What is Adenoid cystic carcinoma?
Adenoid cystic carcinoma (ACC) is an uncommon form of malignant neoplasm that arises within secretory glands, most commonly the major and minor salivary glands of the head and neck. Other sites of origin include the trachea, lacrimal gland, breast, skin, and vulva. This neoplasm is defined by its distinctive histologic appearance.

Differential Diagnosis
The differential diagnosis is largely that of other benign and malignant neoplasms that arise in these locations. In the salivary glands these include benign mixed tumor, mucoepidermoid carcinoma and polymorphous low-grade adenocarcinoma (PLGA). The major histologic differential diagnosis in the minor salivary glands is between ACC and PLGA, which share many features. In other sites, ACC may be confused on histologic grounds with a variant form of squamous carcinoma known as basaloid squamous carcinoma.

Signs & Symptoms
These depend largely on the site of origin of the tumor. Early lesions of the salivary glands present as painless masses of the mouth or face, usually growing slowly. Advanced tumors may present with pain and/or nerve paralysis, for this neoplasm has a propensity to invade peripheral nerves. Tumors of the lacrimal gland may present as proptosis and changes in vision. ACC arising in the tracheobronchial tree may present with respiratory symptoms, while tumors arising in the larynx may lead to changes in speech.

Clinical Course
Due to its slow growth, ACC has a relatively indolent but relentless course. Unlike most carcinomas, most patients with ACC survive for 5 years, only to have tumors recur and progress. In a recent study of a cohort of 160 ACC patients, disease specific survival was 89% at 5 years but only 40% at 15 years. Another unusual feature of ACC is that, unlike most carcinomas, it seldom metastasizes to regional lymph nodes. Distant metastasis is the most common presentation of treatment failure. The lung is by far the most common site of metastasis, with the liver being the second most common site. Bone metastases usually indicate a fulminant clinical course. Poor prognostic signs at the time of initial surgery are a solid growth pattern, perineural invasion of major nerves and/or positive margins after histopathologic examination.

Etiology/Epidemiology
A wide age range has been reported for adenoid cystic carcinoma, including cases in the pediatric age group. Most individuals are diagnosed with the disease in the fourth through sixth decades of life. There is a slight female preponderance (female to male ratio approximately 3 to 2). No strong genetic or environmental risk factors have been identified. Damage to the DNA genome occurs in the development of ACC, as it does in all cancers studied to date. Various studies have shown chromosomal abnormalities and genetic deletions occurring in samples of ACC. There is some evidence that the p53 tumor suppressor gene is inactivated in advanced and aggressive forms of this neoplasm. Otherwise the specific molecular abnormalities that underlie this disease process are unknown.

Diagnosis
The diagnosis is made by histologic analysis of a biopsy or resection specimen of a tumor mass. There are three major variant histologic growth patterns of ACC: cribriform, tubular and solid. The solid pattern is associated with a more aggressive disease course. There are no serum markers for this neoplasm. Recurrences are usually identified by radiographic imaging techniques, such as computed tomography.

Treatment
Standard Therapy: Surgical resection, whenever possible, is the mainstay therapy. Based on clinical experience, many centers advocate postoperative radiotherapy to help limit local failure. A few specialized centers offer neutron beam therapy which may be more effective than

conventional radiation therapy. There appears to be no effective chemotherapy for metastatic and/or unresectable ACC, although some patients may receive palliation. Investigational Therapy: Several clinical trials are examining the effects of relatively new chemotherapeutic drugs (paclitaxel, gemcitabine, etc.) alone, or in combination with other drugs, in the control of metastatic or locally recurrent ACC.

References

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