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. In most instances, the first lesions appear in childhood and gradually increase in number. 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 lesions resemble the appearance of pinhead or millet seeds and are often elevated above the skin. The lesions are hard, non-tender, and slightly domed. Occasionally, a single lesion is found, which resembles a nodular basal cell carcinoma.

A histologic examination of the lesion reveals a tumor composed of five different elements that simulate the normal epidermis: epidermis, dermis, subcutaneous tissue, hair follicles, and sebaceous glands. The lesions are also characterized by a proliferation of abnormal sebaceous glands and hair follicles, which are seen in the dermis.

Trichoepithelioma is usually a benign condition, but it can sometimes be mistaken for other more serious conditions, such as basal cell carcinoma or sebaceous cyst. It is important to differentiate between these conditions to ensure appropriate treatment.
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, ... 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, noted as a focus of histiocytic hyperplasia, fibrosis, and calcification. Amyloid deposition within the lesion is also encountered. 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 trichoepitheliomas often have a high degree of differentiation toward hair structures. Solitary lesions
Additional Studies. It is assumed that the basophilic cells surrounding horn cysts are similar to hair matrix cells and that the horn cysts are a primary site for hair development. Histochemical staining with the Gomori stain for alkaline phosphatase has shown positive staining in many invaginations of the horn cysts, suggesting that these structures are immature hair structures with abrupt development of the horn cells from hair matrix cells.
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 cases of trichoepitheliomas. In addition, 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 involved in the development of trichoepitheliomas, have been found on chromosome 9q23.
The close relationship between trichoepithelioma and basal cell carcinoma has been explained on the basis of the patched gene mutations seen in both tumors. Because cells of various degrees of maturity may occur in the same lesion, trichoepithelioma may have areas consistent with the histologic picture of basal cell carcinoma and vice versa.
Differential Diagnosis

The difficulty of differentiating multiple trichoepitheliomas...
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...