Adenoidcystic Carcinoma of the Larynx

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A case of adenoid cystic carcinoma of the larynx is presented. It was managed by wide field laryngectomy and neck dissection.

Adenoidcystic carcinoma is an uncommon epithelial tumor of serous and mucous glands. The histopathology of the tumor was first described in 1859 by Billroth who gave it the original designation of "cylindroma." Since then, it has been called by a variety of names. More recently, Foote and Frazell expressed preference for the name adenoidcystic carcinoma and credited the term to the late James Ewing.

Adenoidcystic carcinoma of the larynx is rare. Cady et al, in a review of 2,500 laryngeal cancers, reported five cases. Rosenfeld et al reviewed 184 malignant tumors of salivary gland origin and found only three cases of adenoid cystic carcinoma involving the larynx. Adams and Duvall found one laryngeal cylindroma in a review of 792 head and neck malignancies. Toomey presented a detailed and comprehensive review of adenocarcinoma of the larynx. Of 37 reported lesions, he found 17 cylindromas, 4 epitheliomas, 1 basal cell carcinoma and 1 other neoplasm of minor salivary gland origin. He concluded that the latter six cases may also have been of the cylindroma type. Recently, two more cases of adenoidcystic carcinoma of the larynx have been reported. Leafsted et al reviewed 81 adenoid cystic tumors and found one involving the larynx. Baxley and Farmer reported another laryngeal primary in their series of 22 carcinomas of the head and neck.

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Because of the earlier confusion with terminology, it is difficult to enumerate the exact number of reported cases of adenoidcystic carcinoma of the larynx. We estimate this number to be no more than 50 such tumors. We add here one more case of the tumor involving the larynx.

Case Report

A 47-year-old Caucasian woman reported a five-month history of discomfort in the throat. This was initially noticed as easy fatigue of the voice, and, later, a "rawness" in the throat associated with intermittent sharp, stabbing pain. The patient was treated for "laryngitis" by her local physician. However, as the symptoms progressed, the patient was referred to an otolaryngologist who noted a lesion of the left aryepiglottic fold. Tissue taken for biopsy was reported as adenoidcystic carcinoma. The patient was then referred to Henry Ford Hospital for further treatment.

Direct laryngoscopy revealed a bulky tumor originating from the left aryepiglottic fold. It involved the left arytenoid, posterior half of the left false cord, and extended across the midline to involve the posterior surface of the right arytenoid. The left vocal cord was fixed and the mobility of the right vocal cord somewhat impaired, so that the patient experienced moderate dyspnea after the examination. There was a tender, mobile lymph node, 1 cm in size, on the thyrohyoid membrane in the neck on the left side. The remainder of the head and neck and other general examination were unremarkable.

Laboratory studies including hemoglobin, WBC, FTA-ABS, alkaline phosphatase and urinalysis showed normal results. Findings of a chest x-ray, liver scan, and metastatic bone sur-
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Figure 2
Shows an intact squamous epithelium with tumor present in the deeper layers of the section.

Pathology

Gross examination of the specimen showed a smooth, non-ulcerated 2 x 3 cm mass occupying the region of the left arytenoid and the left aryepiglottic fold. When the larynx was opened, we noted that the right vocal cord, the anterior two-thirds of the left vocal cord, the ventricles and subglottic region were free of obvious involvement (Figure 1). Microscopic sections of the mass showed an intact stratified squamous epithelium with tumor present in the deeper layers of the larynx (Figure 2). The appearances were of a moderately well-differentiated, unencapsulated adenocarcinoma. Low power views showed a varying glandular pattern. In some areas, the tumor had a loose, trabecular cribriform structure; in others, it demonstrated a more compact and dense form. Figure 3 demonstrates the typical cystic spaces and the cordlike pattern. Figure 4 shows a more compact stroma. Individual cells are small with scant cytoplasm. No mitotic figures are seen. In some areas characteristic perineural tumor infiltration can be seen (Figure 5).

Comment

It is worthy of note that a painless mass or swelling is the most common complaint of patients with adenoidcystic carcinoma in the head and neck. Other signs or
Figure 3
Shows the classical "cylindromatous" pattern with cystic spaces and cords of cells.

Figure 4
Demonstrates a more compact dense glandular stroma.
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Figure 5
Shows several nerve bundles with perineural tumor infiltration.

Symptoms of tumor in this region may be present from 1 to 5 years before the patient is first seen by a physician. In patients with laryngeal involvement, however, the most likely presenting symptoms are hoarseness and dyspnea. This probably accounts for the relatively short interval in our case between the onset of symptoms and definitive treatment. Local pain, a complaint considered to be quite characteristic of this tumor, is reported in as many as 50% of cases and as few as 11%. The lymph node metastasis, clinically evident in our patient, was confirmed by histopathological examination.

The course of the disease is characterized by a high incidence of local recurrence and ultimate distant metastases in spite of aggressive surgical management. Distant metastases are found most commonly in the lungs and bone. The five-year survival of patients with laryngeal adenoid cystic carcinoma is reported as 28.6% by Toomey. This is comparable to 26.8% in recent studies of 54 patients with adenoidcystic carcinoma of the head and neck and no laryngeal involvement. However, five-year survivals do not accurately reflect the ultimate prognosis. Because of the relentless and insidious behavior of this tumor, death may occur 10 or 20 years after the initial diagnosis.

The treatment of choice is radical surgical excision. Radiation therapy is useful as a palliative measure to produce relief of pain and tumor regression in advanced cases.
References


