

Laryngeal alveolar rhabdomyosarcoma involving the true vocal fold in an adult: Case report

Introduction

Rhabdomyosarcoma is rare in adults, especially in the head and neck region. More than 70% of rhabdomyosarcomas are diagnosed in children younger than 10 years.¹ Overall, 35% of rhabdomyosarcomas occur in the head and neck; of these, only about 10% arise in nonorbital, nonparameningeal locations.²

The three basic histologic variants of rhabdomyosarcoma are *embryonal*, *alveolar*, and *pleomorphic*. The embryonal subtype is the most common variant seen in the head and neck.³ With regard to age, the embryonal subtype is the most common variant in children, the alveolar type is seen most often in adolescents, and the pleomorphic subtype is most common in elderly patients.^{3,4}

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In the larynx, the vast majority of cancers are of squamous histology. Spindle cell tumors account for only 1% of all laryngeal neoplasms.⁵ In this article, we report what we believe is the first case of an alveolar rhabdomyosarcoma involving the true vocal fold in an adult to be reported in the English-language literature.

Case report

A 54-year-old black woman presented with a 9-month history of hoarseness following intubation during hip replacement surgery. During the immediate postoperative period, she had been aphonic. As her voice returned, her pitch had become significantly lower. She admitted vocal fatigue and denied dysphagia. Her voice was severely hoarse without evidence of stridor. She was healthy-appearing and in no respiratory distress. She had no history of tobacco or alcohol use.

Stroboscopy detected a submucosal mass of the right true vocal fold with expansion medially and superiorly. Vibratory motion was absent on the right, and glottic closure was incomplete posteriorly. Computed tomography (CT) demonstrated an asymmetric fullness of the right true vocal fold, which had caused it to bulge medially ([figure 1](#)).

Figure 1. CT demonstrates the asymmetric fullness of the right true vocal fold.

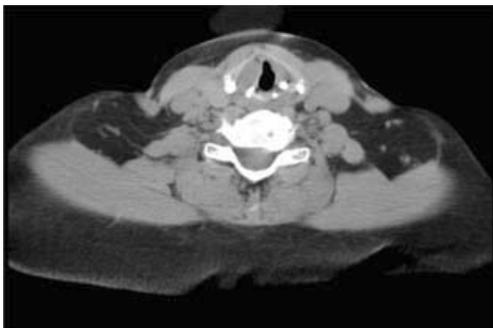
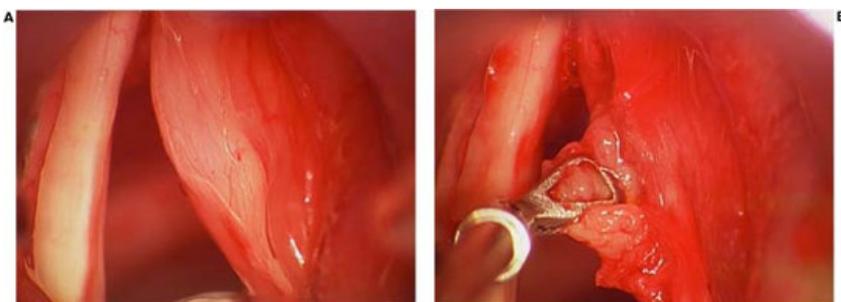


Figure 2. A: Microscopic direct laryngoscopy shows the fullness of the right true vocal fold, which had caused it to bow medially. B: The incisional biopsy is performed.



After providing informed consent, the patient was taken to the operating room for microscopic direct laryngoscopy. The right true vocal fold was erythematous and swollen ([figure 2, A](#)), and a firm submucosal mass was found deep to the vocal fold margin. A CO₂ laser was used to incise the mucosa 2 mm lateral to the vibrating margin. The mass had replaced the entire length of the true vocal fold. An incisional biopsy was performed ([figure 2, B](#)). On hematoxylin and eosin (H&E) staining, the mass was identified as a spindle cell neoplasm; immunohistochemistry was positive for MyoD1, desmin, and alpha-smooth-muscle actin (α-SMA) ([figure 3](#)). These features were consistent with alveolar rhabdomyosarcoma.

Following the biopsy analysis, positron-emission tomography (PET) demonstrated hypermetabolic activity within the larynx but no local or distant metastases. On repeat CT, a significant interval progression of the soft-tissue mass of the right true vocal fold was noted. The lesion measured 2.0 x 1.2 x 1.7 cm, and it had caused a partial obstruction at the level of the glottis. No evidence of thyroid cartilage invasion was seen.

After extensive discussion with the patient and consultation with the radiation and medical oncology units, we formed a treatment plan that included endoscopic surgical debulking followed by induction chemotherapy and then chemoradiation therapy. At surgery, a bulky, multilobulated, exophytic tumor was found occupying the entire right membranous vocal fold and extending 1 cm inferiorly. No involvement of the anterior commissure was observed, and the right cricoarytenoid joint was passively mobile. A CO₂ laser was then used to vaporize the tumor and remove all gross disease. Findings on pathology were again consistent with an alveolar rhabdomyosarcoma.

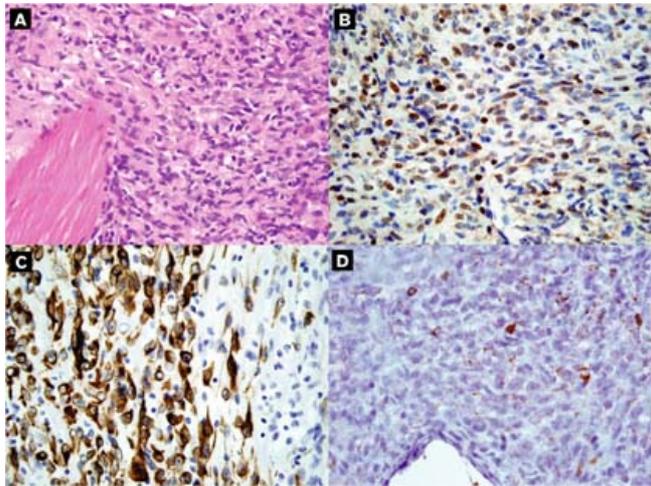
Postoperatively, the patient underwent three cycles of induction chemotherapy with vincristine, ifosfamide, and etoposide at 3-week intervals. She then underwent concomitant treatment with vincristine/ifosfamide and intensity-modulated radiotherapy. She received a mean radiation dose of 63 Gy to the laryngeal area. Following the completion of treatment, examination of the larynx revealed radiation change with some thickening of the arytenoid cartilages, good vocal fold mobility, and a clear subglottic airway. The patient's voice was slightly raspy, but her projection was adequate and she was easily understandable. At 7 months after the completion of therapy, follow-up PET revealed a complete resolution of the hypermetabolic activity. At 1 year after the completion of treatment, the patient remained clinically and radiographically free of disease.

Discussion

In the English-language literature, only 17 well-documented cases of laryngeal rhabdomyosarcoma in the adult population have been previously reported (table).⁶⁻²⁰ As far as we know, only 2 of these cases involved an alveolar rhabdomyosarcoma.^{9,12} Our patient represents the first reported case of an alveolar rhabdomyosarcoma of the true vocal fold in an adult.

In adults, rhabdomyosarcoma is the rarest mesenchymal tumor of the larynx. When it does occur, it is more prevalent in males.¹⁶ Of the various histologic subtypes, embryonal rhabdomyosarcomas commonly involve the head and neck, alveolar lesions usually occur in the upper extremities and perianal area, and pleomorphic tumors are typically located in an extremity. Rhabdomyosarcomas of the larynx appear to be less aggressive than those found at other locations in the head and neck.²¹ However, the alveolar subtype, which accounts for fewer than one-third of all head and neck rhabdomyosarcomas, tends to carry a worse prognosis.²²

Figure 3. H&E staining of a biopsy specimen shows short spindle cells arrayed in sheets with large atypical nuclei and moderate eosinophilic cytoplasm (A). Immunohistochemistry is positive for MyoD1 (B), desmin (C), and α -SMA (D). (Original magnification for all slides is x40.)



Rhabdomyosarcomas are staged according to the “clinical grouping system” developed by the Intergroup Rhabdomyosarcoma Study Group (IRSG).²³ This system takes into account resectability and the extent of disease. The four categories are:

- I. Localized disease completely resected
- II. Grossly resected tumor
- III. Incomplete resection or biopsy only with gross residual disease
- IV. Metastatic disease present at diagnosis

In addition to the IRSG clinical grouping system criteria, other factors that determine the prognosis are tumor site, histologic subtype, and response to treatment.^{23,24} At one time, the prognosis was extremely poor, but as a result of refinements in radiotherapy and multidrug chemotherapy, survival statistics have since improved markedly.^{3,25,26} Thus, the management of this disease has evolved from radical surgeries to less morbid procedures supplemented with radio- and chemotherapy. This paradigm shift can be attributed to the multimodality protocols initiated by the IRSG over several decades. With the discovery that many rhabdomyosarcomas are responsive to chemoradiation, surgery is now typically limited to organ-sparing procedures. Since rhabdomyosarcomas are rare in the adult population, most of the data regarding treatment responses are based on pediatric series. In children with localized disease, 5-year survival with current multimodality treatment protocols exceeds 70%.^{25,26}

As mentioned, only 2 cases of alveolar rhabdomyosarcoma of the adult larynx have been previously reported in the English-language literature.^{9,12} Winter and Lorentzen described the case of a 17-year-old boy who was found to have alveolar rhabdomyosarcoma of the epiglottis with metastatic disease to the left supraclavicular lymph nodes.⁹ (For the purposes of this article, we classified this 17-year-old as an adult.) The boy was treated with neck irradiation and chemotherapy with vincristine, cyclophosphamide, and prednisone. He died of his disease 9 months after treatment. In the other previously reported case of adult laryngeal alveolar rhabdomyosarcoma, Haerr et al described a case of alveolar rhabdomyosarcoma in the interarytenoid notch in a 62-year-old man.¹² That patient was also found to have bilateral metastatic cervical lymphadenopathy. He underwent a total laryngectomy, a left radical neck dissection, and a right modified radical neck dissection. Postoperatively, he was started on adjunctive chemotherapy with cyclophosphamide, lomustine (CCNU), and methotrexate. However, he developed a recurrence in the neck within 1 month of beginning chemotherapy, and the recurrence was treated with radiotherapy. Subsequently, he developed widespread disease and died within 5 months.

In accordance with the current treatment paradigm for children, our patient was treated with a conservative surgical approach that involved surgical debulking rather than a total laryngectomy. Postoperatively, she underwent induction chemotherapy followed by chemoradiation. One year following the completion of treatment, she exhibited no clinical or radiographic evidence of disease.

Table. Summary of reported cases of laryngeal rhabdomyosarcoma in adults

Author/yr	Age/sex	Histologic subtype	Location	Treatment	Results
Rodriguez and Ziskind, ⁶ 1970	57/M	Pleomorphic	True vocal fold	Total laryngectomy	No follow-up reported
Hall-Jones, ⁷ 1975	54/M	Embryonal	Posterior vestibular wall of the larynx	Total laryngectomy	No evidence of disease at 16 mo

Frugoni and Ferlito, ⁸ 1976	33/M	Pleomorphic	Vestibule, false vocal fold, and true vocal fold	Partial laryngectomy plus radiotherapy	No evidence of disease at 6 yr
Winter and Lorentzen, ⁹ 1978	17/M	Alveolar	Epiglottis	Radiotherapy plus chemotherapy	Died of disease at 9 mo
	72/M	Pleomorphic	Vocal fold	Partial laryngectomy	No follow-up reported
Srinivasan and Talvalkar, ¹⁰ 1979	55/M	Pleomorphic	Arytenoid, aryepiglottic fold	Laryngectomy planned	Died prior to surgery of acute laryngeal obstruction
Gorenstein et al, ¹¹ 1980	45/M	Not specified	Aryepiglottic fold	Laryngectomy plus radiotherapy and chemotherapy	Died of disease at 15 mo
Haerr et al, ¹² 1987	62/M	Alveolar	Interarytenoid notch	Total laryngectomy plus radiotherapy and chemotherapy	Died of disease at 5 mo
Da Mosto et al, ¹³ 1996	69/M	Pleomorphic	True vocal fold	Total laryngectomy plus radiotherapy	No evidence of disease at 2 yr
Ruske et al, ¹⁴ 1998	66/F	Pleomorphic	Arytenoid area	Total laryngectomy plus radiotherapy	No evidence of disease at 1 yr
Libera et al, ¹⁵ 1999	66/M	Embryonal (botryoid)	Vestibule	Partial laryngectomy	No evidence of disease at 10 yr
Dikbas et al, ¹⁶ 2005	28/M	Embryonal	Ventricle	Induction chemotherapy plus radiotherapy	No evidence of disease at 22 mo
Schrock et al, ¹⁷ 2007	60/M	Pleomorphic	True vocal fold	Total laryngectomy plus radiotherapy and chemotherapy	No evidence of disease at 20 mo
	64/M	Pleomorphic	True vocal fold	Cordectomy	No evidence of disease at 20 mo
Shayah et al, ¹⁸ 2007	77/F	Pleomorphic	Arytenoid, aryepiglottic fold	Total laryngectomy	No evidence of disease at 1 yr
Papacharalampous et al, ¹⁹ 2009	83/F	Pleomorphic	Subglottis	Endoscopic resection	No evidence of disease at 16 mo
Pittore et al, ²⁰ 2010	75/M	Pleomorphic	True vocal fold	Partial laryngectomy	No evidence of disease at 9 mo
Leventhal et al,* 2010	54/F	Alveolar	True vocal fold	Endoscopic surgical excision plus radiotherapy and chemotherapy	No evidence of disease at 1 yr

*Present case.

In conclusion, rhabdomyosarcoma involving the larynx is rarely diagnosed in the adult population, and to the best of our knowledge, a case of adult alveolar rhabdomyosarcoma of the true vocal fold has not been previously reported. The documented success of the current treatment regimen for the pediatric population supports its use in the adult population, as well. The pediatric protocol of conservative surgery supplemented with radio- and chemotherapy may be the most efficacious for adults with laryngeal alveolar rhabdomyosarcoma.

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