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11<sup>th</sup> International Meeting on General Thoracic Surgery



10<sup>th</sup> International Workshop on Surgical Exploration of the Mediastinum and Systematic Nodal Dissection



5<sup>th</sup> Meeting of the Thoracic Oncology, Thoracic Surgery, Techniques & Transplant, Respiratory Nursing and Respiratory Physiotherapy Areas of the Spanish Society of Pneumology and Thoracic Surgery (SEPAR)



3<sup>rd</sup> Joint Meeting of the Spanish Society of Thoracic Surgery (SECT)



30<sup>th</sup> Congress of the "Asociación Iberoamericana de Cirugía Torácica" AIACT



10<sup>th</sup> International Workshop on Surgical Exploration of the Mediastinum and Systematic Nodal Dissection



## TREATMENT OF ADENOID CYSTIC CARCINOMA

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**Introduction:** Adenoid cystic carcinoma (ACC) of the trachea is a rare, slow-growing malignant neoplasm originating from the salivary glands. Its management presents unique challenges due to its location, potential for late recurrence, and resistance to conventional therapies. Tracheal ACC is the second most common primary tracheal tumor, although the most common in surgical series. Its indolent nature, combined with a tendency for perineural invasion, local recurrence, and late metastasis, necessitates a comprehensive and multidisciplinary approach to management.

**Diagnosis:** Diagnosis of tracheal ACC typically involves a combination of imaging, endoscopic examination, and histopathological analysis. Computed tomography (CT) is instrumental in assessing the extent of the tumor and its relationship to surrounding structures. ACC is generally limited to the trachea, carina, and proximal mainstem bronchi. Bronchoscopy allows for direct visualization and biopsy, defining the anatomy and confirming the diagnosis.

**Endoscopic Management:** Patients may present with symptoms of airway obstruction. Endoscopic palliation of airway obstruction is often an important step in stabilizing the airway to allow completion of workup, clearance of distal inflammation, and allowing definitive surgical resection to be elective rather than emergent. Obstructive adenoid cystic tumors can be temporarily palliated by a mechanical core-out of endoluminal tumor. Unresectable or recurrent tumors with airway obstruction can also be palliated with endoluminal debulking and sometimes augmented by stenting.

**Surgical Management:** Surgery remains the cornerstone of treatment for localized tracheal ACC. In most patients, up to 50% of the tracheal length can be resected with primary anastomosis, but this can be less for older patients or those with kyphosis. Knowledge of tracheal mobilization techniques – pretracheal dissection, neck flexion, hyoid release, hilar release – are critical to achieving a tension-free anastomosis. Depending on location and extent, resection may be a cervical or transthoracic tracheal resection and reconstruction, a carinal resection and reconstruction, or carinal pneumonectomy, necessitating a skilled surgical team.

to manage a variety of approaches and reconstructive techniques. Straightforward tracheal resections can be managed well with standard cross field ventilation during the airway reconstruction but more complex carinal reconstructions can be facilitated by ECMO support.

**Radiation Therapy:** Adjuvant radiation therapy is recommended in almost all cases due to the propensity for perineural invasion beyond visible margins, even when the histologic margin is negative. Radiation is also an alternative to surgery to control local disease in patients who are unresectable, high-risk for surgery, or decline surgery. Neutron radiation has been shown to be superior to photon radiation in ACC and other minor salivary gland tumors.

**Chemotherapy:** The role of chemotherapy in the management of tracheal ACC is limited, as these tumors exhibit low responsiveness to conventional chemotherapeutic agents. However, chemotherapy may be considered in advanced or metastatic cases, often in combination with radiation therapy. Targeted therapies and immunotherapy are emerging areas of interest, with ongoing research exploring their potential efficacy in ACC.

**Follow-Up and Surveillance:** Long-term follow-up is crucial due to the risk of late recurrence and metastasis. Regular surveillance includes periodic imaging and endoscopic evaluations. The slow-growing nature of ACC means that recurrences can occur many years after initial treatment, underscoring the need for 10-15 years of surveillance.

**Prognosis and Outcomes:** Prognosis for tracheal ACC varies based on the extent of the disease at diagnosis and the success of surgical resection. Patients with localized disease who undergo complete resection generally have better outcomes. However, the overall prognosis remains guarded, with a significant risk of local recurrence and distant metastasis, often many years after initial treatment.

**Conclusion:** The management of adenoid cystic carcinoma of the trachea requires a multidisciplinary approach, combining surgical expertise, advanced radiation techniques, and diligent long-term follow-up. Despite the challenges associated with this rare malignancy, advances in surgical and radiation therapies offer hope for improved outcomes. Ongoing research into novel therapeutic approaches, including targeted therapies and immunotherapy, holds promise for the future management of this complex disease.