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Leiomyosarcoma of the Larynx Presenting as a Laryngopyocele

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Summary: An 87-year-old man had an 8-month history of hoarseness, respiratory distress, and dysphagia. Physical examination, including direct laryngoscopy, revealed a mass on the right anterolateral side of the neck and a submucosal mass of the supraglottic larynx. A contrast-enhanced CT scan showed a more superior cystic mass, a laryngopyocele resulting from a more inferior, solid-appearing and obstructing mass at the level of the true vocal cord. The obstructing mass was also entirely submucosal at direct laryngoscopy; however, a biopsy specimen revealed a malignant tumor. Subsequent total laryngectomy and pathologic review showed it to be a leiomyosarcoma.

Index term: Larynx, neoplasms

Cancers of the larynx represent 2% of all malignant neoplasms. Less than 1% of malignant laryngeal cancers are sarcomas or other tumors of mesenchymal origin. This report describes a submucosal leiomyosarcoma of the larynx that obstructed the laryngeal saccule and presented as a laryngopyocele.

Case Report

An 87-year-old man came to our institution with an 8-month history of progressive hoarseness, shortness of breath, dysphagia, and right-sided otalgia. A physical examination revealed a 3-cm mass in the anterior triangle of the upper part of the neck and a submucosal mass on the right side of the larynx. A computed tomographic (CT) study showed a rim-enhancing fluid-density mass on the right side of the paraglottic space, extending from the level of the false vocal cord through the thyrohyoid membrane into the upper part of the neck (Fig 1A). At the level of the true vocal cord the mass abruptly changed character, appearing solid at that level and on sections obtained more inferiorly (Fig 1B). The working diagnosis, made on the basis of the CT findings, was submucosal tumor, most likely squamous cell carcinoma, with a secondary laryngopyocele. This assumption must always be made when an apparently solid mass is noted at the opening of the appendix of the laryngeal ventricle (saccule).

Direct laryngoscopy showed a firm, submucosal lesion on the right side of the true vocal cord that extended across the anterior commissure and onto the anterior one third of the left side of the true vocal cord. The biopsy specimen showed mucosal ulceration and a submucosal tumor mass. The tumor was composed of spindle and multinucleated anaplastic giant cells (Fig 1C). Immunoperoxidase staining demonstrated muscle actin and vimentin in the tumor cells (Fig 1D). No keratin was present. This picture suggests a leiomyosarcoma. A laryngopyocele on the right side of the supraglottic region was drained of 10 to 15 mL of purulent material.

The following month, the patient underwent a total laryngectomy. Gross pathologic examination of the specimen showed a 1.6 × 1.5-cm firm, nodular mass extending from the right side of the false vocal cord into the right side of the laryngeal ventricle and to the right side of the true vocal cord and 2 mm across the midline. No cartilage involvement was noted. The histologic findings on examination of the laryngectomy specimen were similar to the biopsy findings. The tumor cells stained for muscle actin and smooth muscle actin but not for desmin, multiple keratins (AE 1/3, MAK 6, CAM 5.2), S-100 protein, KP1, or alpha$_1$-antitrypsin. The diagnosis of leiomyosarcoma was confirmed. The patient was disease free at the 30-month follow-up examination.

Discussion

A laryngocele by definition is an abnormal dilatation of the laryngeal saccule that is filled with air when its orifice is patent (1); when obstructed and infected, these structures are called *laryngopyoceles*. Approximately 25% of entirely submucosal laryngeal masses are caused by a laryngocele, and most do not result from an obstructing mass lesion. Nevertheless, the radiologist is obligated to exclude an obstruction of the saccular orifice by a neoplasm as the cause of a laryngocele or laryngopyocele (2). Even CT or magnetic resonance (MR) im-

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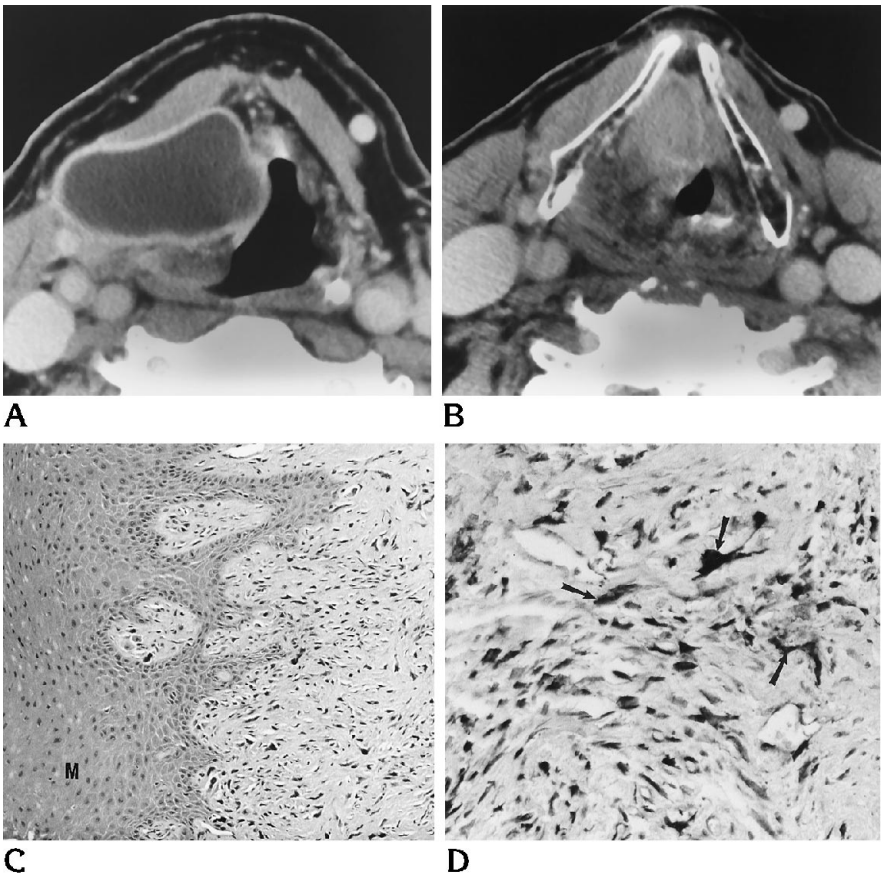

Fig 1. Eighty-seven-year-old man with 8-month history of hoarseness, respiratory distress, and dysphagia.

A, Axial CT scan shows a laryngocele extending from the paraglottic space through the thyrohyoid membrane to the deep portion of the neck.

B, Axial CT scan obtained about 6 to 9 mm inferior to that in *A* shows the solid soft-tissue mass, which proved to be a leiomyosarcoma, obstructing the saccule.

C, A low-power view of the original biopsy specimen shows hyperplastic but otherwise normal squamous mucosa at the edge (*M*). Beneath the mucosa is a tumor mass composed of spindled and large pleomorphic giant cells (hematoxylin-eosin stain, original magnification ×106).

D, A higher magnification of the tumor shows staining (*arrows*) of the spindled and giant cells for smooth muscle specific actin (original magnification ×210).

aging fails to show an obstructing lesion, the otolaryngologist is still obligated to look for one at the time of marsupialization and drainage of the laryngocele (2, 3).

In the larynx, submucosal tumors may arise from the laryngeal ventricle (eg, squamous cell carcinoma), minor salivary glands (eg, adenocystic carcinoma, adenocarcinoma, mucoepidermoid carcinoma), cartilage (eg, chondrosarcomas), and mesenchymal tissue (eg, fibrosarcoma, rhabdomyosarcoma, malignant fibrous histiocytoma, hemangiosarcomas, malignant nerve sheath tumors) (3).

Soft-tissue sarcomas are malignant tumors of extraskeletal connective tissue derived from cells of mesenchymal origin (4): 60% occur in the extremities, 30% occur in the trunk, and less than 10% occur in the head and neck region. Sarcomas account for less than 1% of all malignant head and neck lesions (5–7). They are more prominently included in the differential diagnosis when they are entirely submucosal, as in our case (2). The accurate histologic classification of spindle cell carcinomas was originally based in part on findings at electron microscopy (8), but it has been markedly facilitated by the advent of immunohistochemical markers. Specific markers are available for most tumors that fall into this group; spindle cell squamous carcinoma (keratins), neural tumors (S-100 protein), malignant fibrous histiocytoma ($alpha_1$-antitrypsin and $alpha_1$-antichrymotripsin), and angiosarcoma (factor VIII and ulex uropa agglutinin). In this patient, the differential diagnosis, made on the basis of histologic examination, was primarily between spindle cell squamous carcinoma and leiomyosarcoma. Immunohistochemical staining for muscle actin and smooth muscle actin was significant, and all other markers were nonreactive, indicating leiomyosarcoma (9, 10).

Sarcomas have a wide histologic spectrum and a broad range of malignant potential, which reflects their likelihood to recur locally and to metastasize. Head and neck sarcomas larger than 5 cm in diameter have a worse prognosis than do smaller tumors; this parallels the outlook for sarcomas arising in the retroperitoneum, pelvis, major blood vessels, or other peripheral soft tissues (11). The histologic type

and grade of the sarcoma and the presence of metastasis at the time of diagnosis also affect prognosis and treatment (6, 12, 13).

In an adult population, the mean age at which head and neck sarcomas occur is 48 years (6). Patients with leiomyosarcoma have a mean age of 54 years at presentation (6). There is no significant difference in occurrence rates between males and females (5, 13). The most commonly involved head and neck sites are the sinonasal region, the neck, the face, the scalp, and the orbit (12, 13). Predisposing factors include a history of radiation exposure, tuberous sclerosis, neurofibromatosis, Gardner syndrome, retinoblastoma, multiple basal cell carcinoma syndrome, Werner syndrome, or Turcot syndrome (5, 12). The histologic subtypes of head and neck sarcomas include fibrosarcomas, orbital or nonorbital rhabdomyosarcomas, neurofibrosarcoma and malignant schwannomas, angiosarcoma, leiomyosarcoma, hemangiopericytoma, liposarcoma, and synovial cell sarcoma (6, 12). In adults, the most common types are malignant fibrous histiocytoma, fibrosarcoma, esthesioneuroblastoma, angiosarcoma, and nonorbital rhabdomyosarcoma, each accounting for 10% of the soft-tissue sarcomas. Among children, 50% of cases are neuroblastomas and rhabdomyosarcomas, of which 36% are nonorbital and 15% are orbital rhabdomyosarcomas (13).

Tumors of mesenchymal origin encountered in the larynx include fibrosarcomas, chondrosarcomas, rhabdomyosarcomas, osteosarcomas, leiomyosarcomas, and hemangiosarcomas (2, 7, 11). We know of nine published cases of leiomyosarcomas (14, 15). Sarcomas, as well as other neoplasms affecting the larynx, usually cause progressive hoarseness or dysphagia and associated difficulty in swallowing. The causative mass will frequently be entirely submucosal (2). The reported duration of the symptoms may vary from 2 weeks to 2 years (6, 11). In our case, the symptoms were present for 8 months before the diagnosis was made.

Metastases are usually not noted initially. The most common sites of metastatic disease are the lung, liver, bone, brain, and skin (5, 6). Once the diagnosis is established, the radiologist, by means of CT or MR imaging, must show the size and local extent of the tumor and may provide adjunctive information about possible regional lymph node metastasis or spread along neurovascular bundles. The patient has a better prognosis if wide local excision can control a sarcoma under 5 cm in size and if there are no regional and distant metastases.

The treatment options include wide local excision of the tumor, radiation therapy, and chemotherapy, depending on the site, age, and type of sarcoma (5, 13). Surgery performed with tumor-free margins provides the best prognosis and survival rate. Neck dissection is not routinely performed for sarcomas of the head and neck (12, 13). In our case, the tumor-free margins of the excision made postoperative radiation therapy unnecessary, and the patient was disease free at the 30-month follow-up examination.

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