30-year-old Female with Nausea, Vomiting, and Abdominal Pain

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Patient Presentation

• HPI: 30 y.o. F presenting with intermittent nausea, vomiting, and diffuse abdominal pain for the past week. She has also been experiencing paroxysmal chills and cough with SOB for the past few days.

• PMH: asthma

• PSH: none

• FH: stroke in uncle

• SH: no recent travel

• Allergies: no known allergies

• PE: 100.1 F, HR 117, BP 134/72 General-no acute distress; HEENT- mucous membranes moist; Neck-supple; Respiratory- lungs clear to auscultation bilaterally; Cardiovascular- tachycardic with no murmurs, rubs, or gallops; Abdominal - soft with diffuse tenderness to palpation; Skin- no rashes or skin lesions; Musculoskeletal- no tenderness or swelling; Neurologic- sensation to touch intact and CNII-XII intact

• Pertinent Labs: WBC wnl, Covid +, Mono-, Echinococcus +
### Variant 1:

**Acute nonlocalized abdominal pain and fever. No recent surgery. Initial imaging.**

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Appropriateness Category</th>
<th>Relative Radiation Level</th>
</tr>
</thead>
<tbody>
<tr>
<td>CT abdomen and pelvis with IV contrast</td>
<td>Usually Appropriate</td>
<td>☒ ☒ ☒ ☒</td>
</tr>
<tr>
<td>MRI abdomen and pelvis without and with IV contrast</td>
<td>May Be Appropriate</td>
<td>☐</td>
</tr>
<tr>
<td>US abdomen</td>
<td>May Be Appropriate</td>
<td>☐</td>
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<td>May Be Appropriate</td>
<td>☒ ☒ ☒ ☒ ☒</td>
</tr>
<tr>
<td>Radiography abdomen</td>
<td>May Be Appropriate</td>
<td>☒ ☒ ☒ ☒</td>
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<tr>
<td>FDG-PET/CT skull base to mid-thigh</td>
<td>Usually Not Appropriate</td>
<td>☒ ☒ ☒ ☒ ☒ ☒</td>
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<tr>
<td>WBC scan abdomen and pelvis</td>
<td>Usually Not Appropriate</td>
<td>☒ ☒ ☒ ☒ ☒ ☒</td>
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<tr>
<td>Nuclear medicine scan gallbladder</td>
<td>Usually Not Appropriate</td>
<td>☒ ☒ ☒ ☒ ☒ ☒</td>
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<tr>
<td>Fluoroscopy contrast enema</td>
<td>Usually Not Appropriate</td>
<td>☒ ☒ ☒ ☒ ☒ ☒</td>
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<tr>
<td>Fluoroscopy upper GI series with small bowel</td>
<td>Usually Not Appropriate</td>
<td>☒ ☒ ☒ ☒ ☒ ☒</td>
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</table>

These imaging modalities were ordered by the provider.
Radiography Abdomen (Findings Unlabeled)
KUB demonstrates enlarged spleen
CT w/Contrast (Findings Unlabeled)
Markedly enlarged spleen with a multiloculated solid and cystic mass spanning the entirety of the spleen and demonstrating scattered punctuate calcifications.
US Abdomen (Findings Unlabeled)
Spleen is heterogenous and enlarged, measuring 13.9 x 14.0 x 9.9 cm, with volume of 1005mL. There are complex, mostly solid, partially cystic splenic masses with the largest discrete lesion delineated (+) and measuring 5.6 x 5.1 x 4.9 cm.
Final Diagnosis:

Littoral Cell Angioma of the Spleen
Case Discussion

• Littoral cell angioma of the spleen is a rare, benign primary vascular tumor arising from the littoral cells lining the splenic red pulp sinuses

• Epidemiology
  • Rare with few cases documented in literature
  • Occurs in all ages but most cases are reported in middle-aged adults
  • Not associated with gender

• Clinical Findings
  • Often found incidentally but may present with abdominal pain or splenomegaly on physical exam
  • Laboratory results may show signs of hypersplenism such as anemia or thrombocytopenia
Case Discussion

• Imaging Differential Diagnosis
  • metastases
  • lymphoma
  • sarcoidosis
  • abscesses
  • other primary splenic vascular tumors
    • Benign - hemangioma, hamartoma, hemangioendothelioma
    • Malignant - angiosarcoma

• Imaging Findings
  • CT: hypoattenuating masses with progressive homogeneous enhancement, later than normal splenic parenchyma
  • MR: T1 and T2 – hypointense masses (due to hemosiderin)
  • US: heterogeneous echotexture; variable vascularity
## Case Discussion

### Comparative Imaging Findings:

<table>
<thead>
<tr>
<th>Condition</th>
<th>Littoral Cell Angioma</th>
<th>Splenic Hemangioma</th>
<th>Angiosarcoma</th>
<th>Lymphoma</th>
<th>Abscess</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>CT</strong></td>
<td>Hypoattenuating masses</td>
<td>Hypoattenuating masses</td>
<td>Multiple nodular masses of heterogenous hypoattenuation</td>
<td>Iso attenuating masses</td>
<td>Centrally hypoattenuating lesions</td>
</tr>
<tr>
<td></td>
<td>Progressive homogenous contrast enhancement</td>
<td>Centripetal enhancement</td>
<td>Heterogenous enhancement</td>
<td>Hypoenhancement (best appreciated in late venous phase)</td>
<td>Peripheral enhancement</td>
</tr>
<tr>
<td><strong>MRI</strong></td>
<td>T1 and T2- Hypointense</td>
<td>T1- Iso to hypointense to splenic parenchyma</td>
<td>MRI</td>
<td>MRI</td>
<td>MRI</td>
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<td>T2- Hyperintense to splenic parenchyma</td>
<td>T2- Hyperintense to splenic parenchyma</td>
<td>T1 and T2- Nodular, hypointense to splenic parenchyma</td>
<td>T1- Hypointense (increased signal with proteinaceous content)</td>
<td>T2-Hyperintense</td>
</tr>
<tr>
<td><strong>US</strong></td>
<td>Heterogeneous echotexture</td>
<td>Homogeneous echotexture</td>
<td>US</td>
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<td></td>
<td>Predominantly hyperechoic</td>
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<td>Heterogeneous echotexture (cystic and solid components)</td>
<td>Homogeneous echotexture</td>
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<td></td>
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<td>Increased vascularity on doppler</td>
<td>Hypoechoic</td>
<td>Ranges from predominantly hypoechoic with some internal echoes to hyperechoic</td>
</tr>
</tbody>
</table>

**Notes:**
- CT: Computed Tomography
- MRI: Magnetic Resonance Imaging
- US: Ultrasonography
Case Discussion

• Diagnosis and Treatment:
  • Splenectomy performed for:
    • symptom relief
    • histological diagnosis to differentiate benign, littoral cell angioma from malignant, littoral cell angiosarcoma and from littoral cell hemangioendothelioma
  • Pathology: multiple masses composed of numerous anastomosing vascular channels filled with blood and lined with tall, hemophagocytic, endothelial cells
  • Immunophenotype: Vascular endothelial markers (CD31, CD34, and factor VIII) and Histiocytic markers (CD68)
  • Littoral cell angioma has been associated with extra-splenic, visceral organ malignancies and Crohn’s Disease, so patients should be assessed accordingly
  • Massive splenomegaly (weight >/= 1500gm) has been associated with malignancy requiring post-splenectomy patients to undergo surveillance for recurrence

• Our case: Patient experienced gastric outlet obstruction requiring splenectomy. Pathology of the lesion showed co-expression of CD31, CD68, and CD163
References

- Littoral cell angioma of the spleen | Radiology Reference Article | Radiopaedia.org
- Venkata, Krishna MD; Patel, Ronak; Sarmad, Rehan; Hasanin, Mohsen; Thani, Kannan Littoral Cell Angioma of the Spleen, American Journal of Gastroenterology: October 2015 - Volume 110 - Issue - p S114