CASE REPORT

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ENDOSCOPIC CO2 LASER SURGERY FOR AN ATYPICAL CARCINOID TUMOR OF THE EPIGLOTTIS MASQUERADING AS A SUPRAGLOTTIC CYST

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Abstract: Background. Atypical carcinoid tumor is a neuroendocrine tumor; its occurrence in the larynx is uncommon, and clinical manifestations are rare. We report an unusual case of atypical carcinoid tumor of the epiglottis mimicking a supraglottic retention cyst.

Methods. A 44-year-old woman complained of an intermittent globus sensation of the throat of 2 years' duration. A 1.0- x 0.8-cm cystic lesion was found over the tip of the epiglottis. A supraglottic retention cyst was initially diagnosed, and the patient was treated medically. Her symptoms persisted, so we performed a laryngoscopic biopsy, which suggested an atypical carcinoid tumor.

Results. Transoral endoscopic CO2 laser surgery and bilateral elective neck dissection were subsequently performed. The 2-year follow-up did not reveal any locoregional recurrence or distant metastasis.

Conclusions. This aggressive neoplasm may cause only a few, unremarkable symptoms and masquerade as a supraglottic cyst. Endoscopic CO2 laser surgery can be used to resect this uncommon tumor, with oncologically sound results and without surgical morbidity. © 2005 Wiley Periodicals, Inc. Head Neck 27: 1004–1007, 2005

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Neuroendocrine tumors of the larynx are rare, accounting for only 0.6% of all laryngeal neoplasms.1 They consist of two major groups, carcinomas of epithelial origin and paragangliomas of neural origin, and include well-differentiated typical carcinoid tumors, moderately differentiated atypical carcinoid tumors, and undifferentiated small cell carcinomas. Paragangliomas are usually discussed separately, because they almost always behave benignly and originate...
from the paraganglion cells of the parasympathetic system.2

Among the laryngeal neuroendocrine carcinomas, atypical carcinoid tumors are a relatively frequent nonsquamous carcinoma of the larynx. Most occur in the supraglottic larynx, with a predilection for the aryepiglottic fold, the arytenoid, and the laryngeal aspect of the epiglottis.3 They have a male preponderance, are associated with a history of heavy smoking, and have a peak incidence between the sixth and seventh decades of life.3 Atypical carcinoid tumors are aggressive neoplasms with a high risk for widespread metastasis. They seem to be unresponsive to chemotherapy, and the role of radiotherapy alone has been regarded as controversial.4–6 Therefore, wide surgical excision should be the treatment of choice. To date, only a few reports have described surgical approaches, including partial laryngectomy for limited lesions and total laryngectomy for advanced tumors or as postirradiation salvage management.4,7 Because of its rare occurrence, a consensus for a surgical rationale seems impractical.

We present the clinical and pathologic findings in an unusual case of epiglottic atypical carcinoid tumor masquerading as a supraglottic cyst. The patient was a middle-aged woman who never smoked and rarely consumed alcohol.

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An otherwise healthy 44-year-old woman was initially seen with a progressively worsening, intermittent globus sensation of the throat of 2 years’ duration. She did not complain of dysphagia or odynophagia. No significant personal or family history of malignancy was noted. The patient had never smoked and rarely consumed alcohol. Flexible transnasal laryngoscopy revealed a 1.0-×0.8-cm, reddish, smooth, and cystic subepithelial lesion over the tip of the epiglottis (Figure 1). No metastatic cervical lymphadenopathy was found. An epiglottic retention cyst was initially diagnosed, and the patient was treated medically. However, because of her persistent discomfort, she underwent a transoral laryngoscopic biopsy under general anesthesia. The results suggested an atypical carcinoid tumor. Subsequently, CT revealed no abnormalities in the head and neck regions, and systemic preoperative workups, including routine chest x-ray, bone scan, and liver ultrasonography, were all negative for distant metastasis.

Three weeks after the biopsy, we resected the epiglottic tumor by transoral endoscopic CO₂ laser surgery. Considering the high frequency of occult cervical metastasis, bilateral prophylactic neck dissection (zone II, III, and IV) was also performed at the same time. The surgical specimen from the neck dissection showed no cervical metastasis bilaterally. The patient’s postoperative course was uneventful, and she had no surgical morbidity. She was discharged 1 week after surgery. No postoperative radiotherapy was ad-
The symptoms caused by laryngeal atypical carcinoid tumors usually depend on the site of the lesion. Carcinoid syndrome, such as flushing, diarrhea, and hypertension has been infrequently reported.4 Some patients have complained of symptoms of “glossopharyngeal neuralgia” before their tumors have become clinically apparent. The tumor usually manifests clinically as a nodular, polypoid, pedunculated subepithelial lesion, with or without ulceration of the overlying surface epithelium.3 However, these symptoms and signs are usually too ambiguous to support a correct clinical diagnosis. Consequently, delayed diagnosis or misdiagnosis for laryngeal atypical carcinoid tumors can easily occur.

Pathologic classification is crucial for correct management of laryngeal neuroendocrine carcinomas, because the behavior of neuroendocrine neoplasms varies by subtype. Typical carcinoid tumors have the best prognosis and seldom metastasize; conservative surgical excision with regular follow-up is usually sufficient. Elective neck dissection is not necessary if no lymph node metastasis is found at diagnosis.

In contrast to typical carcinoid tumors, small cell neuroendocrine carcinomas are the most lethal neuroendocrine carcinomas of the larynx, with a 5-year survival rate of only 5%.8 Because radical surgical procedures fail in almost every reported case, therapeutic radiotherapy of the larynx and neck with accompanying systemic chemotherapy is the currently accepted method of treatment.7

Atypical carcinoid tumors, on the other hand, have a regional lymph node metastasis rate of approximately 43% and a distant metastasis rate of approximately 45%. After curative management of atypical carcinoid tumors, the 5-year survival rate is 48%, and the 10-year survival rate is only 30%.9 The size of the atypical carcinoid tumor is an important prognostic factor, with primary tumors greater than 1 cm demonstrating twice the mortality rate of those ≤1 cm (44% vs 23%).6,9,10 Because chemotherapy is of no value in managing atypical carcinoid tumors, except in cases of metastatic disease, and because the therapeutic effect of radiotherapy is also questionable, surgical resection should be advocated for patients with primary atypical carcinoid tumors, with accompanying elective neck dissection if the cervical lymph nodes are clinically negative at diagnosis. Radical or modified radical neck dissection should be performed in cases of positive cervical lymph nodes.5–7 To the best of
our knowledge, only a few reports have described surgical approaches, including partial laryngectomy for limited lesions or as postirradiation salvage management.4,7 Under the consensus that conservative surgery with larynx preservation should always be considered as long as adequate tumor resection can be ensured, endoscopic CO$_2$ laser surgery, with its growing popularity, provides a wonderful alternative approach.

Microscopically, typical and atypical carcinoid tumors are composed of nests of uniformly organized, round cells, whereas small cell carcinomas have sheets or ribbons of small, hyperchromatic, pleomorphic cells with a high nuclear-to-cytoplasmic ratio, frequent mitoses, and necrosis.4 The tumor cells of atypical carcinoid tumors may be variably arranged and may have an organoid growth pattern or may grow in nests, ribbons, trabeculae, cords, acini, or a pseudoglandular pattern. Mitosis and nuclear pleomorphism are more frequent in atypical carcinoid tumors than in typical carcinoid tumors.3 Because cells may be arranged in a pseudo-Zellballen pattern, atypical carcinoid tumors have also been known to mimic paragangliomas, which are benign tumors that are treated conservatively. Immunohistochemical studies have proved to be very helpful in differentiating atypical carcinoid tumors from paragangliomas.3

The supraglottis is the most common site of laryngeal cysts, which occurs most frequently where mucus-secreting glands are abundant. In general, mucous retention cysts of the supraglottis are usually found incidentally on routine examination. In most cases, they are asymptomatic, <1 cm in diameter, and superficial within the mucous membrane. Infrequently, mucous retention cysts are large enough to produce cough and stridor, warranting surgical management. In our opinion, cystic-appearing lesions of the supraglottis that cause throat pain (such as “glossopharyngeal neuralgia”), show a polypoid or nodular contour subepithelially, and/or enlarge progressively under regular follow-up may warrant endoscopic biopsy to differentiate the common mucous retention cysts from unusual benign-appearing neoplasms.

The case presented here is somewhat different from the usual clinical scenario of a supraglottic atypical carcinoid tumor, because our patient was a middle-aged woman with no history of smoking. The only symptom she had was just an intermittent globus sensation of the throat. The tumor presented as a small epiglottic cystic lesion, which was easily ignored by clinicians and was misdiagnosed as a supraglottic retention cyst. We present the clinicopathologic manifestations of this rare case to remind clinicians that sufficient attention and subsequent laryngoscopic biopsy are needed for imminent management of this aggressive, but clinically indolent, malignancy. In addition, endoscopic CO$_2$ laser surgery can be used to resect this neoplasm, with oncologically sound results and without surgical morbidity.

REFERENCES