Rare Case of 27 Year Old with Esophageal Cancer

Saakshi Joshi M.D., Kallie Mae Jimmerson D.O., Alexandra Davies D.O., Jonathan Markus M.D.

McLaren Macomb Medical Center, Mount Clemens, MI
Michigan State University School of Medicine

Introduction

Esophageal cancer is one of the most common malignancies globally and the sixth leading cause of cancer mortality; associated with a high mortality, more specifically a 5 year survival rate of 25%.[1] Two main histological types are adenocarcinoma and squamous cell carcinoma (SCC), there is a recent increase in incidence of adenocarcinoma with a decrease in squamous cell carcinoma.[1] Neuroendocrine tumors of the esophagus account for 0.03 to 0.05% of all esophageal cancers and majority of research is based on the SCC and adenocarcinoma.[8] Both are more common in men and caucasians.[2] Strauss et al analyzed the trend in age of esophageal cancer diagnosis from 1991 to 2018 as seen in Figure 1 emphasizing the importance of reporting such cases.

Years	Total number of patients	Number (%) of patients ≥50	Number (%) of patients <50	Number (%) of patients <40	Number (%) of patients <30	Mean age	Standard deviation
1991–1995	20	18 (90%)	2 (10%)	0 (0%)	0 (0%)	58.9	12.7
1996–2000	57	47 (82.4%)	10 (17.5%)	4 (7.0%)	0 (0%)	60.3	11.8
2001–2006	116	104 (89.7%)	12 (10.3%)	4 (3.4%)	0 (0%)	63.6	10.6
2007–2011	140	123 (87.8%)	17 (12.1%)	3 (2.1%)	2 (1.4%)	62.4	10.3
2012–2018	291	267 (91.7%)	24 (8.24%)	7 (2.4%)	4 (1.3%)	64.2	11.2

Figure 1: From 1991 to 2018, increased incidence in patients <30 year old with esophageal cancer from 0% to 1.3%[3]

Risk factors include obesity, human papillomavirus, smoking, gastroesophageal reflux disease, hot beverages, alcohol abuse, and achalasia. Common symptoms include dysphagia and weight loss. Asymptomatic patients may have anemia, mediastinal lymphadenopathy, or hoarseness of voice due to the encasement of the recurrent laryngeal nerve.[4]

Case Description

27-year-old female with history of obesity and depression, presented to the outpatient clinic for acid reflux. Additionally, patient had globus sensation and stated symptoms are worse after eating. Patient started on famotidine 20mg daily with no improvement, patient developed dysphagia to solids daily. Esophagogastroduodenoscopy (EGD) demonstrated 4cm mass at distal esophagus bridging the gastroesophageal junction with a 2cm cratered ulceration at the distal end of the mass as seen in Figure 2, 3 and 4. Computed tomography of abdomen showed abdominal adenopathy and 11cm metastatic lesion at liver as seen in Figure 5. Pathology showed advanced neuroendocrine carcinoma of the esophagus with mets to the liver. Patient started on carboplatin and VP-16. Currently, patient continuing to receive chemotherapy, and will receive PET scan within the next month.

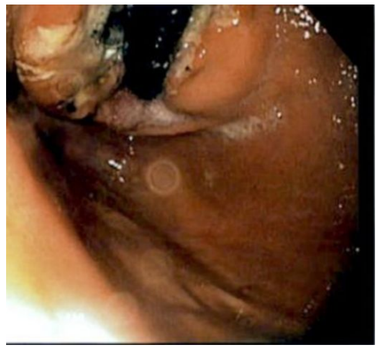

Figure 2

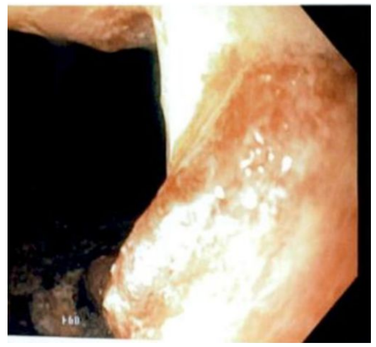

Figure 3

Figure 2 and 3: 2cm cratered ulceration at distal end of mass

Figure 4: 4cm mass

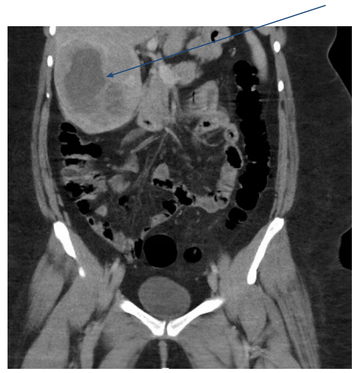

Figure 5: 11cm liver lesion

Discussion

This case addresses multiple topics amongst the latest gastroenterology research, including prognosis of patients with esophageal cancer, endoscopic resection of tumors and the use of neoadjuvant chemoradiation.

In regards to contraindicating studies regarding prognosis, due to the rarity of neuroendocrine tumors of the esophagus; there are limited studies to assess optimal therapy and prognosis of such patients.[8] Although studies to exist comparing adenocarcinoma and SCC, recent studies indicate increased survival rates amongst patients with adenocarcinoma with a difference of 10.6% in 1 year survival and 4.5% in 5 year survival. A higher risk of mortality associated with grade 3, stage 4, lack of surgery and chemotherapy. Contrary to prior literature stating larger tumor size indicates higher risk of mortality, these studies were found to have low statistical significance and in recent studies; no correlation is found between size of tumors and mortality.[5]

The therapeutic approach to esophageal cancers is dependent on a multitude of factors. These factors include the preoperative staging, location, size, histology and tissue of origin. Treatment options include endoscopic resection, surgical resection for lesions invading the submucosa, chemo or radiation therapy of resectable lesions invading the muscularis propria and palliative systemic therapy for locally advanced, unresectable and metastatic disease.[4]

A recent meta analysis compared endoscopic mucosal resection (EMR) and endoscopic submucosal dissection (ESD). Study demonstrated increased en bloc resection, lower recurrence and more marginally negative resection rates in ESD. Although, increased perforations and more time spent with ESD. Furthermore, these benefits of ESD occurred more often with lesions more than 20mm in size, therefore, recommending EMR appropriate for <10mm, EMR and ESD appropriate for 11 to 20mm and ESD preferred for >20mm.[6]

For adenocarcinoma and SCC options for nonsurgical therapy include chemotherapy, radiation, neoadjuvant therapy and more recent research is suggesting immunotherapy. Study comparing neoadjuvant chemotherapy and surgery alone stated an increase in R0 resection rate of 84% and 74% respectively. There are ongoing studies comparing neoadjuvant chemotherapy with chemoradiotherapy showing controversial evidence and more research is needed in this area.

Conclusion

As esophageal neuroendocrine tumor is extremely rare with limited research and most clinical trials are based on adenocarcinoma and SCC, more cases need to be reported of such pathology.[8] Amongst controversy between ESD and EMR and the various types of nonsurgical treatments. We recommend further analysis and studies regarding treatment of esophageal cancers including neuroendocrine tumors.

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