Trichoepithelioma Papulosum Multiplex
Trichoepithelioma

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. In rare instances, lesions are located in the axillas or perineum. A generalized distribution has been observed in a few cases. In rare cases, multiple trichoepitheliomas, desmoids, and adenomas may occur together in a single individual, and in rare cases, an association with cylindromas has been observed (55).
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, scalp, neck, or upper chest. 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.
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. Occasionally, some lesions in patients with multiple trichoepitheliomas show relatively little differentiation toward hair structures, resembling basal cell carcinoma, which may also show horn cysts. 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...
**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. This suggests that trichoepithelioma represent 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. 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
The close relationship between trichoepithelioma and basal cell carcinoma has been explained by the presence of similar genetic alterations. Specifically, mutations in the patched gene, which is known to function as an ubiquitin-specific protease, are present in some cases. This gene is crucial for the development of hair structures and is normally present in the outer root sheath cells of the hair follicle. Mutations in patched may allow for the development of hair-like structures in the affected skin, leading to the formation of trichoepitheliomas. Additionally, 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...
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