Leiomyoma and spectrum of submucosal tumors of the GI tract

Gateorn Pongarnar
Chulalongkorn University, Year VI
Gillian Lieberman, MD
Overview

• Patient presentation
• Radiologic findings
• Differential diagnosis
• Spectrum of submucosal tumors
• Endoscopic ultrasonography
• Hospital course summary
• Take home messages
Patient Presentation

• A 20-year-old healthy woman desired to attend the beauty contest.
• Screening chest X-ray was abnormal.
• No past history of medical illness
• Physical examination and laboratory results are within normal limits.
Our patient: chest X-ray
Time is up !!!

....... 

Could you pick up anything abnormal?
Our patient: chest X-ray

- Soft tissue density with the crescent of adjacent air in the left upper abdomen (stomach)
- Otherwise are unremarkable

Source: PACS, KCMH
Our patient: mass on CT

Axial abdominal CT+oral contrast
Our patient: mass on CT

How do you describe the findings of this CT image?
Our patient: mass on CT

- Homogeneous low attenuation polypoid intraluminal mass
- well-defined margin
- smooth surface
- without evidence of invasion or obstruction
- located at the cardia of stomach

Source: PACS, KCMH
Axial abdominal CT+oral contrast
Our patient: differential diagnosis

- Our patient was asymptomatic and young.
- Nothing found remarkably on her physical examination.
- Malignancy is less likely
- So what kind of benign gastric tumor can present with this features?

Let’s see the classification of benign gastric mass ......
Benign gastric mass: Classification

Mucosal mass
- Non-neoplastic polyp
  - Harmatomatous polyp
  - Juvenile polyp
  - Hyperplastic polyp
- Neoplastic polyp
  - Adenomatous polyp
  - Fundic gland polyp

Submucosal mass
- Neoplastic polyp
  - mesenchymal
    - GISTs (*may not be benign*)
      - Leiomyoma
      - Schwannoma
      - Lipoma
      - Vascular tumors
  - Non-mesenchymal
    - Heterotopic pancreas
    - Brunner’s gland harmatoma
    - Duplication cyst
    - Gastric cystic profunda
    - Intramural pseudocyst
Our patient: differential diagnosis

Some types of gastric mass can be differentiated by CT scan

Here are examples of CT scan of common benign gastric tumors
Companion pt#1: Inflammatory fibroid polyp

Axial C+ abdominal CT shows an intraluminal pedunculated mass in the antrum of the stomach.

Companion pt#2 : small GIST

Axial C+ abdominal CT shows a homogeneous low-density mass located in the fundus of the stomach with the intact overlying mucosa enhancing to a greater degree than the mass.

Axial C+abdominal CT shows a heterogeneous low-attenuation mass with the focal areas of necrosis arising from the stomach (high risk of malignancy).
Axial C+ abdominal CT shows a homogeneous low-attenuation mass with central ulceration in case of large size.

Companion pt#5: Schwannoma

Axial C+ abdominal CT obtained with water as an oral contrast agent shows the well-circumscribed mass with homogeneous enhancement.

Axial C+ abdominal CT shows an ill-defined ovoid endoluminal submucosal mass in the gastric antrum. The lesion shows homogeneous enhancement and higher attenuation than the pancreas.
Axial C+ abdominal CT shows a **well-circumscribed** submucosal mass with uniform fat attenuation.

Lipoma can be definitely diagnosed by CT scan.
CT scan could be used to narrow down the differential diagnoses

........

Now let’s go back to our patient’s abdominal CT image......what is the most likely diagnosis?
Our patient: mass on CT

- Homogeneous low attenuation polypoid intraluminal mass
- well-defined margin
- smooth surface
- without evidence of invasion or obstruction
- located at the cardia of stomach

Source: PACS, KCMH
Axial abdominal CT + oral contrast

Gateorn Pongarnar, Year VI
Gillian Lieberman, MD
Our patient: further investigation

No definite diagnosis could be made by only CT scan, except for Lipoma

The next appropriate investigation for intraluminal mass of the stomach should be gastroscopy

Let’s see the companion patient’s image of gastroscopy
Companion pt#8: Endoscopic findings of submucosal tumor

- An elevated submucosal mass at the upper part of the stomach
- The overlying mucosa is intact
- Suggesting a submucosal lesion

It’s not mucosal lesion because the overlying mucosa was intact
And not extraluminal compression indicated by CT scan

Now, the diagnosis is focused on submucosal lesion
Let’s go through a bit more about submucosal lesion of the upper GI tract ......
Submucosal lesions: definition

Definition: A variety of neoplastic and non-neoplastic condition arising from deeper layers of the wall of the GI tract, the overlying mucosa is not involved.
Submucosal lesion : GI tract wall

Submucosal lesions: overview

- Incidence: 0.4% of diagnostic scope
- Usually asymptomatic
- Discovered as accidental findings
- Symptoms: abdominal pain, bleeding, obstruction and intussusception

**CT scan and Endoscopic ultrasonography (EUS)** are used to narrow down the differential diagnoses and to assess the risk of malignancy

Endoscopic ultrasonography (EUS)

• The first choice for examining submucosal lesions in the upper GI tract.

• Important application of EUS is to stage GI malignancy, also to differentiate the types of submucosal tumors by their original layers.

• The sensitivity of cytological samples achieved through EUS-FNA has been reported to be 88%-91% and the specificity was 100% for the diagnosis of malignant lesions.

EUS images of stomach wall

Alternating echogenicity of layers of the GI tract wall

Companion pt#9: EUS images of tumor origins

MM = Muscularis mucosae
SM = Submucosal layer
PM = Muscularis propia

## Submucosal tumors: Origins

<table>
<thead>
<tr>
<th>Histological layer</th>
<th>Superficial mucosa</th>
<th>Deep mucosa, muscularis mucosa</th>
<th>Submucosa</th>
<th>Muscularis propria</th>
<th>Serosa/adventitia</th>
</tr>
</thead>
<tbody>
<tr>
<td>EUS layer</td>
<td>1st</td>
<td>2nd</td>
<td>3rd</td>
<td>4th</td>
<td>5th</td>
</tr>
<tr>
<td>Mesenchymal tumors</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Leiomyoma</td>
<td></td>
<td>+</td>
<td></td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>Schwannoma</td>
<td></td>
<td></td>
<td></td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>GIST</td>
<td></td>
<td>+</td>
<td>+</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Other tumors</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ectopic pancreas</td>
<td>+</td>
<td></td>
<td>(+)</td>
<td></td>
<td>(+)</td>
</tr>
<tr>
<td>Cysts</td>
<td></td>
<td>+</td>
<td></td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>Lipoma</td>
<td></td>
<td></td>
<td></td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>Carcinoid</td>
<td></td>
<td>+</td>
<td></td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>Glomus tumor</td>
<td></td>
<td></td>
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<td>+</td>
</tr>
</tbody>
</table>

Gastric submucosal tumors: classification

Mesenchymal tumors
- Myogenic
- Neurogenic
- GISTs
- Leiomyoma
- Schwannoma

Non-mesenchymal tumors
- Heterotopic pancreas
- Brunner’s gland harmatoma
- Duplication cyst
- Gastric cystic profunda
- Intramural pseudocyst

> 50% of submucosal tumors

Mesenchymal tumors : DDx by EUS

- Differentiation among leiomyoma, schwannoma and GIST is extremely difficult by imaging modalities.
- 50% of GIST and schwannoma: complete or incomplete marginal hypoechoic halo from EUS findings. (distinct marginal halo is not seen in leiomyoma).
- Echogenicity of tumor comparing to the surrounding proper muscle from EUS findings.

<table>
<thead>
<tr>
<th>Tumor</th>
<th>Echogenicity</th>
</tr>
</thead>
<tbody>
<tr>
<td>GIST</td>
<td>slightly higher</td>
</tr>
<tr>
<td>Leiomyoma</td>
<td>nearly equal</td>
</tr>
<tr>
<td>Schwannoma</td>
<td>extremely low</td>
</tr>
</tbody>
</table>

In the past, we thought GISTs = Leiomyoma but not any more

......

Let’s go through more details about GISTs and leiomyoma
Submucosal tumors: GISTs

- GIST occurs more in stomach and small bowel than esophagus.
- 10-30% of GIST are eventually malignant.
- GIST expressed KIT (CD 117) and/or CD34 antigen.
- Follow up is needed after surgical resection.
- Imatinib mesylate is used to shrink large GISTs before surgery and it is the first line treatment for recurrent or metastatic GISTs.

Submucosal tumors: Leiomyoma

- Leiomyomas are benign tumors without malignant potential.
- Most common are found at esophagus but are rare in the stomach and duodenum.
- Leiomyoma: non-expression of KIT (CD 117) but express desmin and/or actin.
- Most tumors are asymptomatic.
- No recurrence after surgical resection

EUS findings:

Gastric GISTs VS Leiomyoma

• Inhomogenicity, hyperechogenic spots, a marginal halo and higher echogenicity as compared with the surrounding muscle layer from EUS findings appeared more frequently in GIST than in leiomyoma.

• GIST size > 3.5 cm: increase risk of malignancy

Now, let’s go to the patient’s hospital course summary and definite diagnosis

..........
Our patient: Hospital course

• Surgery was designed after CT scan and gastroscopy suspecting GISTs or gastric leiomyoma, *EUS was not done.*
• Proximal esophagogastrectomy was performed.
Our patient: Surgical specimens

Courtesy of Dr. Sirichindakul, KCMH
The surgical specimens were sent to the pathologist

.........

Let’s see the results ....
Our patient: Histology result

- Well demarcated border *spindle cell tumor*, lack of pleomorphism and scant mitotic count

**Benign spindle cell tumors**

- GISTs
- Leiomyoma
- Schwannoma
Histological result helps focus on benign spindle cell tumors of the stomach

To confirm definite diagnosis, immunostaining is needed.
Spindle cell tumor: Immunostaining

Table 6. Frequency of Immunostaining for Each Antibody in Gastrointestinal Stromal Tumors (GISTs) and Other Spindle Cell Tumors of the Gastrointestinal Tract

<table>
<thead>
<tr>
<th>Tumor Type</th>
<th>KIT/CD117 (%)</th>
<th>CD34 (%)</th>
<th>Desmin (%)</th>
<th>Smooth-Muscle Actin (%)</th>
<th>S-100 (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>GIST (n = 211)</td>
<td>100</td>
<td>91</td>
<td>4</td>
<td>31</td>
<td>8</td>
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<tr>
<td>Stomach (n = 177)</td>
<td>100</td>
<td>98</td>
<td>5</td>
<td>24</td>
<td>3</td>
</tr>
<tr>
<td>Small intestine (n = 22)</td>
<td>100</td>
<td>32</td>
<td>0</td>
<td>82</td>
<td>41</td>
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<tr>
<td>Smooth-muscle tumor</td>
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<td></td>
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<tr>
<td>Leiomyoma (n = 12)</td>
<td>0</td>
<td>0</td>
<td>100</td>
<td>100</td>
<td>0</td>
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<tr>
<td>Leiomyosarcoma (n = 18)</td>
<td>0</td>
<td>0</td>
<td>100</td>
<td>100</td>
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<td>Peripheral nerve sheath tumor</td>
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<tr>
<td>Schwannoma (n = 14)</td>
<td>0</td>
<td>50</td>
<td>0</td>
<td>0</td>
<td>100</td>
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<tr>
<td>Fibrous tumor</td>
<td></td>
<td></td>
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<tr>
<td>Solitary fibrous tumor (n = 17)</td>
<td>0</td>
<td>100</td>
<td>0</td>
<td>0</td>
<td>24</td>
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<tr>
<td>Desmoid-type fibromatosis (n = 25)</td>
<td>0</td>
<td>0</td>
<td>20</td>
<td>84</td>
<td>0</td>
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</table>
Our patient: Immunostaining

<table>
<thead>
<tr>
<th>CD-117</th>
<th>CD-34</th>
<th>SMA</th>
<th>S-100</th>
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</thead>
<tbody>
<tr>
<td>-</td>
<td>-</td>
<td>+</td>
<td>-</td>
</tr>
</tbody>
</table>

GISTs   GISTs   Leiomyoma   Schwannoma

Leiomyoma
Take home messages

• Certain abdominal pathologies can be detected by a routine screening chest X-ray.

• Current modalities to assess the submucosal tumors of stomach are combination of CT scan and endoscopic ultrasonography (EUS).

• Diagnostic approach of gastric submucosal tumors is to differentiate GISTs from the others which are usually benign but GISTs are by far the top candidate for malignancy.


References

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