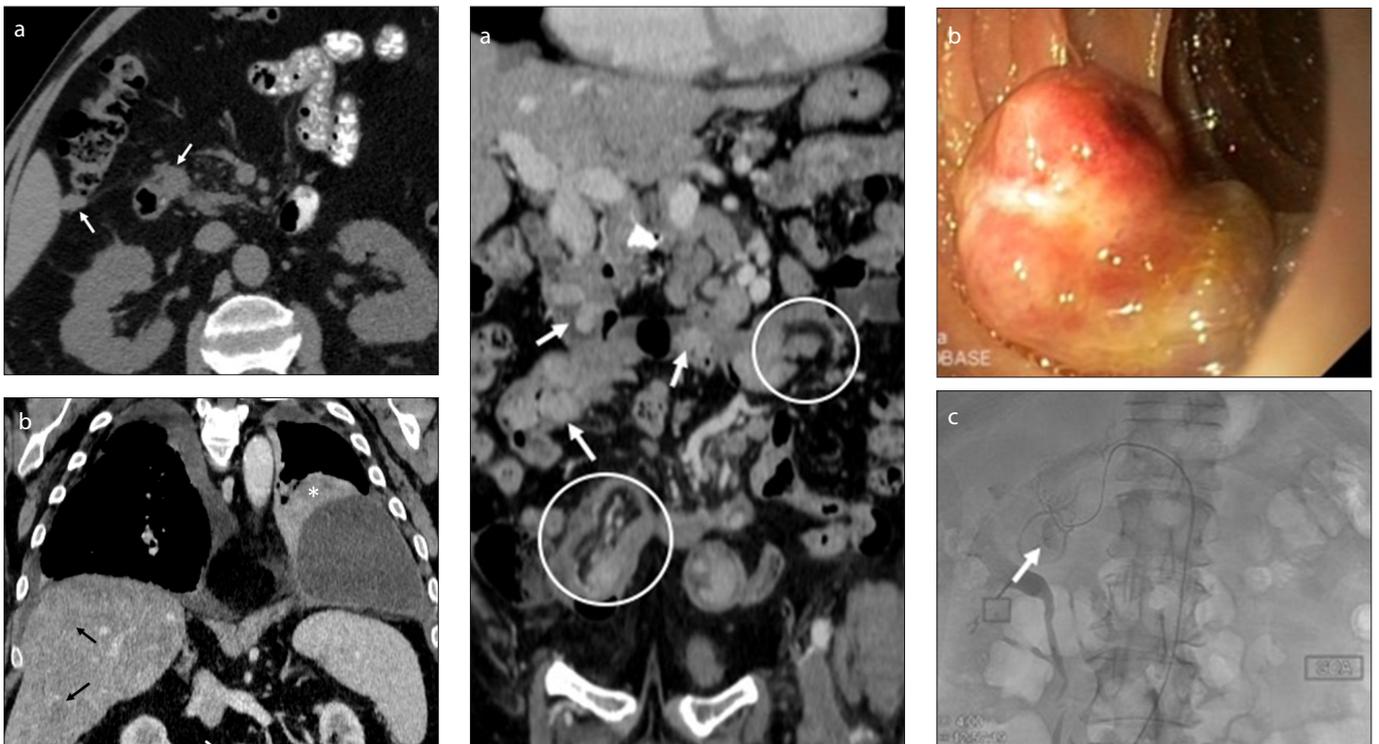


Figure 11. a, b. Axial unenhanced CT image (a) shows periduodenal and pericolic implants (*arrows*) in a patient with a subungual melanoma (hematogenous or peritoneal spread). Curved-coronal reconstructed contrast-enhanced CT image (b) shows a hypervascular duodenal implant (*white arrows*) and liver metastases (*black arrows*) in a patient with lung cancer (*asterisk*).

Figure 12. a–c. A 58-year-old man with a left renal cell carcinoma and intense upper gastrointestinal bleeding. Coronal-curved reconstructed contrast-enhanced CT image (a) shows multiple hyperdense focal nodules within the small bowel lumen compatible with hemorrhagic metastases (*arrows*). Incidentally, small bowel invaginations were identified (*circles*). Upper endoscopy (b) shows friable and bleeding duodenal nodules; for this reason, the gastroduodenal artery (*arrow*) was embolized (c).

(91%–96%) sensitivity and 100% specificity (5). Periampullary adenocarcinomas occur with 2 cm of the major duodenal papilla

and can cause bile and pancreatic duct dilation, which is seen as the “double-duct sign” (Fig. 10). An infiltrative growth pattern is commonly associated with pancreatobiliary-type periampullary tumors, whereas a polypoid growth pattern is typically seen in intestinal-type periampullary tumors. Locoregional invasion and lymph node involvement are often present from the onset

of duodenal adenocarcinomas. Node category is one of the most powerful predictors of survival, and lymphovascular invasion is a significant prognostic factor (15). Distant metastases are identified at onset in more than 50% of cases (2). In preoperative planning, the size of the tumor does not itself affect its resectability, but arterial invasion often makes resection difficult or impossi-

Table. Clinical and radiologic features of the most common duodenal tumors

	Anatomic location	Imaging features	Lymphadenopathy	Manifestations
Gastrointestinal stromal tumor	2nd to 4th portions	Well-circumscribed endophytic or exophytic mass Heterogeneously enhancing mass with variable necrosis, hemorrhage, or cystic changes May calcify	-	Biliary obstruction is rare Liver metastases +/- peritoneal involvement, if malignancy
Neuroendocrine tumor	1st and 2nd portions	Focal endoluminal mass, which may be hypervascular Ampullary tumors are more aggressive Primary tumor usually small, may be difficult to identify	+ Desmoplastic reaction sometimes present	Classic carcinoid syndrome is rare Usually multiple when MEN-1 or NF-1 Most non-functioning Bowel obstruction, jaundice, or UGIB Liver hypervascular metastases if malignancy
Adenocarcinoma	No predominance	Irregular short-segment thickening or intraluminal mass Necrosis and ulceration changes "Double-duct sign" if periampullary	+	50/70-year-old men Abdominal pain, weight loss, UGIB, gastric outlet obstruction, jaundice if periampullary Metastases to other organs
Lymphoma	No predominance	Homogeneously and intensely enhancing concentric wall thickening Classic "aneurysmal bowel dilatation" appearance of affected segment	+	Not occlusive (gastric outlet obstruction is not common) More common in immunosuppressed patients
Metastases	Duodenal or periduodenal	Variable in number, size and appearance	+ / -	History of primary cancer. Most common: melanoma, lung cancer, breast cancer, Kaposi sarcoma and renal cell carcinomas Routes of spread include intraperitoneal seeding, hematogenous and direct invasion

MEN-1, multiple endocrine neoplasia type 1; NF-1, neurofibromatosis type 1; UGIB, upper gastrointestinal bleeding.

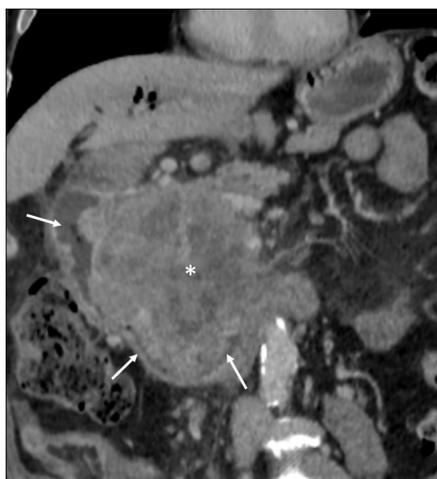


Figure 13. Curved-coronal reconstructed contrast-enhanced CT image shows pancreatic tumor (*asterisk*) progression within duodenum (*arrows*).

ble (16). Also, the surgical treatment would be the same in case of having a distal pancreas carcinoma or distal cholangiocarcinoma mimicking a periampullary adenocarcinoma.

Duodenal lymphomas

Duodenal lymphomas are non-Hodgkin T-cell lymphomas (2), which generally have an indolent, slow-growing course, although

up to 30% of cases can become aggressive (5). Risk factors for development of intestinal lymphoma include acquired immunodeficiency syndrome, inflammatory bowel disease, immunosuppression after solid organ transplantation, systemic lupus erythematosus, and chemotherapy. Segmental nodular wall thickening or a large eccentric mass extending into adjacent tissues are typical (producing circumferential wall thickening and irregular "aneurysmal dilatation" of the bowel lumen); gastric outlet obstruction is uncommon. Splenomegaly and lymphadenopathy in other areas help prospectively raise the diagnosis of lymphoma (6) and its presence helps distinguishing it from Crohn's disease.

Metastases

Metastases have a variety of appearances, even mimicking benign lesions when smoothly marginated or with homogeneous enhancement or mimicking malignant lesions when central ulceration/cavitation or invasion of adjacent structures are present (5, 17).

Routes of spread include intraperitoneal seeding, hematogenous and direct invasion. Regarding intraperitoneal seeding the most frequent are primary mucinous tumors of the ovary, appendix, or colon. Malignant tumors

that typically spread hematogenously are lung cancer, melanoma (Fig. 11), breast carcinoma, and renal carcinoma (Fig. 12). Finally, regarding the tumors that invade the duodenum directly, the most frequent are pancreatic, gastric, colonic, and hepatic carcinomas (Fig. 13), which may produce duodenal displacement, obstruction, or fistulization.

Conclusion

Duodenal neoplasms are uncommon, but they can cause significant morbidity and mortality. Cross-sectional imaging plays an important role in the assessment and management of mass-forming duodenal lesions. MDCT remains the first-line imaging modality for their evaluation. The imaging features of some duodenal tumors are nonspecific, but knowing the common sites of involvement and correlating the imaging presentation with the clinical presentation can often help reach the correct diagnosis. Awareness of these conditions will enable accurate diagnosis and prompt treatment.

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Conflict of interest disclosure

The authors declared no conflicts of interest.

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