AMSER Case of the Month
October 2021

60-year-old male with dysphagia, abdominal pain, and weight loss

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Patient Presentation

• HPI: Several months of inability to keep solid foods down, low-grade fever, and night sweats. Recent worsening dysphagia with mild diffuse abdominal pain and 10 lb weight loss.

• ROS: Fatigue, nausea, regurgitation after eating

• PMH: Crohn’s disease, Hodgkin lymphoma s/p chemotherapy (1981), bladder cancer s/p resection (2018), GERD and Barrett’s esophagus s/p fundoplication (EGD 11/2020 showed enteritis)
Patient Presentation

• **SH**: non-smoker, no alcohol use

• **PE**: vitals normal, thin and ill-appearing, no abdominal tenderness or masses

• **Labs**: Hb 12.1, lipase normal, ALT 65, HCV Ab negative, HIV Ag/Ab Combo negative, LDH 1203
What Imaging Should We Order?
Select the applicable ACR Appropriateness Criteria

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Appropriateness Category</th>
<th>Relative Radiation Level</th>
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</thead>
<tbody>
<tr>
<td>Fluoroscopy upper GI series</td>
<td>Usually Appropriate</td>
<td>3</td>
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<tr>
<td>CT abdomen and pelvis with IV contrast</td>
<td>Usually Appropriate</td>
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<tr>
<td>CT abdomen and pelvis without IV contrast</td>
<td>May Be Appropriate</td>
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<tr>
<td>CT abdomen with IV contrast</td>
<td>May Be Appropriate (Disagreement)</td>
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<tr>
<td>CT abdomen without IV contrast</td>
<td>May Be Appropriate</td>
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<td>CT abdomen with IV contrast multiphase</td>
<td>May Be Appropriate</td>
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<tr>
<td>Fluoroscopy biphasic esophagram</td>
<td>Usually Not Appropriate</td>
<td>3</td>
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<tr>
<td>Fluoroscopy single contrast esophagram</td>
<td>Usually Not Appropriate</td>
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<tr>
<td>MRI abdomen without and with IV contrast</td>
<td>Usually Not Appropriate</td>
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<td>MRI abdomen without and with IV contrast with MRCP</td>
<td>Usually Not Appropriate</td>
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<tr>
<td>MRI abdomen without IV contrast</td>
<td>Usually Not Appropriate</td>
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<td>Usually Not Appropriate</td>
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<tr>
<td>FDG-PET/CT skull base to mid-thigh</td>
<td>Usually Not Appropriate</td>
<td>3</td>
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Given abdominal pain, dysphagia, constitutional symptoms, PMH of cancer – concern for gastric or esophageal malignancy versus other abdominal pathologies

May also consider fluoroscopy or endoscopy for dysphagia
Findings
Findings

Lymphadenopathy around the GE junction, measuring up to 3.3 cm along the gastrohepatic ligament.

Multiple bilobar hepatic lesions measuring up to 5 cm.

Severe thickening of the distal to mid esophagus with fluid and debris.
Endoscopy

Circumferential mass (biopsy taken)

Lower Third of the Esophagus: Mass, Food

Gastroesophageal Junction: Barrett's esophagus

(separate from the mass)
Ultrasound-Guided Liver Biopsy
Final Dx:

Esophageal Sarcoma with metastasis to the liver
Outcome

• Esophageal pathology showed SMARCA4-deficient undifferentiated sarcoma. Liver pathology was the same, indicating metastasis.

• The patient underwent jejunostomy tube placement for feedings, and began chemotherapy with doxorubicin, ifosfamide, and mesna. No further surgery or radiation therapy was planned.
Case Discussion

• Esophageal cancers – squamous cell carcinoma and adenocarcinoma most common; several rarer types (<1% are sarcomas)

• General risk factors include: age, male, tobacco/alcohol use, GERD, Barrett’s, personal cancer history

• AJCC TMN staging for all types

• Take into account grading: well differentiated (G1), moderately differentiated (G2), poorly differentiated (G3), undifferentiated (G4)

• Fluoro/endoscopy aids diagnosis, plus CT or PET for staging
Case Discussion

- Soft tissue sarcomas – aggressive with high recurrence rate
- Can be primary or a secondary malignancy as a result of past cancer treatment with radiation or chemotherapy, as in this patient
- LDH may be elevated in lymphoproliferative disorders, such as Hodgkin’s, as well as sarcomas
- Large tumor size (>5.6cm), LDH>240, Hb<12.4 associated with malignancy of soft tissue tumors
Case Discussion

• SMARCA4 – gene involved in chromatin remodeling
• Variety of rare SMARCA4-deficient neoplasms including thoracic sarcomas – usually in the lungs and often associated with smoking history
• For all esophageal cancers: resection recommended if no invasion/metastasis. However esophagectomy has significant morbidity/mortality; consider adjuvant radiation therapy, chemotherapy alone
• Metastatic disease, such as in this patient, warrants chemotherapy and supportive measures
References


