Adenoid Cystic Carcinoma of the Airway: 30-Year Review at One Institution

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CONCLUSIONS

ACC is an uncommon tumor of the airway. Due to the propensity of ACC to spread along perineural bundles and submucosally, the recommended treatment has been for surgical excision with postoperative radiotherapy. We suggest that prognostic outcomes and rates of recurrence likely vary according to subsite within the airway.

The largest series of laryngeal ACC are summarized in Table IV. 7-8 Combined, these case series support a high rate of early survival in ACC of the larynx (Table III). The discrepancy in late survival rates likely results from the paucity of cases of laryngeal ACC and the difficulty in obtaining adequate follow-up. In addition, our data supports the notion that negative margins are achievable in laryngeal ACC even with high grade lesions (Table II), and that distant disease affects survival.

As opposed to ACC of the larynx, ACC in the trachea is more commonly reported. Survival rates in other series range from 38% to 79% at 5 years, and from 29-70% at 10 years. 9, 10, 11-14 In contrast to laryngeal ACC, tracheal ACC seems to be more commonly associated with poorer local regional control as demonstrated by the 40% local recurrence in this series. Negative surgical margins are more difficult to obtain due to the relative inability to resect more than 6cm of the trachea.10 Prior studies cite positive margins in 8-82% of patients.9-10, 15 In our series, 80% of the patients with tracheal ACC had positive margins, which was significantly increased over patients with laryngeal ACC.

We present the experience of ACC of the airway over 30 years at our institution, highlighting the propensity of ACC to recur especially in the case of positive margins and distal tracheal location. ACC of the airway appears to behave similarly to other head and neck subsites and further multi-institutional studies are needed to evaluate and improve prognostic and therapeutic outcomes.

METHODS AND MATERIALS
All cases of ACC of the airway over a 30-year period at one tertiary care institution were retrospectively reviewed.

RESULTS

11 patients were treated for ACC of the airway with an age range of 25-72 years (median, 48 years, Table I, II, Fig. 1). 6 patients presented with ACC in the larynx, and 5 patients had ACC of the trachea. All patients underwent surgical excision and radiation; 9 of 11 patients had postoperative external beam radiation, one patient had preoperative external beam radiation and the remaining patient had postoperative neutron beam therapy. 4 patients with tracheal ACC, and none with laryngeal ACC, had microscopic or grossly positive margins after surgery (p=0.048, Table III). 80% of patients had perineural invasion on pathology. 2 patients with tracheal ACC had local recurrence of disease, which occurred at 1- and 10-months postoperatively (Fig. 2). One patient with laryngeal ACC died of distant metastatic disease at 16 months. Follow-up varied from 4 months to 168 months (median, 31 months).

DISCUSSION

ACC is a rare tumor of the salivary glands, and accounts for 10% of tumors of the head and neck. It can occur in any head and neck site containing salivary gland tissue. ACC typically presents with late metastases most commonly to the lungs2, but can also metastasize to the brain, bone, liver, thyroid and spleen3,4. ACC is also known for its neurotropic tendency, accounting for local regional recurrences many years after initial presentation and treatment5.

Laryngeal ACC is extremely rare. A review of the literature shows approximately 40 cases reported in the past 41 years1, 2, 7-8. Combined, these case series support a high rate of early survival in ACC of the larynx (Table III). The discrepancy in late survival rates likely results from the paucity of cases of laryngeal ACC and the difficulty in obtaining adequate follow-up. In addition, our data supports the notion that negative margins are achievable in laryngeal ACC even with high grade lesions (Table II), and that distant disease affects survival.

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REFERENCES