

Anthony P. Nguyen MD, Fray Arroyo MD, Marjan Hovaida MD, Christopher Chang MD, PhD

Department of Gastroenterology & Hepatology, University of New Mexico Health Sciences Center, Albuquerque, New Mexico

Department of Gastroenterology & Hepatology, Raymond G. Murphy VA Medical Center, Albuquerque, New Mexico

Introduction | **Figures & Tables** | **Discussion**

- Primary/Idiopathic achalasia describes a condition that leads to impaired esophageal motility related to dysfunction of the myenteric plexus.
- Pseudoachalasia accounts for ~4-5% of cases of achalasia and causes similar clinical symptoms as primary achalasia but is due to a secondary underlying cause such as malignancy.
- It is often difficult to differentiate primary achalasia from pseudoachalasia due to the clinical and diagnostic similarities between the 2 conditions.

Case Presentation

HPI: A 76 year-old man with a history of SCC presented with dysphagia to solid foods and significant weight loss.

Diagnostic/Treatment Course:

- Presented to PCP with 2-3 weeks of dysphagia with significant (12-13kg) weight loss
- Referred to GI clinic and EGD with biopsy was performed with normal mucosa and no evidence of eosinophilic esophagitis.
- CT Chest notable for ill-defined 2.4x1.5 cm mediastinal mass along the posterior aspect of the mid-esophagus w/ circumferential mural esophageal thickening
- MRI notable for 3cm mediastinal mass behind the esophagus concerning for malignancy w/ local lymphadenopathy
- Patient then started having increasing nocturia and intermittent difficulty urinating. PSA elevated at 34.8 w/ enlarged prostate on DRE.
- Prostate biopsy was positive for prostatic adenocarcinoma.
- Transferred to UNM hospital for EUS which showed hypoechoic granular mass w/ FNB pathology returning positive for prostatic adenocarcinoma
- Patient was referred to medical oncology and initiated chemotherapy and radiation.

Table 1. Clinical and Diagnostic Features in Primary Achalasia vs Pseudoachalasia

	Primary Achalasia	Pseudoachalasia
Clinical Presentation	- Variable age - Longer duration of symptoms - Less significant weight loss	- Advanced Age - Short duration of symptoms - Significant weight loss
EGD	- Normal Exam	- Difficulty traversing EGJ
Barium Esophagram	Generally not useful for distinguishing the 2 conditions.	
Manometry	- High LES pressure - Absent lower esophageal peristalsis - Failure of LES relaxation after deglutition	- Low Integration Relaxation Pressure (IRP) - Compartmentalized pressure at the distal esophagus

- Primary and Pseudo-achalasia are often clinically homogenous conditions that are difficult to diagnose based on initial diagnostic studies such as EGD and barium esophagram alone.
- Prior studies have shown that advanced age (>55), significant weight loss (>10kg), shorter duration of symptoms (6 months) and difficulty passing the scope through the EGJ during initial exam were all risk factors for malignant pseudoachalasia with the highest predictive value occurring when >2 risk factors are present
- High-resolution manometry results showing lower integration relaxation pressure (IRP) and compartmentalized pressure in the distal esophagus in the context of the risk factors above can be suggestive of pseudoachalasia.
- EUS can be a useful when infiltrative malignancy is suspected as the cause for esophageal symptoms and initial EGD, HRM and barium esophagram are equivocal.

Learning Points

- Risk factors that suggest pseudoachalasia over primary achalasia include: advanced age, short duration of symptoms, significant weight loss and difficulty passing through the EGJ junction during EGD.
- EUS can be a useful diagnostic tool to rule out infiltrative tumors when EGD and manometry are equivocal.
- Timely diagnosis of pseudoachalasia is important as the treatment plan is completely different from patients with a primary/idiopathic achalasia.

References

Haj Ali, S. N., Nguyen, N. Q., & Abu Sneineh, A. T. (2021). Pseudoachalasia: a diagnostic challenge. When to consider and how to manage? *Scandinavian Journal of Gastroenterology*, 56(7), 747–752. <https://doi.org/10.1080/00365521.2021.1925957>

M, G., MD, M., A, R., & CP, G. (2021). Duration of symptoms and manometric parameters offer clues to diagnosis of pseudoachalasia. *Neurogastroenterology and Motility: The Official Journal of the European Gastrointestinal Motility Society*, 33(1). <https://doi.org/10.1111/NMO.13965>

I, G., VF, E., T, S., & T, J. (2005). Pseudoachalasia: a case series and analysis of the literature. *Scandinavian Journal of Gastroenterology*, 40(4), 378–385. <https://doi.org/10.1080/00365520510012118>

FA, P., MI, van R., SMM, M., AJPM, S., & AJ, B. (2017). Diagnostic features of malignancy-associated pseudoachalasia. *Alimentary Pharmacology & Therapeutics*, 45(11), 1449–1458. <https://doi.org/10.1111/APT.14057>

E, F., AJ, E., C, L., C, U., FM, S.-J., & GJ, K. (2019). Pseudoachalasia as First Manifestation of a Malignancy. *Digestive Diseases (Basel, Switzerland)*, 37(5), 347–354. <https://doi.org/10.1159/000495758>



Figure 1. EUS notable for granular, hypoechoic mass in the mid-esophagus

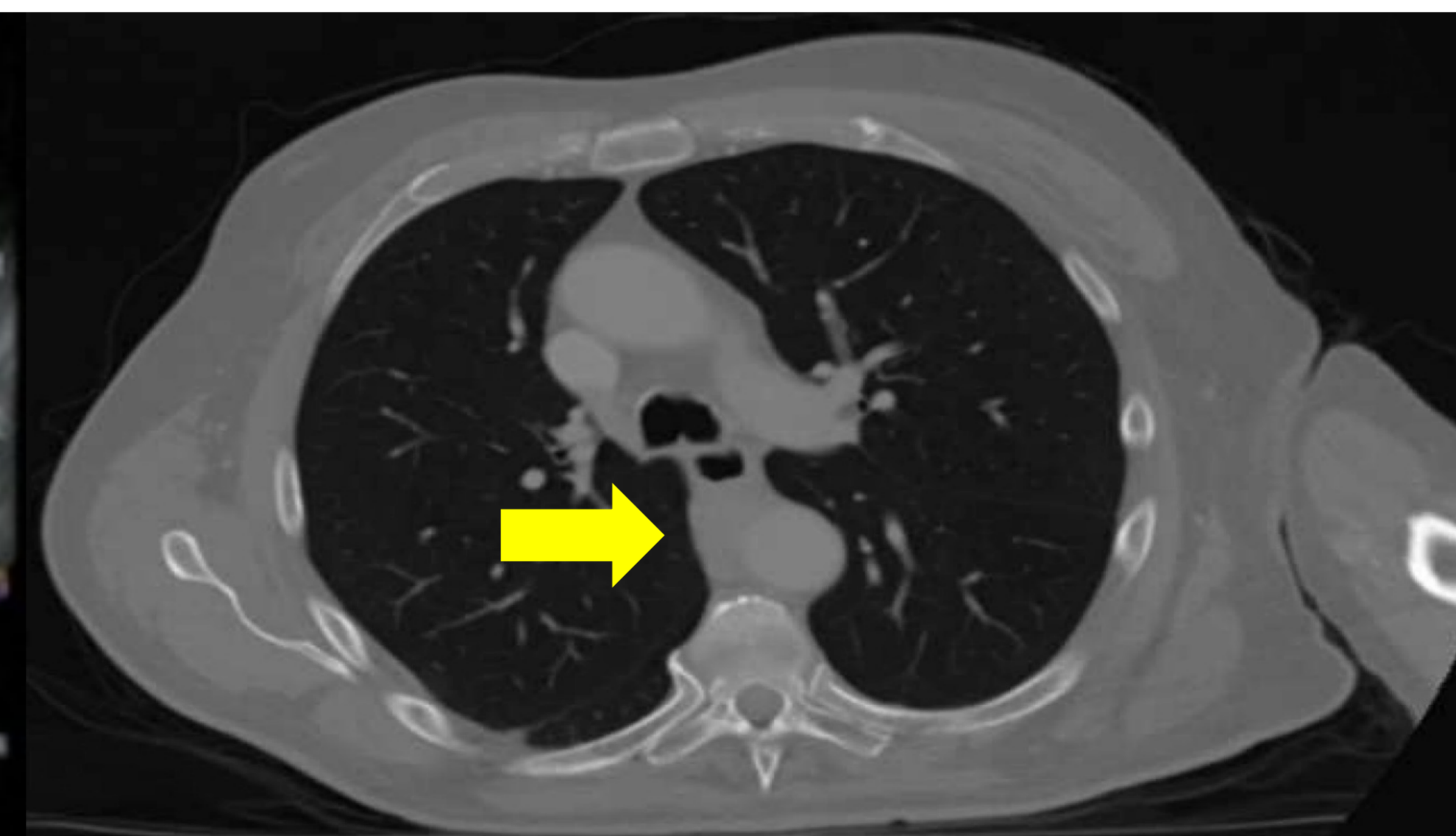


Figure 2. CT Chest showing circumferential mass posterior to the esophagus.

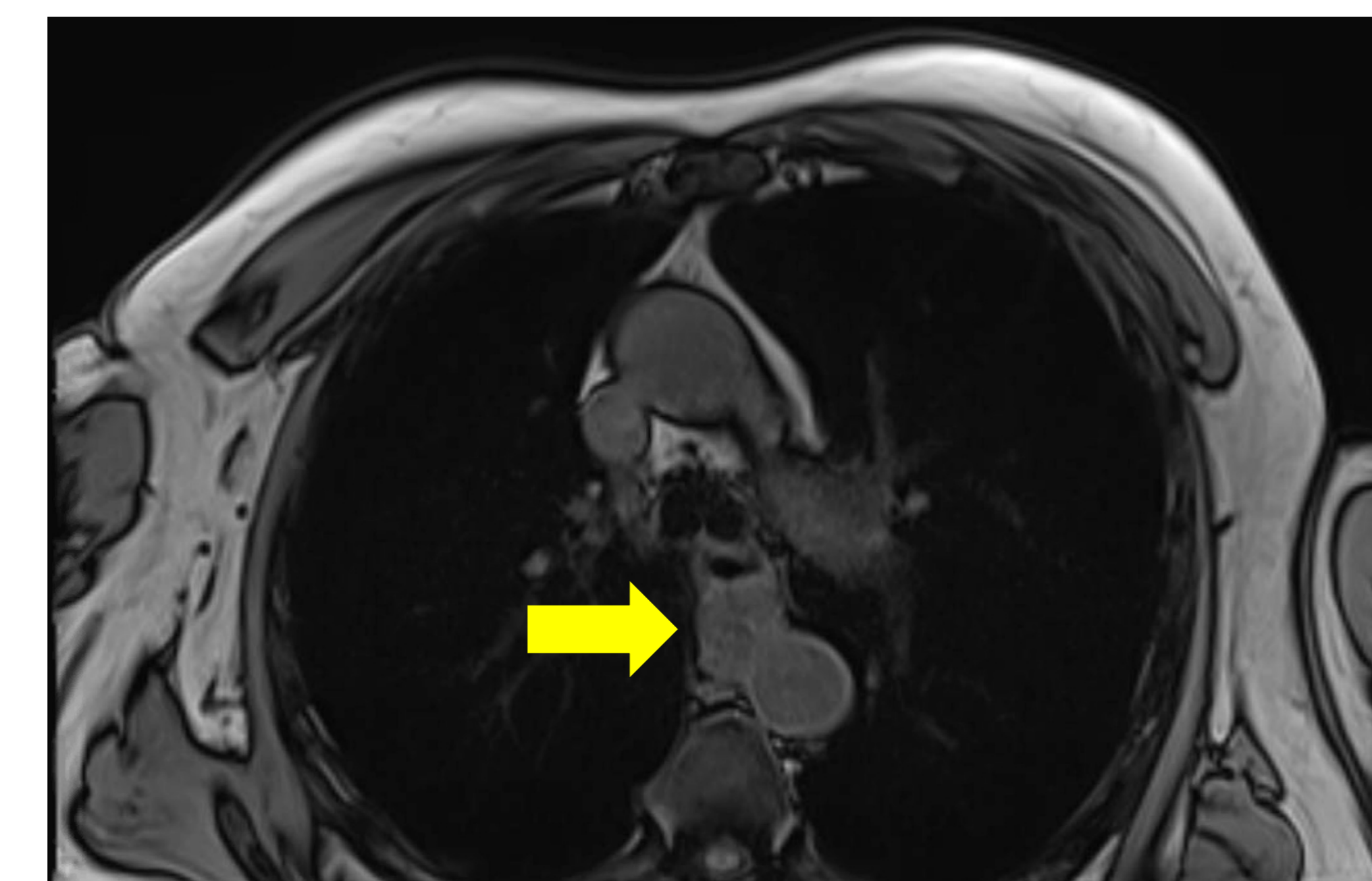


Figure 3. MRI notable for circumferential mass posterior to the esophagus

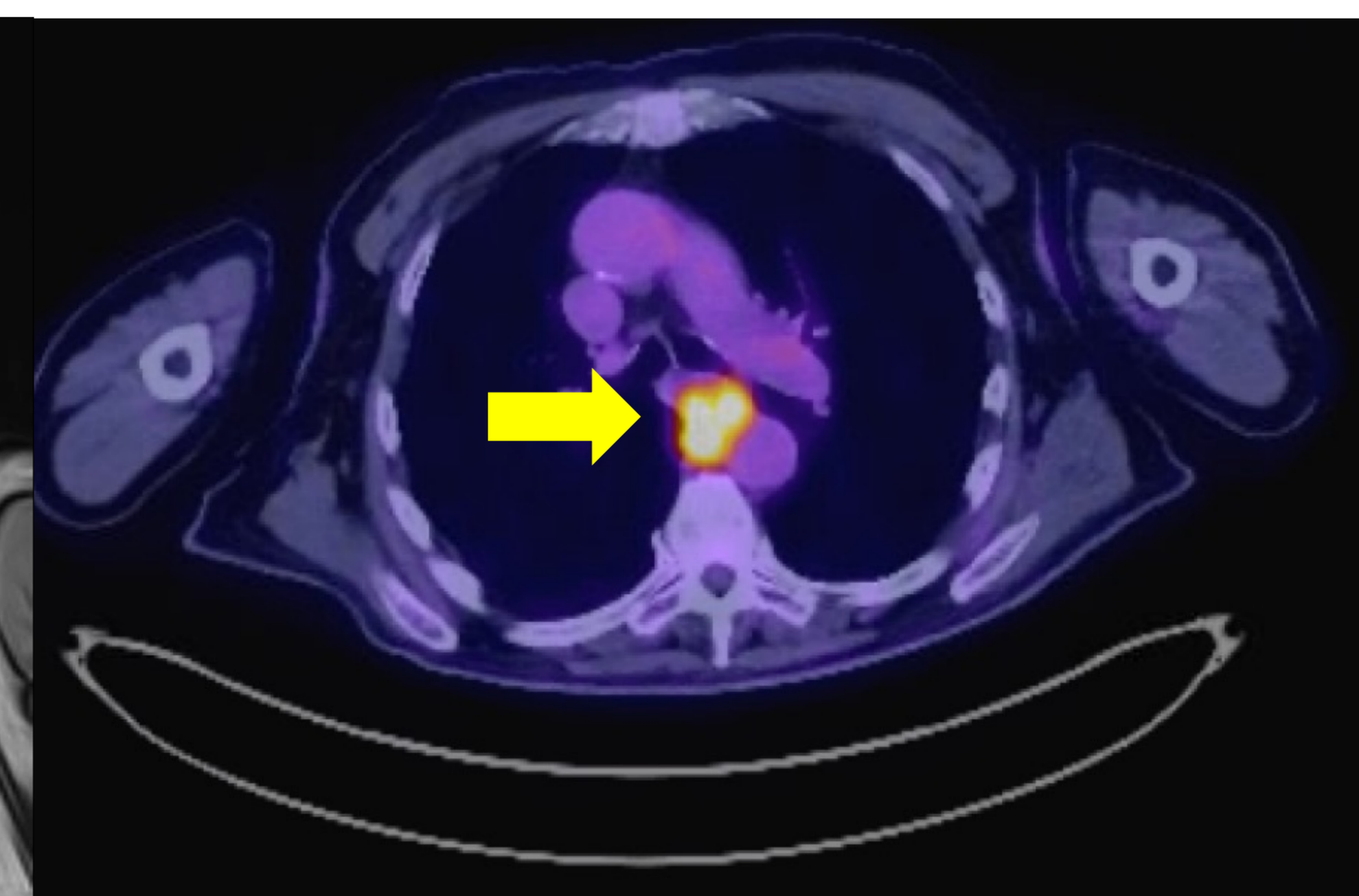


Figure 4. PET CT scan notable for avidity in the posterior peri-esophageal mass.