An Unusual Cause of Hoarseness: Rhabdomyosarcoma of the Larynx

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INTRODUCTION

Squamous cell carcinomas are the most common malignancy of the larynx with laryngeal sarcomas accounting for <1% of all laryngeal malignancies. Only a few reports exist discussing laryngeal spindle cell rhabdomyosarcoma. We report a case of a 30 year old man presenting with hoarseness found to have glottic spindle cell rhabdomyosarcoma. Symptoms, subtypes, histology, treatment options, and surveillance recommendations are discussed.

METHODS AND MATERIALS

Case Report

A 30 year old man with a seven month history of progressively worsening hoarseness was referred from an outside otolaryngologist for surgical management of a large laryngeal polyp. He endorsed dysphagia and shortness of breath upon neck extension. On indirect fiberoptic laryngoscopic exam, a very large mucosa-covered polypoid lesion originating from the right true vocal fold was seen. The mass obstructed nearly eighty percent of the anterior and mid airway and prevented vocal fold adduction.

RESULTS

The patient was taken for urgent laryngoscopy where it was noted the mass appeared to emanate from the anterior portion of the right true vocal fold. It was firm with a broad base extending across the anterior one-third of the right vocal fold. Additionally, at the anterior aspect by the anterior commissure, the mass was intrinsic to the vocal fold and extended laterally into the ventricle.

A computed tomographic scan with contrast of the neck was done post-operatively showing a 1.2 centimeter soft tissue mass at the right anterior vocal fold extending past midline (Figure 1). No significant lymphadenopathy was visualized.

Pathologic consultation confirmed a spindle cell rhabdomyosarcoma of the larynx. The patient subsequently underwent frontolateral partial laryngectomy followed by adjuvant radiation and chemotherapy with Vincristine. The patient is currently disease free at his 15 month follow up visit, is tolerating a regular diet, and has an acceptable voice quality.

DISCUSSION

Rhabdomyosarcoma arises from undifferentiated mesodermal tissue and is a malignant tumor of striated muscle, which rarely occurs in the larynx with even fewer cases reported in adults. Symptoms range from hoarseness to dysphagia or as serious as respiratory failure. RMS can be subdivided into four main types: embryonal, alveolar, pleomorphic, and botryoid. Spindle cell RMS are a subtype of embryonal and were first described in children in 1992 and in adults in 1998. RMS histologically show invasion into skeletal muscle. On immunohistochemistry, RMS stain positive for desmin, vimentin, myoglobin, and MyoD1. MyoD1 protein is highly sensitive and specific for RMS.

Multimodality therapy is the mainstay of treatment for head and neck RMS though not enough data exists regarding the optimal treatment of laryngeal RMS, especially in adults. Historically, radical surgery consisting of a total laryngectomy with post-operative radiation therapy was the treatment of choice; however, conservative surgery followed by adjuvant chemotherapy and radiation may be possible.

Newer imaging techniques are emerging as possible adjuncts to or substitutions for conventional imaging for surveillance. The overall TNM staging was much more accurate using FDG PET/CT rather than conventional imaging especially in reference to finding distant metastases.

CONCLUSIONS

Adult laryngeal RMS is an unusual neoplasm. No true consensus has emerged for the management of adults with spindle cell RMS. Additionally, no case reports of RMS of the larynx have been described in the English literature in the same age range as our patient. After the pathology results were confirmed, our patient underwent frontolateral partial laryngectomy with negative margins followed by radiation and single agent chemotherapy. This treatment plan was designed to provide organ preservation for this young adult and prevent local-regional recurrence. Our patient will be followed regularly in the clinic and will have periodic FDG PET/CT for surveillance.

REFERENCES