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Visual Diagnosis Pearl

Tracheal Adenoid Cystic Carcinoma

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CASE PRESENTATION

A 36-year-old woman with no significant past medical history presented to an outside hospital with progressive shortness of breath. She reported respiratory symptoms over the past three years that were previously attributed to asthma. Despite treatment, her symptoms gradually worsened, with increasing dyspnea on exertion and minimal sustained response to inhaler therapy. She denied tobacco use, occupational exposures, or systemic symptoms. Prior chest x-rays were negative. Given the progressive nature of her symptoms and suboptimal response to typical asthma management, further evaluation was pursued.

On this evaluation, she underwent a CT (computed tomography) scan which showed a large tracheal mass, prompting transfer to a tertiary care center for a higher level of care. A CT chest without contrast on admission demonstrated a 5.41 cm length segment of the trachea with diffuse nodular wall thickening (Figure 1).

The patient underwent rigid bronchoscopy on ECMO (extracorporeal membrane oxygenation) with biopsy and debulking of the tracheal mass. The extent and longitudinal distribution of airway involvement raised concern for airway instability during instrumentation, necessitating extracorporeal support to ensure procedural safety. Pathology showed adenoid cystic carcinoma. There was no evidence of metastatic disease on imaging. The tumor was determined to be unresectable due to extensive longitudinal involvement of the trachea and the inability to achieve safe surgical margins.

She received chemotherapy with carboplatin and paclitaxel as well as radiation therapy, which was completed three months after the initial diagnosis. Subsequent imaging showed interval improvement of the tracheal mass with no evidence of metastatic disease, with associated improvement in respiratory symptoms and airway patency.

DISCUSSION

Adenoid cystic carcinoma (ACC) most commonly arises in the salivary glands but can occur at other sites, including the trachea. Among primary malignant tracheal tumors, squamous cell carcinoma (SCC) is the predominant histologic subtype, followed by ACC.[1] Tracheal ACC is rare, with an estimated incidence of 0.04–0.2%.[2] It is a slow-growing tumor, with a slight female predominance and a peak incidence in the fourth decade of life.[1] Tracheal ACC is not associated with smoking or alcohol use.[1] Given its indolent nature and ability to mimic other respiratory diseases, diagnosis can be delayed until significant airway obstruction develops.

As illustrated in this case, tracheal ACC may present with nonspecific respiratory symptoms that closely resemble asthma, leading to prolonged diagnostic delay. Progressive symptoms, poor response to bronchodilator therapy, or dyspnea out of proportion to examination findings should prompt consideration of structural airway pathology and further imaging.

Imaging and bronchoscopy are important to assess resectability, and endoscopic intervention may serve as a bridge to definitive treatment by restoring airway patency. Endoscopic treatments include mechanical tumor debulking, thermal ablation, and cryotherapy, among others.[3] In cases of critical or near-critical airway obstruction, these procedures carry significant risk and may require advanced airway planning, including extracorporeal support. A 2025 systematic review including more than 1000 patients reported that surgical resection, with or without post-operative radiation therapy, was the most common treatment approach.[1] Radiation is recommended as adjuvant therapy after certain types of resections, as well as for unresectable tumors or palliation. Given the rarity of tracheal ACC, evidence on systemic therapy is limited.

This case highlights the importance of cross-sectional imaging in patients with atypical or refractory obstructive respiratory symptoms and underscores the role of multidisciplinary collaboration in managing rare tracheal malignancies, particularly when surgical resection is not feasible.

CONCLUSION

This case demonstrates the importance of maintaining a broad differential diagnosis, even in nonsmokers with atypical presentations. Tracheal ACC is rare and indolent, often delaying diagnosis. Management requires an individualized, multidisciplinary approach that integrates surgical, endoscopic, radiation, and oncologic expertise to achieve airway stabilization and long-term disease control. Reconsideration of an asthma diagnosis in patients with progressive or treatment-refractory symptoms may facilitate earlier identification of structural airway disease and prevent advanced obstruction.

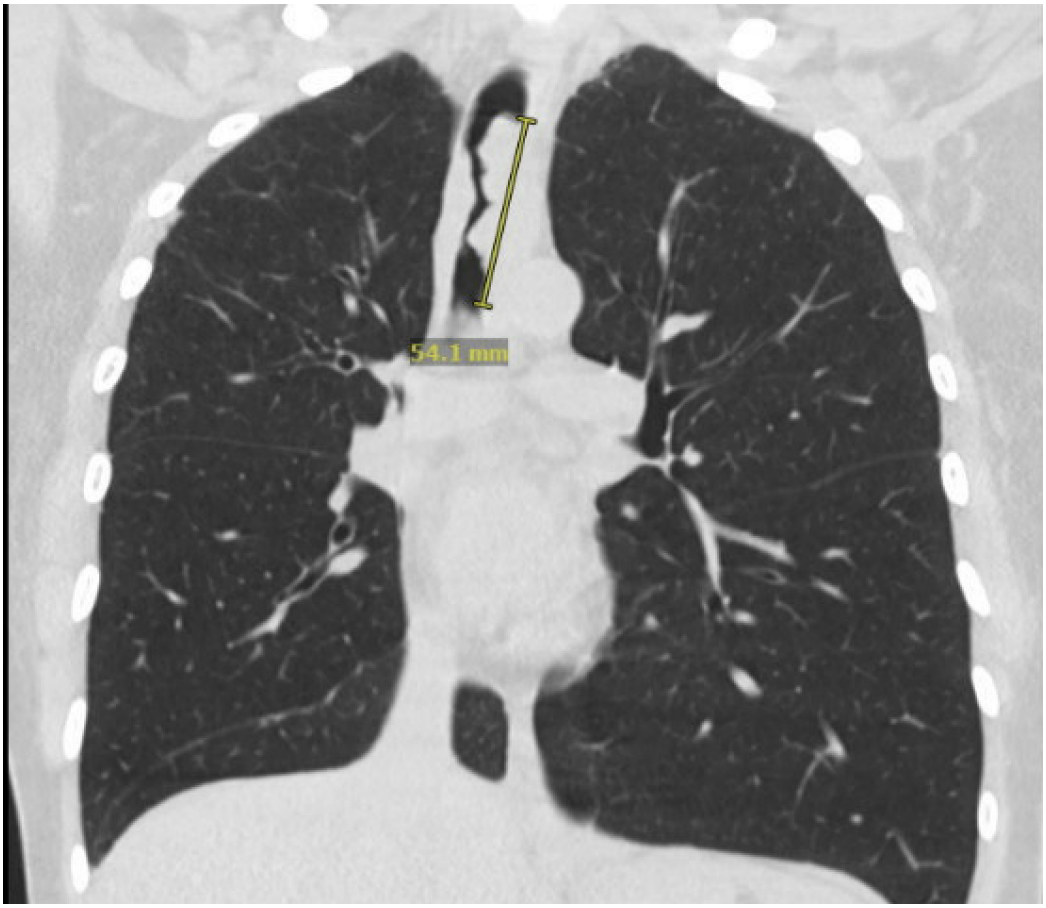

Figure 1. A component of the mass extended into the tracheal lumen, resulting in moderate airway stenosis measuring 5.41 cm identified by the yellow line.

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