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Adenoid Cystic Carcinoma of Trachea Treated with Radical Radiotherapy: A Case Report

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Abstract: Adenoid cystic carcinoma of trachea is rare and is associated with prolonged survival and better prognosis. It is often misdiagnosed as chronic bronchitis or asthma. The ideal treatment option is surgical resection with tracheal reconstruction followed by adjuvant radiotherapy. Post operative radiotherapy may decrease the risk of local recurrence, but has no effect on the survival. In this report, we present a case of a patient with adenoid cystic carcinoma of trachea treated only by radical radiotherapy by intensity modulated radiotherapy technique. Initial coring followed by radical radiotherapy may be considered in unresectable cases or in those patients who refuse surgery. Radical radiotherapy may alone treat adenoid cystic carcinoma of trachea without any surgical morbidity, indicating that adenoid cystic carcinoma may be relatively and not absolutely resistant to radiation. There is no role of chemotherapy in adenoid cystic carcinoma.
Keywords: Adenoid cystic carcinoma, Trachea, Coring, Radical radiotherapy, IMRT.

INTRODUCTION

Primary tracheal tumors are rare and represent 0.2% of all thoracic cancers [1]. Adenoid cystic carcinoma (ACC) differs from other malignant tracheal tumors with respect to its clinical features, natural history, and treatment approaches. Tracheal ACC is a slow-growing neoplasm, often diagnosed at advanced stages. It is associated with prolonged survival and better prognosis [2]. The ideal treatment option for tracheal ACC is surgical resection with tracheal reconstruction followed by adjuvant radiotherapy. The addition of radiotherapy improves the local control, but does not affect the survival.

CASE REPORT

A 63 year-old diabetic, hypertensive man with no addictions presented with complaints of progressively worsening dyspnea, cough and weight loss (~10 kg) of six months duration. Physical examination revealed stridor and decreased bilateral air entry with rhonchi heard during both expiratory and inspiratory phase of respiration. He was misdiagnosed as a case of asthma and given symptomatic treatment at local hospital.

CT scan of neck and chest showed a soft tissue mass lesion arising from subglottic larynx and post cricoid region with contiguous involvement of posterior aspect of proximal trachea and abutting the cervical oesophagus with significant compromise of the tracheal lumen. Bronchoscopy showed endotracheal tumor, arising 1.4 cm below true vocal cord, 5 cm in craniocaudal extent and occluding almost 80% of the tracheal lumen. In view of persistent stridor, he underwent rigid brochoscopic tumor debulking. Histology was suggestive of adenoid cystic carcinoma with predominantly tubular and cribriform pattern (Grade II).

Post debulking, MRI neck showed enhancing soft tissue mass involving the trachea for 5.6 cm length from C6 to D1 vertebral body with tracheal ring destruction and extraluminal extension (Fig.1). Cranially the lesion was extending into the subglottis, while posteriorly it was abutting and compressing the esophagus. In view of extensive disease, patient was planned for tracheal resection with reconstruction. However, the patient refused surgery and was therefore

referred for radical radiotherapy. He was planned and started on radical radiotherapy to tracheal mass by intensity modulated radiotherapy (IMRT) to a dose of 60.25 Gy/30# (Fig. 2). Follow up PET-CT after 6 and 12 months showed no evidence of tumor recurrence.

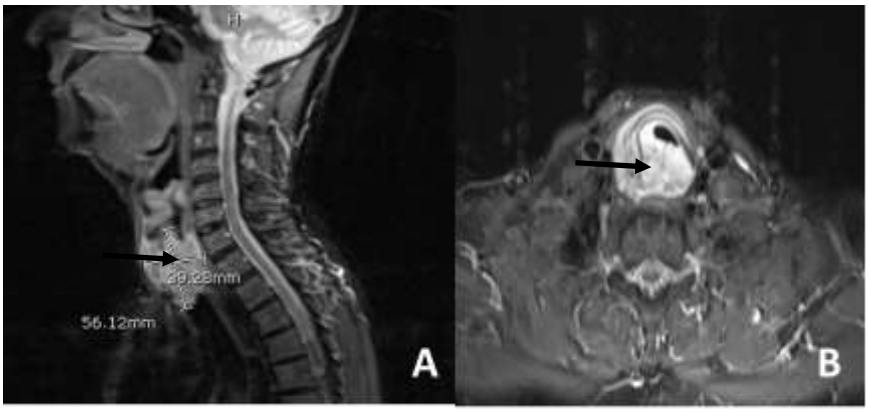

Fig. 1: MRI neck showing soft tissue mass involving subglottic trachea (Post coring) with both intraluminal and extraluminal component in A) sagittal and B) axial section

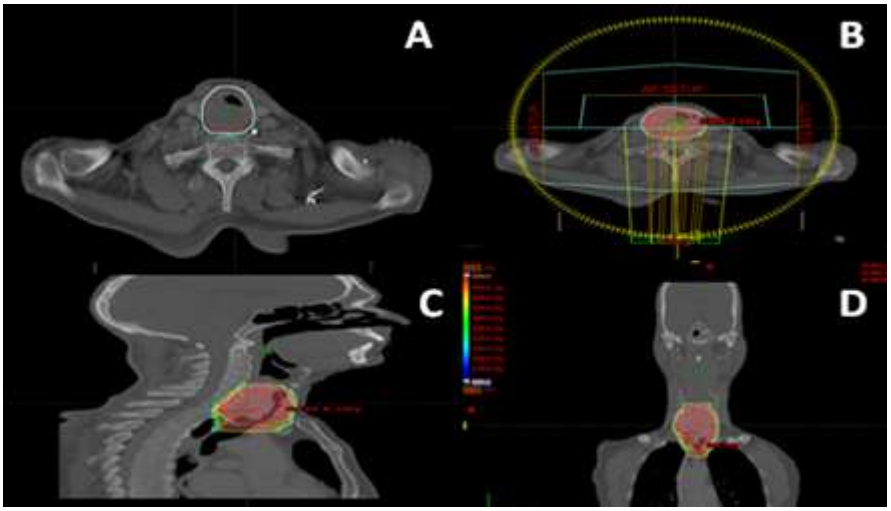

Fig. 2: A) Axial section of planning CT scan showing the Gross tumor volume (GTV) (red) and clinical target volume (CTV) (cyan), B) Tracheal tumor being treated by Arc therapy C) Sagittal dose distribution D) Coronal dose distribution of the radiation treatment plan [Pseudo-colouring shows the high dose (shown in red) and low doses (shown in green and blue) regions]

DISCUSSION

Primary tracheal tumors are rare intrathoracic tumors accounting for approximately 0.2 cases/1,00,000 population/year [3]. Patient typically presents with complaints of dyspnea, cough, hemoptysis, wheeze and stridor. They are often misdiagnosed as asthma or chronic bronchitis and treated for months to years before correct diagnosis is made. Therefore, for those patients with continuous progression or no improvement in symptoms with steroids, a radiographic evaluation to rule out upper airway obstruction is necessary.

Treatment options include surgery, radiation therapy, or a combination of both. Surgical options include primary tracheal resection and reconstruction, and endoscopic resection, either by coring or using a laser. Honings *et al*. [3] recommended tracheal resection followed by reconstruction as the best curative treatment, although this treatment is possible in 10-25% cases.

Both complete and incomplete resections can be done with a low operative mortality and a long overall survival has been reported in a majority of patients even with incomplete resection. Maziak *et al*. [1] demonstrated a trend towards better survival (10 year survival, 69% vs 30%) with complete rather than incomplete resection. Gaissert *et al*. [4] published the largest series of tracheal ACC, with 135 cases being reported at Massachusetts General Hospital over a 40 year period. Majority (71%) were treated with surgical

resection and showed 52% 5 year overall survival. Radiotherapy was administered after operation (54 Gy), except in superficial tumors, or as palliation (60 Gy). Adjuvant radiation may be beneficial and likely delays or reduces the incidence of local recurrence [5].

In unresectable, elderly and debilitated patients, tumor coring followed by radiotherapy (>60Gy) may lead to favourable outcome, with 5 year overall survival of 30% [6]. Unresectable ACC presents an increasingly difficult problem to control, given that the tumors often traverse long regions throughout the thorax. Primary radiotherapy has had mixed results, with local control varying between 20 - 70% (with better results with dose >60 Gy) [7, 8].

CONCLUSION

Complete surgical resection in ACC trachea provides the best chance of prolonged survival or even complete remission. Post-operative radiotherapy may have some effect on local control but does not affect survival. In summary, patients who are unresectable or who refuse surgery may be considered for radical radiotherapy after initial coring thereby decreasing surgical morbidity as ACC may be relatively and not absolutely radioresistant.

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