A rare case of rhabdomyoma of the larynx causing airway obstruction

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Abstract
Rhabdomyomas are benign tumors of skeletal muscle derivation. Rhabdomyomas of the head and neck are unusual, and laryngeal rhabdomyomas of the adult type are rare. When they do occur, patients may present with symptoms that have progressed slowly over several years. In extraordinary cases, stridor and airway obstruction manifest. We report a case of adult rhabdomyoma of the larynx that features some of its typical and atypical presenting characteristics. This case also exemplifies the favorable prognosis that results following complete yet conservative surgical excision.

Introduction
Rhabdomyomas are rare benign neoplasms that are derived from striated muscle. They are more common in the myocardium than in skeletal muscle. Rhabdomyoma can be differentiated into three types: adult, fetal cellular, and fetal myxoid:

• Adult-type tumors, which are confined to the head and neck, typically feature well-defined margins and contain large, densely packed round cells with granular eosinophilic cytoplasm and cross-striations.
• Tumors of the fetal cellular type are extremely rare. They occur in children younger than 4 years of age and in the head and neck of elderly men. These tumors are poorly defined and are made up of immature, elongated, spindle-shaped skeletal muscle elements in various stages of differentiation; mature cells are few.1 Fetal cellular rhabdomyomas in children have a predilection for the posterior auricular area.2
• Fetal myxoid rhabdomyomas are tumorlike polypoid masses with loose, edematous, and myxoid stroma. These tumors may be found in the vagina and vulva of middle-aged women.1

Most extracardiac rhabdomyomas occur in the head and neck,2 and most are of the adult type.1 In 1988, Helliwell et al reported a large series of 115 extracardiac rhabdomyomas and demonstrated that most occurred in the head and neck.2 In 1995, Johansen and Illum published a review of 23 reported cases of rhabdomyoma of the larynx and reported that 15 of them were of the adult type.1 In the latter study, most patients had presented with progressive hoarseness and dysphagia as their chief complaints; only 1 of these patients had presented with acute airway obstruction, and 1 other patient had presented with dyspnea along with dysphagia. In 1994, Roberts et al reported a case of laryngeal rhabdomyomathat featured worsening dyspnea with positional stridor.3

In this article, we report a case of adult rhabdomyoma of the larynx that typified the common indolent progression of dysphagia and dysphonia but featured a more acute presentation of upper airway obstruction.

Case report
A 66-year-old man was referred to the otolaryngology service for evaluation of a possible supraglottic mass that had been suggested by routine endoscopic gastroscopy performed during an assessment of dysphagia. The patient relayed a 3-year history of progressive dysphagia (primarily with solids) and a vocal coarseness. He also related that he had experienced some difficulty breathing over the previous several months and that his breathing was significantly noisy when he was supine and during minimal exertion. He reported no odynophagia, unexpected weight loss, fevers, or other aerodigestive or systemic symptoms. He had no history of congenital defects, head and neck cancer, radiation therapy, or relevant trauma.

On physical examination, a notable inspiratory upper airway noise could be appreciated on auscultation. Nasopharyngoscopy detected a large, smooth, pedunculated mass emanating from the vocal process of the right ary-
tenoid. With each inspiration, the mass would ball-valve into the laryngeal introitus and cause a near-total airway obstruction.

The patient was taken urgently to the operating suite, where extirpation of the mass via suspension laryngoscopy was performed (figure 1). The mass was attached by an insubstantial stalk that was pedicled to the vocal process of the right arytenoid. The patient did not require a surgically created airway, and he experienced no complications.

On gross examination, the excised specimen was a smooth soft-tissue mass with a homogenous tan-brown surface. It measured $2.0 \times 1.4 \times 1.3$ cm. Microscopic examination demonstrated that the large tumor mass was made up of large polygonal cells with distinct cell borders and abundant granular eosinophilic cytoplasm. Cross-striations and large vacuoles were noted in the cytoplasm of many of the cells. The nuclei were uniformly large with prominent nucleoli. No mitotic activity was identified. The cytoplasmic granules were positive on periodic acid-Schiff staining and diastase-labile, indicating the presence of glycogen. Immunohistochemical analysis revealed strong reactivity in the tumor cells for desmin (figure 2).

The patient made a full recovery and reported a complete cessation of dysphagia, dysphonia, and airway obstruction. At the 18-month follow-up, he exhibited no evidence of tumor recurrence.

Figure 1. The smooth, round, pedunculated supraglottic mass is seen during direct suspension laryngoscopy.
The term rhabdomyoma was first used in 1864 by Zenker to describe a striated muscle tumor. Rhabdomyomas are generally benign, never metastasize, and rarely recur. Rhabdomyomas of the heart are usually hamartomatous growths, and they have been associated with phakomatoses such as tuberous sclerosis. Extracardiac rhabdomyomas account for only 2% of skeletal muscle tumors, and 70% of them occur in the head and neck. They are thought to arise from unsegmented mesoderm in the branchial arches. Most reported cases of extracardiac rhabdomyoma have involved solitary masses; the few that have occurred multifocally were associated with no clinically significant difference in treatment or outcome. Adult rhabdomyoma occurs more frequently in men than women (4:1). The reported ages of affected patients have ranged from 16 to 76 years (mean: 52).

The most common symptom of laryngeal rhabdomyoma is hoarseness, a foreign-body sensation, and dysphagia. The duration of symptoms is usually long, typically years. In rare cases, progressive airway obstruction has occurred; such obstructions were usually secondary to pedunculated supraglottic mass that blocked the airway during inspiration or when it was in a dependent position.

Histologic analysis is essential for diagnosis. Adult rhabdomyomas have been confused with granular cell tumors during initial evaluation. Battifora et al used light and electron microscopy to compare the two tumors. They found that rhabdomyomas are characterized by the presence of a sarcolemma sheath, rodlike cytoplasmic bodies, and cross-striations on light microscopy. Ultrastructural studies showed actin filament and Z bands. There were no contractile elements found in the granular cell tumor. The differential diagnosis also includes rhabdomyosarcoma, oncocytoma, and alveolar soft-tissue sarcoma.

The curative treatment of rhabdomyomas of the supraglottic airway is complete surgical excision. This is most easily approached via suspension laryngoscopy. Successful removal of these typically poorly vascularized tumors can be achieved with microlaryngeal instrumentation. Overall, the long-term prognosis is excellent, and recurrence rates are very low.

References