Trichoepithelioma occurs either in multiple lesions or as a solitary lesion. The name trichoepithelioma is preferable to other designations, such as epithelioma adenoides cysticum and multiple benign cystic epithelioma, because it is more indicative that the differentiation of this tumor is toward hair structures.

Multiple trichoepitheliomas are transmitted as an autosomal dominant trait (53). In most instances, the first lesions appear in childhood and gradually increase in number (54). Numerous rounded, skin-colored, firm papules and nodules usually between 2 and 8 mm in diameter are seen located mainly in the nasolabial folds, but also the nose, forehead, and upper lip. The association of trichoepithelioma and cylindroma, the latter of which is also dominantly inherited, has been observed repeatedly.
Solitary trichoepithelioma occurs more commonly than multiple trichoepitheliomas. It is not inherited and consists of a firm, elevated, flesh-colored nodule usually less than 2 cm in diameter. Its onset usually is in childhood or early adult life. Most commonly, the lesion is seen on the face, although it can occur on the scalp or elsewhere. The presence within the same tumor of a solitary trichoepithelioma and an apocrine adenoma has been described.
Giant solitary trichoepithelioma, measuring several centimeters in diameter, is a distinct variant of trichoepithelioma.
Histopathology

As a rule, multiple trichoepitheliomas are superficial dermal lesions. They appear...
The fibroblasts encircle and are tightly associated with the basaloid islands, lacking the retraction artifact.
Additional findings, observed in some but not all trichoepitheliomas, are the presence of a foreign-body giant cell reaction, foreign-body giant cells, and multinucleate cells, which may or may not be in association with horn cysts. Calcium deposits may be noted, either within or without the cysts.

Occasionally, some lesions in patients with multiple trichoepitheliomas show relatively little differentiation toward hair structures. Such lesions can be difficult to distinguish from those of keratotic basal cell carcinoma. Thus, on a histologic basis, it may be difficult definitively to distinguish between multiple trichoepitheliomas and basal cell carcinoma (see Differential Diagnosis).

Solitary trichoepithelioma often has a high degree of differentiation toward hair structures. Solitary lesions commonly are firm or hard, less likely than multiple lesions to ulcerate or hemorrhage, and may more frequently occur on the trunk and extremities.
Additional Studies. It is assumed that the basophilic cells surrounding horn cysts are similar to hair matrix cells and that the horn cysts are similar to the nucleated cells seen in normal hair shafts at the keratogenous zone.

Histochemical staining with the Gomori stain for alkaline phosphatase has shown positive staining in many invaginations of the horn cysts.
The putative gene for multiple familial trichoepitheliomas has been localized to chromosome 9p21. Several known tumor suppressor genes, including p15, p16, and p19, have been assigned to this region. However, loss of heterozygosity on chromosome 9p21 has not been found in sporadic tumors. In contrast, deletions causing overexpression of the human homologue of the Drosophila patched gene (Ptch) have been found in trichoepitheliomas as in basal cell carcinoma. A large body of recent work has demonstrated that mutations in the CYLD2 gene, which appears to be located on chromosome 9q23, have been found in trichoepitheliomas.
to function as an ubiquitin-specific protease, are present in some cases
Differential Diagnosis

The difficulty of differentiating multiple trichoepithelioma...
transmission. In addition, certain histologic features, as well as immunohistochemical stains, can assist in
The differentiation of multiple trichoepitheliomas from the nevoid basal cell carcinoma syndrome on histologic grounds...