CT role in distinguishing GIST from non-GIST mesenchymal gastric tumors

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Learning objectives

The purpose of this exhibition is to characterize the CT findings which could allow differentiation of GIST from other intramural gastric tumors.

Background

Mesenchymal gastric tumors are generally intramural gastric masses which present overlapping radiologic appearances. Gastrointestinal stromal tumors (GISTs) are the majority of intramural gastric tumors varying widely in appearance, from small intraluminal lesions to exophytic masses. Other less common mesenchymal tumors occur in the stomach namely lipoma, leiomyoma, sarcomas, plexiform fibromyxoma, glomic tumors and neuroendocrine tumors with different prognosis and treatment vs the much more common gastric GIST tumor.

Findings and procedure details

Submucosal lesions comprise a variety of neoplastic and non-neoplastic condition arising from deeper layers of the wall of the gastrointestinal tract (figure 1); the overlying mucosa is not involved.

Due to the submucosal location tumors arising from this layer usually attain a large size without causing obstruction by the time of diagnosis.

DISTINGUISHABLE FEATURES OF MESENCHYMAL GASTRIC TUMORS

Some CT features aid in the differential diagnosis of a mesenchymal gastric tumor (figure 2 - 5):

Location - Location of the tumor is important in establishing a differential diagnosis, because some tumors are seen more frequently in certain parts of the stomach. Leiomyomas are almost always found in the gastric cardia. GISTs and schwannomas often manifest in the body of the stomach. Glomus tumors, IFPs, lipomas, and ectopic pancreas are commonly seen in the gastric antrum.
**Attenuation** - Leiomyomas and schwannomas are usually low in attenuation. Even large schwannomas are characteristically well-circumscribed with homogeneous attenuation, a feature that may be useful in differentiating schwannomas from the more heterogeneous GISTs. Avid enhancement is often seen with glomus tumors, carcinoid tumors, small GISTs, metastases, ectopic pancreas, and hemangiomas. A mass of -70 to -120 is a lipoma.

**Growth pattern** - A polypoid growth pattern suggests a differential diagnosis of carcinoid tumor, IFP, GIST, adenomatous polyp, or polypoid cancer. Exophytic growth patterns include GIST, schwannoma, lymphoma, or poorly differentiated adenocarcinoma. Among this group of tumors, GIST is typically heterogeneous with areas of cystic or necrotic change and lacks lymphadenopathy.

**Size** - leiomyomas, glomus tumors, IFPs, and ectopic pancreas are usually less than 5 cm. The presence of multiple lesions should raise the possibility of carcinoid tumors, multiple polyps, or metastases.

**SPECIFIC MESENCHYMAL GASTRIC TUMORS**

**GASTROINTESTINAL STROMAL TUMOR (GIST)**

In the esophagus, leiomyomas are more common than GIST; however, in the stomach, small intestine, and colon, GISTS are the most common mesenchymal tumors.

Gastrointestinal stromal tumors (GISTs) are mesenchymal neoplasms of the gastrointestinal (GI) tract and are thought to develop from the interstitial cells of Cajal. GISTs are typically defined by the expression of c-Kit (CD117) in the tumor cells, as these activating KIT mutations are seen in 85-95% of GISTs. Although radiologic or histologic results may suggest GISTs, the diagnosis must be made immunochemically. Independent of location, most GISTs express the CD34 antigen (70-78%) and the CD117 (72-94%) antigen.

About 50-70% of gastrointestinal stromal tumors (GISTs) occur in the stomach; 33%, in the small bowel; 5-15%, in the rectocolon; and only 1-5%, in the esophagus. GISTs are multicentric in fewer than 5% of cases.

Grossly GISTs are well-demarcated spherical masses that appear to arise from the muscularis propria layer of the gastrointestinal wall. Intramural in origin, they often project exophytically and/or intraluminally, and they may have overlying mucosal ulceration. The enhancement pattern can vary from homogenously enhancing to heterogeneously enhancing, with or without ulceration (figure 6 - 9). Smaller GISTs appear as smooth, sharply defined intramural masses with homogeneous attenuation (figure 10). Larger
GISTs nearly always outgrow their vascular supply, leading to extensive areas of necrosis and haemorrhage (figure 11, 12). Lymph node metastases are uncommon.

CT is the ideal image modality in defining the endoluminal and exophytic extent of tumor.

Differential diagnosis of GISTs includes schwannomas, true leiomyomas, and solitary neuroendocrine tumors, particularly for smaller lesions. Occasionally, gastric adenocarcinoma or lymphoma may demonstrate intramural growth and mimic a GIST. However, advanced gastric carcinomas and lymphomas are usually associated with bulky perigastric or celiac lymphadenopathy, which is rare for GISTs. Other more rare entities can also mimic the appearance of a GIST (figure 13).

NON-GIST SARCOMA

Although they are rare, sarcomas other than GIST, such as liposarcomas, leiomyosarcomas, and unclassified sarcomas, may form in the stomach. These lesions are usually large, aggressive tumors with heterogeneous enhancement and areas of necrosis. Their appearance is nonspecific, and histologic confirmation is required for diagnosis.

It is extremely important to accurately differentiate non-GIST sarcomas from GISTs because GISTs respond favorably to imatinib, whereas non-GIST sarcomas have a less predictable response to conventional chemotherapy.

LEIOMYOMA

True gastric leiomyomas are rare and are negative for c-KIT and strongly and diffusely positive for desmin and smooth muscle actin.

The distinction between GISTs and leiomyomas is clinically important, because leiomyomas are benign and GISTs are associated with a variable risk of progression and metastasis. In the stomach, small bowel, and colon, GISTs far outnumber leiomyomas. The esophagus is the only site where leiomyomas predominate.

Leiomyomas are almost always seen in the gastric cardia as homogeneous, low-attenuation masses with an endoluminal growth pattern, often ranging from 1.3 to 4.7 cm in diameter (figure 14, 15, 16). Tumors larger than 2 cm can have central ulceration.

Differential diagnosis of leiomyomas includes GISTs (figure 17) and schwannomas.

LIPOMA
On CT images, an intramural mass with attenuation values of #70 to #120 HU is diagnostic of a lipoma (Figure 18). Three-quarters of lipomas are solitary lesions, with the most common location being the gastric antrum. The majority (95%) of lipomas are endoluminal, arising in the submucosa. One exception to this is the presence of ulceration, which can result in inflammation that manifests with soft-tissue attenuation, and can be endoluminal, arising in the submucosa. Lipomas infrequently develop into pedunculated lesions which can prolapse through the pylorus and cause intermittent gastric outlet obstruction. Unless the patient is symptomatic, treatment is not indicated.

**NEUROGENIC TUMORS**

Schwannomas (benign tumors of the nerve sheath) account for 2%-7% of gastrointestinal mesenchymal tumors. Schwannomas are believed to arise from the myenteric plexus within the muscularis propria of the gastrointestinal tract, with the stomach being the most common site (60%-70% of cases). Schwannomas arise most often in the gastric body, demonstrating an exophytic or intramural pattern of growth. A notable feature of schwannomas is their homogeneous attenuation, independently of the size (Figure 19). It shows low attenuation on unenhanced CT images because of the dense spindle cell composition and contrast-enhanced CT may show no or minimal enhancement during the arterial phase and delayed enhancement during the equilibrium phase.

Cystic degeneration is uncommon for gastric schwannomas and calcification is rare. Absence of hemorrhage, necrosis, or cavity formation is a useful feature of schwannomas, and can be used to distinguish schwannomas from the more heterogeneous GISTs.

**GLOMUS TUMOR**

Glomus tumors arise from modified smooth muscle cells of the glomus bodies, which are neuromyoarterial receptors that regulate body temperature. Nearly all gastrointestinal glomus tumors arise in the muscularis propria of the stomach, and they account for 2% of all gastric tumors.

Glomus tumors most commonly manifest as a solitary hypervascular lesion in the gastric antrum and range from 1 to 4 cm in diameter. They show dense arterial phase enhancement which persists in the delayed phase (Figure 20). A peripheral nodular pattern of enhancement with delayed filling-in, similar to a hemangioma, has been reported as a specific feature of gastric glomus tumors.

Cystic degeneration is uncommon for gastric schwannomas and calcification is rare. Absence of hemorrhage, necrosis, or cavity formation is a useful feature of schwannomas, and can be used to distinguish schwannomas from the more heterogeneous GISTs.
Differential diagnosis of glomus tumors includes GIST, carcinoid tumor, metastasis, ectopic pancreas, and hemangioma.

HEMANGIOMA

It is unknown whether these lesions are true neoplasms or congenital malformations. Hemangiomas are usually solitary, vascular tumors that may occur anywhere in the gastrointestinal tract. Gastric hemangiomas represent 1.6% of all benign gastric tumors. Gastrointestinal hemangiomas can manifest at any age; however, they are commonly seen in early childhood and gradually involute.

Contrast-enhanced CT shows an avidly enhancing, often intraluminal mass. It can also mimic its hepatic counterparts (figure 21). Phleboliths within the lesion are virtually pathognomonic.

INFLAMMATORY FIBROID POLYP (IFP)

They are usually solitary and can be found anywhere in the gastrointestinal tract, but 75% occur in the gastric antrum, followed by the gastric body and fundus. The tumors are often 2-5cm in diameter, with a smooth or slightly lobulated contour.

Because they arise in the submucosa, they manifest as intraluminal lesions, which may mimic the appearance of adenomatous polyps, intraluminal GISTs, carcinoid tumors or schwannomas.

PLEXIFORM FIBROMYXOMA

Plexiform fibromyxoma is a benign tumor that appears to be unique to the stomach, almost always occurring in the gastric antrum. They range in size from 2 cm to 15 cm and can demonstrate ulceration or mucosal invasion.

On CT images the tumor demonstrates areas of low attenuation owing to presence of myxoid tissue, interspersed with foci of vascularity. On MR images the myxoid stroma of the tumor is T2-hyperintense with persistent enhancement after administration of contrast material.

The differential diagnosis of plexiform fibromyxoma includes schwannoma, GIST, neurofibroma, and myxoid liposarcoma.

NEUROENDOCRINE TUMORS
These tumors occur most frequently in the gastrointestinal tract, the small bowel is the most common location, followed by the rectum, appendix, and stomach. Gastric carcinoid tumors are rare, accounting for 1.8% of all gastric malignancies (figure 22).

Carcinoid tumors usually originate from enterochromaffin-like cells in the gastric mucosa and are therefore epithelial in origin. However, often the bulk of the tumor is submucosal and they should be included in the differential diagnosis of a submucosal gastric tumor.

Clinicopathologic characterization of gastric carcinoid tumors has revealed three subtypes. Type I tumors are associated with enterochromaffin-like cell hyperplasia, hypergastrinemia, and chronic atrophic gastritis, with or without pernicious anemia. Type I gastric carcinoid tumors generally represent benign disease. Nodal and hepatic metastases are very rare, occurring in 2% of the patients in the larger studies and causing no tumor-related deaths.

Type II gastric carcinoid tumors are the least common type, representing 5-10% of gastric carcinoid tumors. They are seen in the hypergastrinemic state of Zollinger-Ellison syndrome in association with MEN-I (multiple endocrine neoplasia). Approximately 30% of patients with MEN-I will have gastric carcinoid tumors. Type II carcinoids are multicentric and variable in size but are prone to developing local lymph node metastasis. Tumor-related death is rare, as is carcinoid syndrome. The appearance of these tumors on CT scans can be striking because there are multiple masses in the setting of diffuse gastric wall thickening (figure 23).

Type III gastric carcinoid tumors are sporadic tumors and are not associated with a hypergastrinemic state. They represent about 13% of gastric carcinoid tumors. Unlike types I and II tumors, type III gastric carcinoid tumors are large solitary tumors that may show ulceration and are more likely to be invasive with distant metastases (figure 24). Among patients diagnosed with type III carcinoid tumors, 80% are men. Such tumors are more aggressive and should be treated with total gastrectomy with enbloc removal of regional lymph nodes when liver metastases are not present.

Images for this section:
Fig. 2: Schematic illustration of the most common mesenchymal gastric tumors. IF. Inflammatory polyp. NE. Neuroendocrine. GIST. Gastrointestinal stromal tumor.

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**Fig. 6:** Gastric GIST. Axial unenhanced (a) and arterial (b) and venous phase (c) CT images showing a large, well-defined, exophytic and heterogeneously enhancing mass centered in the gastric body, with low density areas suggestive of necrosis.

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**Fig. 19:** Gastric schwannoma. Axial unenhanced (a) and enhanced arterial (b) and venous phase (c) CT images showing a lobulated, exophytic, spontaneously low density mass located in the gastric body. It shows slightly heterogeneous arterial phase enhancement (b) and delayed homogeneous enhancement (c). Both the location, growth and enhancement pattern are suggestive of a gastric schwannoma which was biopsy proven.

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**Fig. 20:** Gastric glomus tumor. Axial unenhanced (a) and venous phase (b) CT images showing a solitary, intramural, nodular lesion at the gastric antrum with intense, slightly heterogeneous enhancement. Although an arterial phase was not obtained, the location, size and avid enhancement are characteristic of a gastric glomus tumor which was biopsy proven.

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Fig. 21: Gastric hemangioma. Axial CT images showing a lobulated nodular mass in the gastric body. It shows faint peripheral enhancement in the late arterial phase (a) and progressive globular enhancement in the venous phase (b), suggestive of a gastric hemangioma. Note the presence of an additional hepatic hemangioma (arrow).

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Fig. 14: Gastric leiomyoma. Axial CT images showing a homogeneous low-attenuation mass with an endoluminal growth pattern and homogeneous enhancement. This type of enhancement is suggestive of a gastric schwannoma or leiomyoma, however schwannoma tend to be exophytic.

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**Fig. 18:** Gastric lipoma. Axial CT images showing a well defined intramural antral mass with fat density (-115 HU) which is diagnostic of a gastric lipoma.

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Fig. 15: Gastric leiomyomas. Axial CT non-enhanced (a,c) and venous phase (b,d) images from two different patients showing gastric leiomyomas - characteristically well-defined, hypodense and homogeneous intraluminal mass located in the cardia.

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**Fig. 10:** Small GISTs. Axial CT images demonstrating two small GISTs of the gastric antrum (arrows) - well-defined, slightly enhancing intramural nodules. This appearance mimics a glomus tumor that could be differentiated by a more pronounced enhancement (arterial phase images could be helpful in the distinction).

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**Fig. 16:** Atypical gastric leiomyoma. Axial CT images showing a lobulated low-attenuation mass at the gastroesophageal junction narrowing the distal esophagus. It was histologically proven to be a gastric leiomyoma.

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**Fig. 17:** GISTs of the gastric fundus mimicking leiomyomas. Axial enhanced CT images showing fairly homogenous hypoenhancing well defined nodules at the gastric fundus, resembling gastric leiomyomas. a. The lesion (oblique arrow) is inconspicuous and cannot be differentiated from a gastric leiomyoma. b. The nodule shows tiny foci of low attenuation that can be due to hemorrhage. c. The nodule demonstrates very faint internal heterogeneity favoring GIST over leiomyoma.

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**Fig. 11:** Massive gastric GIST. Axial (a) and coronal (b) CT images illustrating a very large, well-defined, markedly heterogeneous lesion with mass effect on the nearby organs (apparently not infiltrative). The lesion was resected with no evidence of recurrence.

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Fig. 9: Various antrum GISTs. Axial enhanced CT images demonstrating various appearances of antrum GISTs. From an intraluminal well-defined, smoothly margined nodule (a) to an exophytic fairly homogeneous mass (b) (note the the integrity of the mucosa line) or heterogenous hypoenhancing (c) and hyperenhancing (d) masses with infiltrative margins.

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**Fig. 12:** GIST of the gastric fundus with liver metastases. Axial CT image demonstrating a large ulcerated heterogeneous mass in the gastric fundus and several metastatic lesions in the hepatic parenchyma.

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Fig. 7: Typical GISTs of the gastric body. Axial non-enhanced (a) and enhanced (b) CT images depicting an intraluminal, well-defined, smoothly marginated, heterogeneously enhancing mass. Axial CT image (c) showing a similar lesion although more homogenous and exophytic. Axial non-enhanced CT image (d) illustrating a slightly hyperdense nodular exophytic lesion with a coarse calcification.

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**Fig. 8:** GISTs of the gastric fundus. Axial CT enhanced images presenting the variety of appearances of gastric fundus GISTs showing a well-defined intraluminal mass slightly heterogeneous (a), a fairly heterogeneous exophytic mass (b) and an ulcerated, markedly heterogeneous enhancing and invasive mass (c).

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**Fig. 13:** Axial (a, b) CT images showing a large, solitary, heterogeneous nodular mass apparently centered in the gastric wall (small curvature). Note the apparent integrity of the gastric mucosa and the absence of lymphadenopathy. This appearance is somewhat characteristic of a GIST but was histologically classified as a rare tunica intima gastric artery sarcoma.

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**Fig. 22:** Two gastric neuroendocrine tumors histologically confirmed. a. Unenhanced CT images showing diffuse gastric wall thickening (no contrast was given to this patient which
could depict intramural enhancing nodules). b. Enhanced CT images demonstrating an infiltrative, enhancing, exophytic gastric wall mass. The anterior portion of the mass configures an inconspicuous heavily enhancing nodule.

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**Fig. 23:** Gastric multicentric type II neuroendocrine tumor. PET-CT images showing intramural and exophytic gastric wall nodules with avid 18FDG uptake. Liver metastasis also show radionuclide uptake.

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**Fig. 24:** Sporadic (type III) gastric neuroendocrine tumor. CT images with oral contrast (a) and CT images with oral and intravenous contrast (b) showing a large intramural mass markedly ulcerated and associated liver metastasis.
Fig. 3: Distinguishable features of gastric mesenchymal tumors - emphasis on "attenuation". Adapted from Kang et al. RadioGraphics 2013; 33:1673-1690
**Fig. 4:** Distinguishable features of gastric mesenchymal tumors - emphasis on location. Adapted from Kang et al. RadioGraphics 2013; 33:1673-1690

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**Fig. 5:** Distinguishable features of gastric mesenchymal tumors - emphasis on growth pattern, size, margins and associated findings. Adapted from Kang et al. RadioGraphics 2013; 33:1673-1690

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**Fig. 1**: Most common differential diagnosis of a gastric submucosal mass.

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Conclusion

A thorough knowledge of characteristic findings is necessary to differentiate between mesenchymal gastric tumors types. A combination of features such as location, attenuation, enhancement and growth pattern may suggest one diagnosis over another. GISTs account for the majority of intramural tumors and can vary from small intraluminal lesions to exophytic masses, usually with areas of hemorrhage or necrosis. Lipomas are well-circumscribed masses from -70 to -120 HU; leiomyomas generally present as low-attenuation mass at the gastric cardia; a hypervascular mass in the antrum is a common manifestation of glomus tumors; gastric schwannomas characteristically have a homogeneous attenuation; inflammatory fibroid polyps usually arise as a polypoid mass in the antrum and plexiform fibromyxomas are rare, usually antral tumors. Although gastric neuroendocrine tumors are epithelial in origin often they present submucosal location and therefore should be distinguished from other intramural lesions.

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