Adenoid Cystic Carcinoma  
(Cylindroma)

This malignant tumor is poorly encapsulated and while seemingly well defined within the affected gland, there is usually infiltration of surrounding tissue on closer examination. The cut surface is firm and there are not the myxomatous areas seen in pleomorphic adenoma or the cysts seen in Warthin’s tumor.

Histologically, the tumor classically shows a “Swiss cheese” appearance with cribriform (sieve-like) growth. Other areas of the same tumor may show nests or cords or sheets of cells in various combinations. Perineural involvement is the rule. Encapsulation usually is incomplete. There may be minimal nuclear pleomorphism and no mitotic activity so that the overall rather bland microscopic appearance belies the clinical course of this highly malignant neoplasm.

Adenoid cystic carcinoma; cribriform or “Swiss cheese” pattern that is unmistakable but not always present. The cells show scanty cytoplasm and round to ovoid basophilic nuclei of regular form. The circular areas containing weakly eosinophilic amorphous basement membrane material accounting for the synonym “cylindroma.” Mucous and serous glands and the ducts of normal submandibular salivary gland are seen on the left. The gland was not invaded.
Adenoid cystic carcinoma with perineural invasion. There are clumps of tumor cells (arrows), some with cribriform pattern. Neural invasion by adenoid cystic carcinoma is so common that it is expected and is thought to contribute to the poor prognosis. Triangle indicates neural tissue.

High power photo of the tissue seen. The spaces are said to be attempts at duct formation. Note the relative lack of pleomorphism. There are no mitoses.
Adenoid cystic carcinoma. Additional example of neurotropism with islands of tumor cells surrounding and invading neural bundles (arrows).

Adenoid cystic carcinoma, high power. Tiny island of adenoid cystic carcinoma in nerve fibers (triangle). While little nuclear pleomorphism is present in most adenoid cystic tumors, some is seen here. Nuclei stain dark (arrow) and there is minimal cytoplasm.
Adenoid cystic carcinoma, invasion of adventitia of medium-sized artery (arrow). The media and intima are not involved.

Adenoid cystic carcinoma, cords of cells and individual cells separated by mucoid material; high grade tumor. Cross-section of nerve at top (arrow).
Adenoid cystic carcinoma, infiltrating bone of maxillary sinus.

Adenoid cystic carcinoma. Formation of hyaline or mucoid material may be extensive, breaking up the tumor cells (arrow) into strands or chains.
Adenoid cystic carcinoma, parotid, capsular invasion. A fragment of parotid is present in one corner (large arrow) with tumor (double arrows) invading the thick fibrous parotid capsule. Adenoid cystic carcinomas are only poorly encapsulated at best.

Adenoid cystic carcinoma, external auditory canal, outer one-half. Triangle indicates wall of external auditory canal; small double arrows, squamous epithelium; large single arrows, adenoid cystic carcinoma in small nests. Tumor apparently arose in this site.
Adenoid cystic carcinoma. FNAB of a parotid mass. Spherical balls of metachromatic staining stroma (which represents reduplicated basement membrane material) are surrounded by small cells with very high nuclear cytoplasmic (N/C) ratios. This pattern faithfully reproduces the cribriform pattern (also called cylindromatous pattern in the older literature) that one sees in tissue sections. Diff-Quik stain.

**Clinical Aspects:**

Diagnosis may be verified in some cases by FNA. The presence of adenoid cystic carcinoma is suspected in patients with a painful tumor, especially one that is ill-defined. Facial nerve paralysis is more common with adenoid cystic carcinoma than with other tumors of the parotid and is an unfavorable sign.

Treatment has varied over the years and now radical resection of the neoplasm with a generous cuff of normal tissue is favored even if sacrifice of the facial nerve or bones of the face or skull is required. Postoperative radiotherapy may be of some benefit.

This tumor accounts for about fifteen percent of all malignant salivary gland neoplasms and involves chiefly the parotid and submandibular salivary glands, but also affects the minor salivary glands and the ceruminous and lacrimal glands. The disease is extremely difficult to cure. A patient may live five to twenty years after apparently successful therapy only to have a local recurrence or lung metastasis.