ADENOID CYSTIC CARCINOMA TRACHEA UNUSUAL CASE
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Case report:
40 year old gentleman presented with the complaints of cough and progressive dyspnea of 9 months duration and hoarseness for 1 month duration. Nasopharyngolaryngoscopy showed reddish growth in trachea about 2-3 cm below subglottis almost occluding lumen. Bronchoscopy showed shiny soft polypoid lesion almost completely obstructing the tracheal lumen. MRI of the neck showed there was a mid tracheal growth from 4 cm to 5.7 cm of carina partially obstructing the lumen with peritracheal fat planes involvement and few peritracheal nodes. He underwent tracheal resection with primary anastomosis. Per operative trachea was 90% occluded at the level of fifth to eighth tracheal rings. There was a large mass measuring 4 cm x 3 cm x 2.5 cm within the lumen of the trachea arising mainly from the posterior wall of trachea and infiltrating posteriorly into the right tracheo-oesophageal groove. Growth was polypoidal with soft friable consistency. Post operative histopathology specimen was reported as Intermediate to high grade adenoid cystic carcinoma with perineural invasion, infiltrating entire thickness with extension into soft tissue involving the margins. He subsequently received postoperative adjuvant radiation therapy using Cobalt 60 beam and a dose of 40 Gy in 20 fractions to midplane by AP-PA technique during phase 1 and 20 Gy in 10 fractions to tumor volume with oblique pair beams during phase 2. He tolerated the treatment well. He was disease free till August 2015 (8 years of follow up).

Discussion:
Adenoid cystic carcinoma of the trachea originates from submucosal glands of the tracheobronchial tree. Primary Adenoid cystic carcinoma occurs predominantly in the trachea (36%), less frequently in the major bronchi (24%), lobar bronchi (18%), uncommonly in the larynx(3%) and rarely in peripheral locations in the lungs(3). Adenoid cystic carcinoma (ACC) is the second most common malignant tracheal tumor after squamous cell carcinoma. It has no gender predilection and not associated with smoking. It occurs in the age of 20-80 years with mean in the fifth decade(4). Patients with Adenoid cystic carcinoma of trachea usually present with symptoms such as coughing, wheezing and dyspnea and are often treated for asthma for months to years before being diagnosed. Hemoptyis is the most common symptom (43%)(5). The tumour is alike an iceberg, with the base wider than the intraluminal projection of nodular or lobulated masses covered with mucosa revealing increased vascularity. It has propensity for insidious longitudinal and circumferential extension.

Abstract:
BACK GROUND Adenoid cystic carcinoma of the lower respiratory tract is a rare indolent neoplasm with prolonged survival, propensity for recurrences and metastasis years after initial therapy(1). We report the presentation, treatment and outcome an uncommon tumor adenoid cystic carcinoma of trachea.

METHOD We collected retrospective data about our patient, treated in 2007 from our Hospital Information system. RESULTS 40 year old gentleman presented with the complaints of cough and progressive dyspnea of 9 months duration and hoarseness for 1 month duration. Bronchoscopy showed shiny soft polypoid lesion almost completely obstructing the tracheal lumen. MRI of the neck showed there was a mid tracheal growth from 4 cm to 5.7 cm of carina partially obstructing the lumen with peritracheal fat planes involvement and few peritracheal nodes. He underwent tracheal resection with primary anastomosis. Per operative trachea was 90% occluded at the level of fifth to eighth tracheal rings. There was a large mass measuring 4 cm x 3 cm x 2.5 cm within the lumen of the trachea arising mainly from the posterior wall of trachea and infiltrating posteriorly into the right tracheo-oesophageal groove. Growth was polypoidal with soft friable consistency. Post operative histopathology specimen was reported as Intermediate to high grade adenoid cystic carcinoma with perineural invasion, infiltrating entire thickness with extension into soft tissue involving the margins. He subsequently received postoperative adjuvant radiation therapy using Cobalt 60 beam and a dose of 40 Gy in 20 fractions to midplane by AP-PA technique during phase 1 and 20 Gy in 10 fractions to tumor volume with oblique pair beams during phase 2. He tolerated the treatment well. He was disease free till August 2015 (8 years of follow up).

Conclusion:
The challenge is to find the best therapeutic regimen aiming for a true cure and also to discover the best systemic therapy by expounding on molecular studies leading to targeted cell therapy to address a currently notable chemoresistant cancer. A multicenter long-term study including ACC of the head and neck region will be desirable.

Keyword: adenoid cystic carcinoma, trachea, Cobalt -60, radiation therapy

Introduction:
Adenoid cystic carcinoma of the lower respiratory tract is a rare indolent neoplasm with prolonged survival, propensity for recurrences and metastasis years after initial therapy(1). Tracheal malignancies occur in only 0.2 per 100000 people per year(2). We report the presentation, treatment and outcome an uncommon tumor – adenoid cystic carcinoma of trachea.
submucosally and perineurally long distances beyond where it is grossly visible (6). Lymphatic spread is uncommon (13%) (7). More than 50% of patients with tracheal ACC have hematogenous metastases. Pulmonary metastases are the most common. Histological subtypes described are cribriform (most common), tubular and solid tumors. Since they are infiltrative, complete resection are often difficult to achieve (1). Wide surgical resection is the most often advocated initial treatment, as many reported superior results in terms of duration and quality of survival. Surgery is fraught with technical difficulties because of the tendency for extensive local invasion and the common location at anatomically challenging sites. Despite advancement in thoracic surgical techniques that upgraded the respectability of many airway lesions, between 38 and 63% of patients were found to have positive airway resection margins (8).

Radiotherapy plays an important role in adjuvant setting and tumors that cannot be resected. Adjuvant radiation therapy improves local control in those having unknown or positive margins. The tumour is radiosensitive and disease remission, and therefore prognosis, is well correlated with radiation dose (9). Local control with definitive external beam radiation range from 20 to 79%, with better control and possible curative intent when a radical dose of >60 Gy is used (10). It is shown to provide local control and may improve survival (3). In the era of conformal techniques, we are able to achieve similar tumor control and lesser toxicity profile through the traditional conventional technique. The response of ACC to systemic therapy thus far has been poor. Chemotherapy has no established role in either the palliative or curative setting. Clinical trials have shown that KIT-activating mutations are correlated with response to TKIs. There are potential avenues for novel target treatment (11).

Conclusion

ACC of the trachea is a rare presentation of an indolent neoplasm. Successful tumor control with optimal conventional techniques still holds promise in the current era of conformal techniques. The challenge is to find the best therapeutic regimen aiming for a true cure and also to discover the best systemic therapy by expounding on molecular studies leading to targeted cell therapy to address a currently notable chemoresistant cancer. A multicenter long-term study including ACC of the head and neck region will be desirable.

References
