Trichoepithelioma Papulosum Multiplex = ﻣﺘﻌدﺪ ﺣﻄاﻄﻲ ﺷﻌرﻲ ﻇﻬارﻲ ورﻢ

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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 coexistence 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, scalp, or upper chest. In multiple trichoepitheliomas, usually more than 20 lesions are present. 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, horn cysts, and, less commonly, amyloid. 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 a keratotic 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 arise from the hair matrix cells. With initial keratinization, 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 trichoepithelioma representing 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 forms of the disease. In addition, deletions causing overexpression of the human homologue of the *Drosophila* patched gene (Ptch) have been found in trichoepitheliomas as well 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 regulation of the Wnt signaling pathway, are associated with a subset of familial and sporadic cases of trichoepithelioma.
to function as an ubiquitin-specific protease, are present in some cases.
Differential Diagnosis

The difficulty of differentiating multiple trichoepitheliomas from keratotic basal cell carcinoma on histologic grounds...
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...