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Leiomyosarcoma of theLarynx: a case report

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Leiomyosarcoma of the larynx: A case report

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Abstract

Leiomyosarcoma of the larynx is an extremely rare neoplasm; only about 50 cases have been reported in the English-language literature. We report a new case of laryngeal leiomyosarcoma in a 65-year-old man. The metastatic work-up was negative, and the patient underwent total laryngectomy. He remained disease-free 12 months postoperatively.

Introduction

Compared with epithelial tumors, malignant mesodermal neoplasms of the larynx are very rare. In fact, they account for less than 1% of all malignant laryngeal tumors.[1] Leiomyosarcomas originate in the smooth muscles of the cutaneous and subcutaneous tissues, the lower gastrointestinal/genitourinary tract, and the vascular walls. Approximately 85% of leiomyosarcomas develop in the extremities; only 3% occur in the head and neck.[2] Most lower-torso tumors arise in the female genital tract, the walls of the gastrointestinal tract, and the retroperitoneal area. Most leiomyosarcomas of vascular origin originate in the walls of the veins.[3] Patients with leiomyosarcomas of the cutaneous and subcutaneous tissue generally have a good overall prognosis.

When head and neck leiomyosarcomas have occurred, most have arisen in the oral cavity and in the superficial soft tissues of the scalp, paranasal sinuses, and jaws.[4] Other affected sites in the head and neck have included the tongue, trachea, hypopharynx, and cervical esophagus. Leiomyosarcoma of the larynx, first reported by Jackson and Jackson in 1939,[5] is extremely rare.[6] Only about 50 cases have been reported in the English-language literature. In a review of 31 of those cases, Marioni et al reported a wide variety of laryngeal subsites: glottis, 15 cases (48.4%); supraglottis, 10 (32.3%); supraglottis-glottis, 2 (6.5%); subglottis, 2; supraglottis-glottis-subglottis, 1 (3.2%); and glottis-subglottis, 1.[4] Smoking and alcohol use are not considered to be risk factors for laryngeal leiomyosarcoma.[7]

In this article, we report a new case of laryngeal leiomyosarcoma that was successfully treated with total laryngectomy.

Case report

A 65-year-old man was referred to our head and neck clinic for evaluation of a 5-month history of worsening hoarseness and a 3-day history of breathing difficulty. He had no history of smoking or alcohol intake. Physical examination revealed obvious stridor at rest, but he was able to achieve oxygen saturation on room air. Fiberoptic laryngoscopy revealed that a polypoid mass was occupying almost the entire glottic space. A very narrow glottic space could be seen posteriorly. The large size of the tumor prevented us from identifying its site of origin or the degree of vocal fold mobility. No cervical lymph nodes were palpable.

Emergency tracheotomy with local anesthesia was performed to secure the airway. Suspension laryngoscopy showed that the polypoid lesion had arisen from the right true vocal fold and anterior commissure. Subglottic extension could not be determined. The surface of the mass was smooth. The left vocal fold was clear. Examination of multiple biopsy specimens revealed that the histologic picture was consistent with a leiomyosarcoma (figure 1, A). Immunohistochemical studies demonstrated positive staining with antibodies to smooth-muscle actin (figure 1, B) and vimentin, and negative staining with S-100, cytokeratin AE1/AE3, and cytokeratin MNF.

Computed tomography (CT) of the neck confirmed that the well-enhancing mass originated in the right true vocal fold and anterior commissure (figure 2). The mass had infiltrated the entire glottis, but we noted only minimal subglottic extent and no supraglottic involvement. No cervical lymphadenopathy or destruction of the thyroid cartilage was evident, and the chest x-ray was clear. Ultrasonography of the liver found no evidence of metastatic disease, and the results of liver function tests were within normal limits.

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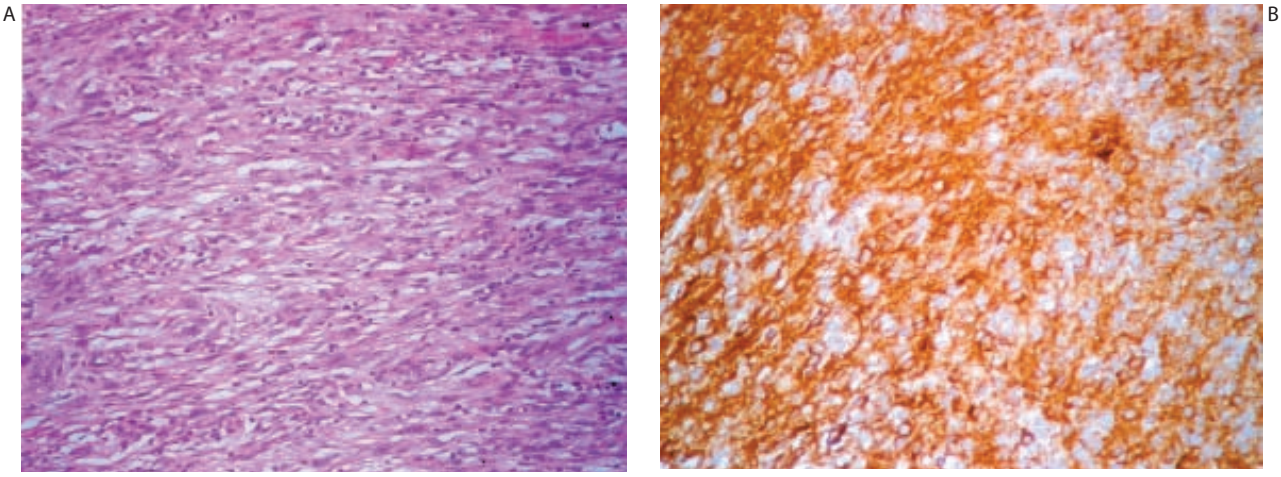

Figure 1. ***A:*** *Histologic examination reveals fascicles of smooth-muscle fibers with cigar-shaped nuclei (H&E, original magnification ×10).* ***B:*** *Immunohistochemistry demonstrates membrane staining of tumor cells with smooth-muscle actin (original magnification ×20).*

The patient underwent total laryngectomy, and his recovery was uneventful. He was fitted with an electrolarynx, which helped him maintain satisfactory communication. One year following surgery, he was well and exhibited no evidence of local or distant tumor.

Discussion

The morphologic diagnosis of laryngeal leiomyosarcoma can be difficult to make on the basis of conventional microscopy alone. Morphologic findings should be supported by immunohistochemistry and ultrastructural investigations whenever possible. Histopathologically, laryngeal leiomyosarcoma is characterized by prominent interlacing bundles and fascicles of spindle-shaped cells with cigar-shaped, blunt-ended nuclei and eosinophilic cytoplasm. On immunohistochemical staining, leiomyosarcoma is positive for smooth-muscle actin and negative for S-100 protein and desmin.[4] Immunohistochemical analysis is necessary to differentiate a leiomyosarcoma from other spindle cell tumors, such as spindle cell carcinoma, schwannoma, leiomyoma, and fibrosarcoma.

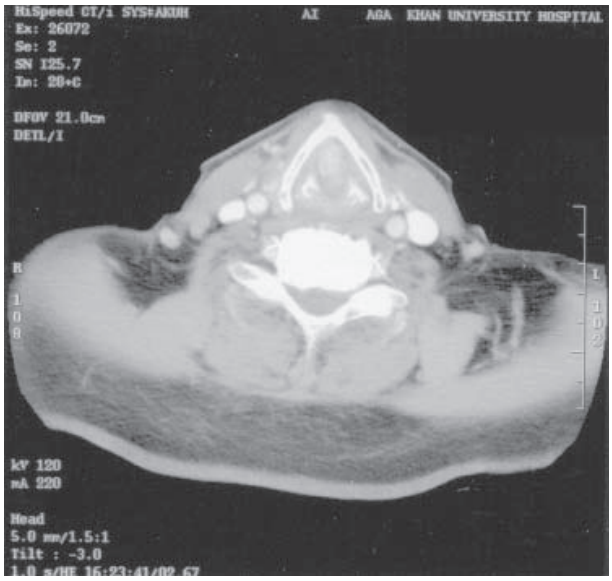

Figure 2. Preoperative postcontrast axial CT at the level of the glottis shows that the enhancing mass occupies the entire glottis. No destruction of the thyroid cartilage is evident.

Mitotic activity is the primary marker for a diagnosis of leiomyosarcoma, but cellular atypia is also taken into the account. Neoplasms with 5 or more mitoses per 10 high-power field and moderate to marked cellular atypia are considered to represent leiomyosarcoma.[5] There have been reports of leiomyosarcoma associated with Epstein-Barr virus in immunosuppressed patients.[8] In determining the extent of these tumors and when planning surgery, CT and magnetic resonance imaging are useful because they not only indicate the extent of the primary lesion, they provide reliable information regarding cervical lymphadenopathy, as well.

The paucity of reports of laryngeal leiomyosarcoma in the literature has hindered efforts to reach valid conclusions about different modalities of treatment. However, one generally accepted treatment is curative resection of the tumor with wide surgical margins. Limited surgical experience in dealing with laryngeal leiomyosarcoma and the unpredictable behavior of this disease have prompted some head and neck surgeons to be aggressive and treat extensive lesions with total laryngectomy rather than with partial surgery. However, some recent reports suggest that endolaryngeal resection or partial laryngectomy results in better functional outcomes whenever the extent of the disease permits.[1] Radical neck dissection is usually withheld unless cervical lymphadenopathy is obvious. Reported

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