Cylindroma
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Epidemiology

Women are more commonly affected than men.

Etiology

Multiple cylindromas are often familial and inherited as an autosomal dominant trait, mainly in the setting of Brooke-Spiegler syndrome (familial autosomal dominant cylindromatosis).

Clinical Findings

Two main clinical forms of cylindroma are recognized; namely, a common, solitary type and a less-frequent form showing multiple lesions. Solitary cylindromas occur as slowly growing, asymptomatic, or painful, skin-colored, reddish, or bluish papules, nodules, or tumors on the head and neck region of adults. Multiple cylindromas occur as numerous papules, nodules, or variously sized tumors distributed mostly on the scalp and sometimes on the face or trunk. They show a tendency to confluence and may cover the entire scalp; or display a linear arrangement. Rarely, patients reveal widespread nodules, a clinical picture that mimics neurofibromatosis. Patients with Brooke-Spiegler syndrome show multiple cylindromas and trichoepitheliomas, occasionally in association with multiple basal cell adenomas of the parotid glands, milia, organoid nevi, basal cell carcinomas, and spiradenomas (see Spiradenoma). The susceptibility gene has been mapped to chromosome 16q12-q13 and has features of a recessive oncogene/tumor-suppressor gene.

Histopathology

There is usually a well-circumscribed dermal and/or subcutaneous lesion composed of irregularly shaped tumor islands and cords of basaloid cells arranged in a jigsaw puzzle-like pattern. Islands of basaloid cells are surrounded by a prominent rim of uniformly thickened periodic acid-Schiff (PAS)-positive basement membranes. They consist of two types of basaloid cells, namely, a central population with large nuclei and abundant cytoplasm and a peripheral population with smaller nuclei, arranged in a palisade at the periphery.

Treatment

Cylindromas may recur if incompletely excised. Local aggressive behavior and malignant transformation has occasionally been observed, especially in long-standing turban tumors of the scalp.