Failed Organ Preservation Strategy for Adult Laryngeal Embryonal Rhabdomyosarcoma
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ABSTRACT

OBJECTIVE:
To present a case of embryonal rhabdomyosarcoma of the intrinsic laryngeal musculature and discuss the treatment of this rare tumor.

STUDY DESIGN:
Case report and review of the literature.

METHODS:
A 45 year-old male presented with an 11 month history of hoarseness. A mass of the posterior glottis was noted on fiberoptic laryngoscopy, with biopsy consistent with embryonal rhabdomyosarcoma involving the interarytenoid muscle. A multidisciplinary tumor board recommended multimodality therapy including total laryngectomy.

RESULTS:
The patient refused surgery and was treated based upon pediatric rhabdomyosarcoma protocols with induction chemotherapy followed by radiation. There was no noted response to chemotherapy and the patient was taken off protocol to increase radiation dose without chemotherapy. 15 weeks following radiation, repeat biopsy revealed viable tumor. The patient currently is alive at 12 months post-treatment and continues to refuse surgery.

CONCLUSIONS:
Embryonal rhabdomyosarcoma involving the larynx is an extremely rare tumor usually seen in children. Although pediatric tumors can be effectively treated with organ preservation strategies, adult tumors may have a poorer response. Based upon our experience and existing literature regarding adult embryonal rhabdomyosarcoma of the larynx, multimodality therapy including surgical resection should be the treatment of choice.

INTRODUCTION

Embryonal rhabdomyosarcoma is a rare tumor which is usually described in children. While it is extremely rare to find this tumor in adults, involvement of the larynx has been described only in a limited fashion, which leads to difficulty in managing these lesions. Wide local excision, including total laryngectomy, is generally prescribed given the recurrent nature of these lesions. This article highlights the difficulty of managing patients with an organ sparing strategy.

CASE:

A 45 year old male presented with an 11 month history of hoarseness and mild laryngitis symptoms. An outside hospital otolaryngologist had noted a cyst on the right posterior glottis and had completed a direct laryngoscopy with biopsy of the firm lesion, with final pathology at that time consistent with fibrous muscular tissue with inflammation. A CT was obtained pre-operatively, and noted a 2.5 x 1.5cm laryngeal mass extending to the level of the cricoid without involvement of the cartilage.

Flexible laryngoscopic examination at presentation noted a bulky, posterior submucosal glottic mass, larger on the right side, which seemed to originate from the interarytenoid mucosa. This was obstructing the majority of the glottic inlet, and the subglottis was only poorly visualized. Direct laryngoscopy was advised, and deeper biopsies were obtained. Intraoperative frozen section histopathologic review was of limited value. The final pathology, however, demonstrated spindle cells invading the local musculature and entrapping nerve structures. The morphology and immunohistochemical profile were most consistent with embryonal rhabdomyosarcoma, stage I grade III.

DISCUSSION

There are three major subtypes of rhabdomyosarcoma: pleomorphic, alveolar, and embryonal. The embryonal variant is more frequently found in children, and the finding of laryngeal embryonal rhabdomyosarcoma in the adult larynx is exceedingly rare. Treatment algorithms have thus been poorly defined, but failure of chemoradiation ultimately requiring surgical resection has been described. While embryonal rhabdomyosarcoma in children is often treated with chemotherapy and occasionally radiation, the paucity of literature in the adult population makes management decisions more difficult. Although organ sparing strategies have obvious appeal, this case identifies the difficulty in managing laryngeal malignancies in this fashion. The lesion demonstrated little to no response to chemotherapy. Given his poor initial response to radiation, he received a larger than initially prescribed radiation dose. This did not prove successful. Ultimately, the lesion has had a stable to slightly increased size following completion of chemoradiation. Total laryngectomy continues to be recommended to the patient.

REFERENCES


CONCLUSION

Embryonal rhabdomyosarcoma of the adult larynx is a poorly described lesion that should be managed aggressively via a multi-disciplinary approach, with strong consideration of total laryngectomy.