Adult-Onset Supraglottic Rhabdomyosarcoma: A Rare Case Report

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Abstract

Rhabdomyosarcoma (RMS) of the larynx is an exceedingly rare malignancy in adults, typically arising in the pediatric population. We report a case of a 60-year-old male presenting with hoarseness, dysphagia, and intermittent stridor. Laryngoscopic examination revealed a large supraglottic mass with white necrotic debris. Histopathology confirmed the diagnosis of pleomorphic rhabdomyosarcoma. This case underscores the importance of considering non-epithelial malignancies in the differential diagnosis of laryngeal tumors and highlights the role of immunohistochemistry in diagnosis.

Keywords: Laryngeal rhabdomyosarcoma; Supraglottic tumor; Adult sarcoma; Pleomorphic RMS; Laryngeal malignancy

1. Introduction

Rhabdomyosarcoma (RMS) is a malignant tumor of mesenchymal origin showing skeletal muscle differentiation. It is the most common soft tissue sarcoma in children but is rare in adults, accounting for less than 1% of adult soft tissue sarcomas. Laryngeal involvement is particularly rare, with few cases reported in the literature-especially in adults where squamous cell carcinoma predominates.

Among RMS subtypes- embryonal, alveolar, and pleomorphic—the pleomorphic form is most common in adults and typically has a more aggressive course. Due to its rarity, adult laryngeal RMS poses significant diagnostic and therapeutic challenges.

2. Case Presentation

A 60-year-old male presented to our ENT clinic with a three-month history of progressive hoarseness, dysphagia to solids, and intermittent stridor, especially on exertion. He was a heavy smoker (40 pack-years) and had a medical history of controlled hypertension.

Laryngoscopic Examination

Flexible fiberoptic laryngoscopy revealed a large, fungating supraglottic mass involving the epiglottis and aryepiglottic folds, partially obstructing the airway. The surface was irregular with white necrotic debris. Vocal cord mobility could not be assessed due to obstruction.

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Radiologic Findings

Contrast-enhanced CT of the neck showed a heterogeneously enhancing supraglottic mass with no cartilage invasion or regional lymphadenopathy. There was partial airway narrowing.

Biopsy and Histopathology

An incisional biopsy was performed under general anesthesia. Histopathological examination revealed sheets of pleomorphic spindle and polygonal cells with abundant eosinophilic cytoplasm. Numerous mitotic figures were seen.

Immunohistochemical staining was positive for

- Desmin
- Myogenin
- MyoD1

These findings confirmed the diagnosis of pleomorphic rhabdomyosarcoma.

3. Discussion

Laryngeal RMS is an extremely rare diagnosis in adults. To our knowledge, fewer than 30 cases have been reported in the English literature. Unlike pediatric RMS, which frequently affects the head and neck, adult RMS has a predilection for the extremities and trunk.

Clinical Presentation of laryngeal RMS mimics squamous cell carcinoma (SCC), presenting with hoarseness, dysphagia, and airway obstruction. In our case, the presence of stridor and rapidly enlarging mass raised suspicion of aggressive pathology.

Histologic Subtypes

- Embryonal: Common in children, better prognosis.
- Alveolar: Aggressive, often in adolescents.
- Pleomorphic: Predominant in adults, associated with poor prognosis.

Immunohistochemistry is essential for diagnosis, particularly in distinguishing RMS from sarcomatoid carcinoma or poorly differentiated SCC. Expression of desmin, myogenin, and MyoD1 are diagnostic markers.

Treatment Considerations

There is no standardized treatment protocol for adult laryngeal RMS. Management is extrapolated from pediatric guidelines, typically involving:

- Surgical resection (often total laryngectomy)-Radiotherapy
- Systemic chemotherapy (e.g., VAC protocol: vincristine, actinomycin D, cyclophosphamide)

In our case, due to the extensive involvement of the supraglottis and compromised airway, total laryngectomy followed by adjuvant chemoradiotherapy was planned. The patient was counseled regarding prognosis and rehabilitation options.



Figure 1: Endoscopic view of the supraglottic mass showing exophytic, necrotic lesion obstructing the supraglottic inlet.

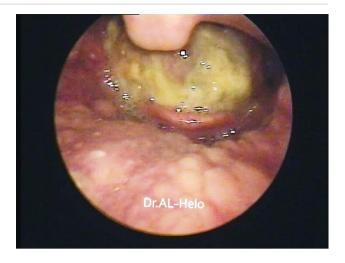


Figure 2: Close-up endoscopic view of the tumor extending to the epiglottis and aryepiglottic folds.



Figure 3: Inferior view of the mass with involvement of the supraglottic structures and obscured vocal cords.

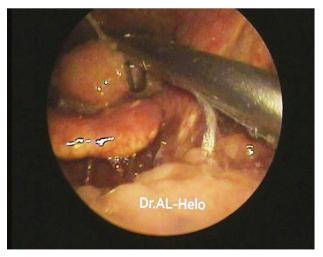


Figure 4: Biopsy for histopathology.

4. Conclusion

This case highlights the need to consider rhabdomyosarcoma in the differential diagnosis of

laryngeal masses, particularly in patients presenting with rapid-onset obstructive symptoms. Early biopsy and immunohistochemical confirmation are critical for diagnosis. Due to its rarity and aggressive behavior, adult laryngeal RMS requires a multidisciplinary approach and further documentation to guide treatment strategies.

Conflicts of Interest

None declared.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

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