CLINICAL VIGNETTE

Basaloid Squamous Cell Carcinoma of the Upper Aerodigestive Tract: A Case Report and Review of the Literature.

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Introduction

Basaloid squamous cell carcinoma (BSCC) is a rare variant of squamous cell carcinoma. Since its classification as a distinct disease in 1986, approximately 200 cases have been reported.[1-3] BSCC is believed to arise from either stem cells in the basal layer of the surface epithelium, or from salivary duct lining epithelium.[4] BSCC primarily affects the head and neck, including the palate, buccal cavity, floor of mouth, tonsils, gingiva, nasopharynx, trachea, sinonasal tract, esophagus, and thymus.[1-3] It has also been reported to involve the lung, anus, and cervix.[2] Most commonly, however, it is found in the supraglottic region.[1]

By the time a patient becomes symptomatic, the disease is often at an advanced stage when the tumor has already metastasized.[2,3] This aggressive behavior, combined with the low incidence of the disease (a retrospective review of 468 surgical specimens of laryngectomy yielded only 7 cases of BSCC, or 0.66%), makes it a formidable diagnostic challenge.[5] The goals of this article are threefold: to report a case of BSCC presenting as a tonsillar primary cancer, to inform practitioners how to recognize the characteristics of BSCC, and to discuss the factors which put patients at risk for this disease.

Case Report

A 65-year-old male presented to primary care clinic complaining of rapid onset of a small, painless mass on the right side of his neck over a 2-day period. He denied any fevers or chills, night sweats, weight loss, difficulty swallowing, changes in his voice, or tooth pain. There were no recent local traumas, dental procedures, insect bites, or prior neck irradiation, which might otherwise have explained the presence of the mass. The patient's family history was negative for head and neck cancers. However, the patient admitted to a 30 pack/year history of smoking, before quitting 20 years ago. He also reported drinking 2 beers every day.

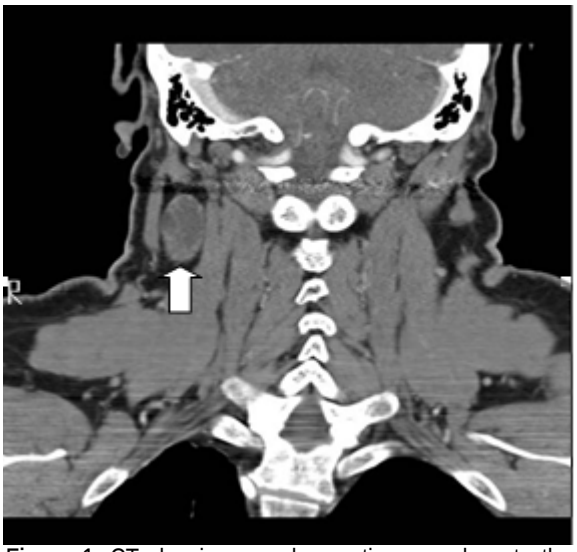

Figure 1. CT showing complex cystic mass deep to the right sternocleidomastoid muscle.

On physical examination the patient was afebrile. His tympanic membranes were translucent and intact. There was no tenderness to palpation over his frontal and maxillary sinuses. His dentition was excellent, and he had no abscesses, erythema, or exudates in his oropharynx. Examination of his neck revealed a 2 cm x 4.5 cm firm, non-tender nodule in the right infra-auricular region, with no changes to the overlying skin. No other lymphadenopathy was appreciated. A computed tomography scan (CT) of the neck revealed this to be an oval-shaped, rim-enhancing complex cystic mass immediately deep to the right sternocleidomastoid muscle.

Cytology from a fine needle aspiration of the mass was interpreted as poorly differentiated cystic squamous cell carcinoma. A subsequent Positron Emission Tomography (PET) scan demonstrated mild hypermetabolic activity in the right internal jugular node as well as the pharyngeal tonsils.

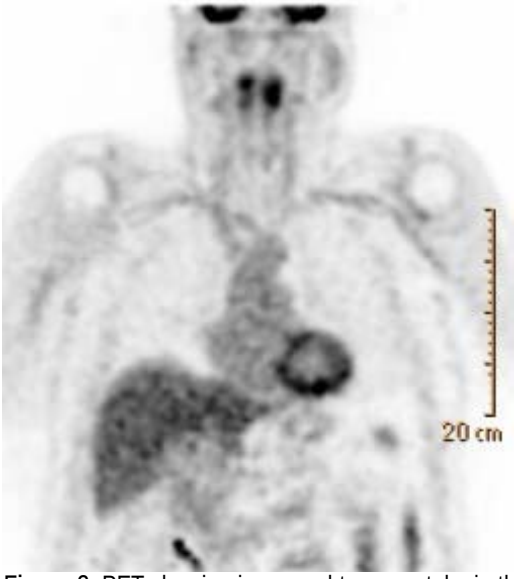

Figure 2. PET showing increased tracer uptake in the tonsils.

The patient underwent bilateral tonsillectomy; histology findings confirmed infiltrating high-grade squamous cell carcinoma of the right tonsil (see below). Based on these findings, the cancer was deemed to be T1N2aM0, or stage IVa. He declined further surgical intervention and opted for a combined regimen of chemotherapy and radiation.

Discussion

Basaloid squamous cell carcinoma is a histologically distinct and highly aggressive malignant tumor. Definitive diagnosis requires a tissue specimen. The pathological features of BSCC include nuclear pleomorphism, hyperchromasia, mitotic activity, and necrosis, which all mark it as a high-grade malignancy.[1] The differential diagnosis of BSCC includes adenoid cystic carcinoma, submucosal salivary gland neoplasm, and small-cell carcinoma, which can be distinguished through the use of immunohistochemical stains.[3]

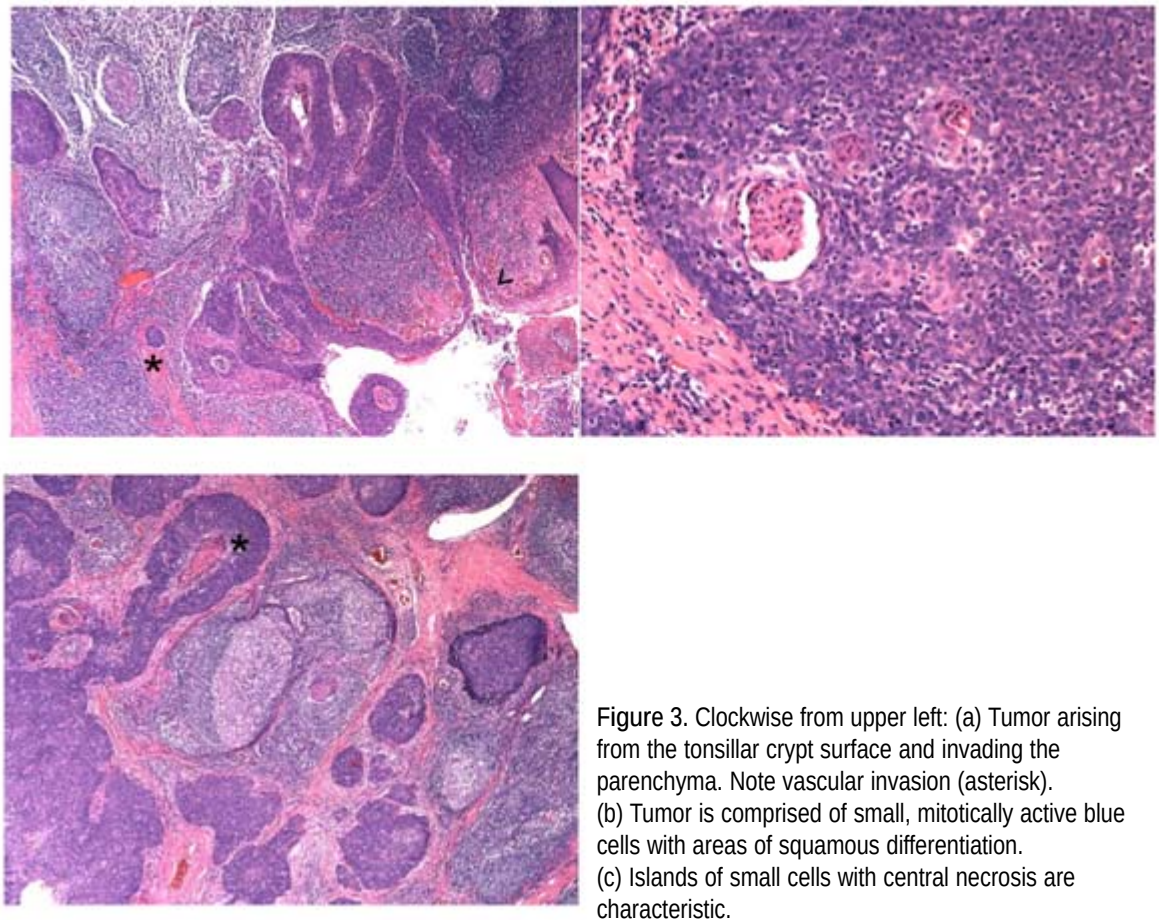

Figure 3. Clockwise from upper left: (a) Tumor arising from the tonsillar crypt surface and invading the parenchyma. Note vascular invasion (asterisk).
(b) Tumor is comprised of small, mitotically active blue cells with areas of squamous differentiation.
(c) Islands of small cells with central necrosis are characteristic.

This clinical case illustrates the presentation of BSCC as a tonsillar primary cancer. In this patient, the cystic component of the tumor likely accounted for its rapid increase in size. He had several of the known risk factors for developing BSCC: male gender, age older than 60 years, and a history of tobacco and alcohol use.[1,4] Prior radiation exposure has also been implicated in the development of BSCC.[1] Infection with the Epstein-Barr virus or human papilloma virus has been suggested as a contributory factor, but this remains unproven.[1] In addition to new onset of a mass, presenting symptoms reported in the literature include bleeding, pain, and hoarseness.[2,4]

In summary, basaloid squamous cell carcinoma is a highly aggressive malignant tumor that occurs primarily in the upper aerodigestive tract. Although a rare diagnosis, BSCC should be suspected in male patients who are older than 60 years, presenting with a head or neck mass, bleeding, pain, or hoarseness, and have a history of tobacco or alcohol abuse.

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