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

Variant 2: Epigastric pain with clinical suspicion for gastric cancer. Initial imaging.		
Procedure	Appropriateness Category	Relative Radiation Level
Fluoroscopy upper GI series	Usually Appropriate	ଜନନ
CT abdomen and pelvis with IV contrast	Usually Appropriate	ଡଡଡ
CT abdomen and pelvis without IV contrast	May Be Appropriate	ଚଚଚ
CT abdomen with IV contrast	May Be Appropriate (Disagreement)	ଚଚଚ
CT abdomen without IV contrast	May Be Appropriate	ଚଚଚ
CT abdomen with IV contrast multiphase	May Be Appropriate	****
Fluoroscopy biphasic esophagram	Usually Not Appropriate	***
Fluoroscopy single contrast esophagram	Usually Not Appropriate	ଚଚଚ
MRI abdomen without and with IV contrast	Usually Not Appropriate	0
MRI abdomen without and with IV contrast with MRCP	Usually Not Appropriate	0
MRI abdomen without IV contrast	Usually Not Appropriate	0
MRI abdomen without IV contrast with MRCP	Usually Not Appropriate	0
CT abdomen and pelvis without and with IV contrast	Usually Not Appropriate	ଉ ତ୍ତର
CT abdomen without and with IV contrast	Usually Not Appropriate	****
FDG-PET/CT skull base to mid-thigh	Usually Not Appropriate	****

This imaging modality was ordered by the physician

- 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

RMSER

Findings





Findings

Multiple bilobar hepatic lesions measuring up to 5 cm Lymphadenopathy around the GE junction, measuring up to 3.3 cm along the gastrohepatic ligament

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



Endoscopy



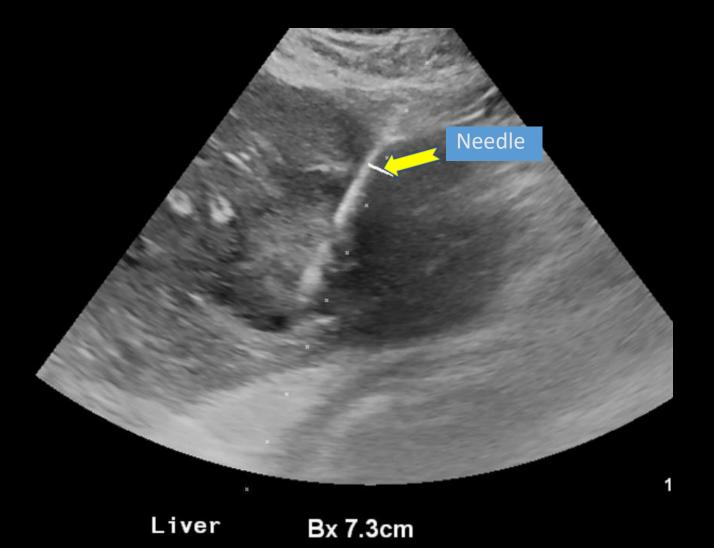
Circumferential mass (biopsy taken)



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

- Berry MF. Esophageal cancer: staging system and guidelines for staging and treatment. J Thorac Dis. 2014;6 Suppl 3(Suppl 3):S289-S297. doi:10.3978/j.issn.2072-1439.2014.03.11
- Brower V. Tracking Chemotherapy's Effects on Secondary Cancers, *JNCI: Journal of the National Cancer Institute*, Volume 105, Issue 19, 2 October 2013, Pages 1421–1422. https://doi.org/10.1093/jnci/djt273
- Fujibuchi T, Miyawaki J, Kidani T, Imai H, Miura H. Prediction of Soft Tissue Sarcoma from Clinical Characteristics and Laboratory Data. *Cancers (Basel)*. 2020;12(3):679. Published 2020 Mar 13. doi:10.3390/cancers12030679
- Hird AE, Magee DE, Matta R, et al. Assessment of Secondary Sarcomas Among Patients With Cancer of the Abdomen or Pelvis Who Received Combinations of Surgery, Radiation, and Chemotherapy vs Surgery Alone. *JAMA Netw Open.* 2020;3(10):e2013929. doi:10.1001/jamanetworkopen.2020.13929
- Le Loarer, F., Watson, S., Pierron, G. *et al. SMARCA4* inactivation defines a group of undifferentiated thoracic malignancies transcriptionally related to BAF-deficient sarcomas. *Nat Genet* 47, 1200–1205 (2015). https://doi.org/10.1038/ng.3399
- Sauter, J., Graham, R., Larsen, B. *et al. SMARCA4*-deficient thoracic sarcoma: a distinctive clinicopathological entity with undifferentiated rhabdoid morphology and aggressive behavior. *Mod Pathol* 30, 1422–1432 (2017). https://doi.org/10.1038/modpathol.2017.61