Take my breath away: A rare cause of dysphagia

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Introduction

- Esophageal adenoid cystic carcinoma (EACC) is an extremely rare malignancy accounting for only 0.1% of esophageal malignancies.
- It is most common in the parotid and salivary glands and metastatic regional lymph nodes are uncommon, hence dysphagia is rarely associated with EACC. EACC carries a very poor prognosis with distant metastases.
- Here we report a case of EACC that debuted with dysphagia.

Case Description

An 81-year-old man with a history of smoking, chronic obstructive pulmonary disease, presented with four years of intermittent dysphagia to solids that improved with drinking water. Associated symptoms are cough and dysphonia. He denied weight loss, chest pain, vomiting, regurgitation of food contents, odynophagia and abdominal pain. Physical examination and laboratories were unremarkable. Barium swallow showed a corkscrew appearance suggestive of esophageal spasm. GI service was consulted and recommended an EGD for direct visualization as symptoms were not consistent with esophageal spams (figure 1). A chest and neck CT was ordered due to persistent cough and dysphonia, revealed a lesion in the subcarinal compartment measuring approximately 2.7 × 2.1 cm proximal to the left mainstem bronchus, inseparable from the esophagus (figure 2). Endobronchial ultrasound (EBUS) with fine-needle aspiration (FNA): reported an adenoid cystic carcinoma. PET-CT shows abnormal metabolic activity of posterior mediastinum retrocarinal space, with no lymphadenopathy or evidence of distant metastases.

Images

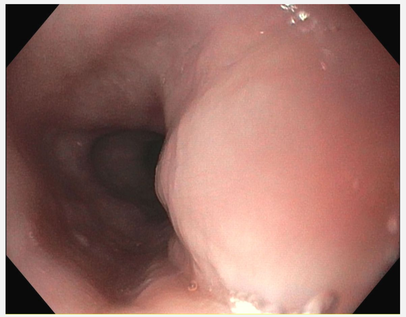

Figure 1 : Normal Esophageal epithelium. Fixed Moderated extramural compression against the esophageal wall at 25-27cm from incisors.

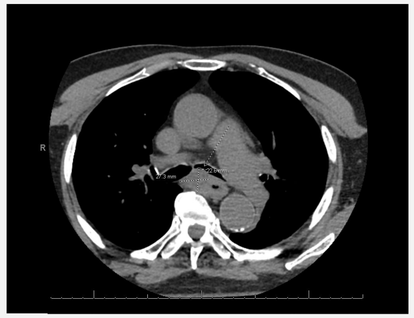

Figure 2: Lesion in the subcarinal compartment proximal to the left mainstem bronchus, inseparable from the esophagus.

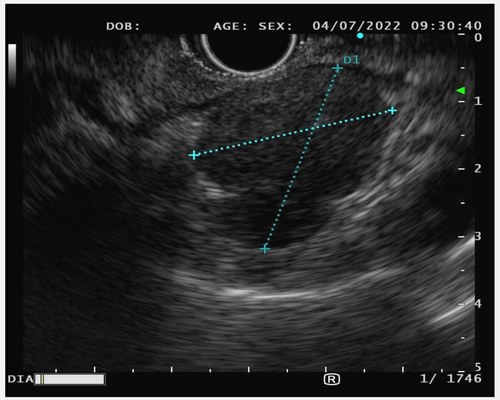

Figure 3: Mediastinum one 2.8cm x 2.7cm hypoechoic mass in the posterior mediastinum with focal areas of heterogeneity abutting the esophagus.

Case Description

Endoscopic ultrasound (EUS) with FNA confirmed the presence of EACC originated from the submucosa (figure 3). Pathology showed cohesive clusters and aggregates of atypical epithelial cells in a tubular and cribriform pattern.

Conclusion

- The most common endoscopic finding of EACC is a polypoid mass.
- Histology shows three growth patterns: cribriform, tubular, or solid, the latter being associated with a worse prognosis.
- Treatment of choice for EACC is radical excision.
- The reported 5-year survival rate of EACC is around 35%.
- This highlights the importance of expedited evaluation in cases of dysphagia and the importance of being aware of rare causes such as EACC.

References

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