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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. 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 on the nose, forehead, and upper lip. In rare cases, tumors have been described on the scalp. The lesions may be single or multiple. The lesional tract usually shows adnexal differentiation with the presence of hair follicles, eccrine glands, and sebaceous glands. The histologic features of trichoepithelioma and cylindroma, the latter of which is also dominantly inherited, have 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, but it can occur on any other skin surface. 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, cysts, calcium deposits either within the foci of the foreign-body reaction or within intact horn cysts, and amyloid.

Occasionally, some lesions in patients with multiple trichoepitheliomas show relatively little differentiation toward hair or skin structures, resembling basal cell carcinoma. 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 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 trichoepithelioma representing immature hair structures, with abrupt development of 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 to be involved in the regulation of the NF-κB pathway, is frequently found in sporadic cases.
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 is a significant challenge. Clinical data, such as the number and distribution of the lesions and the presence of hereditary factors, can help in the differential diagnosis.
transmission. In addition, certain histologic features, as well as immunohistochemical stains, can assist in...
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