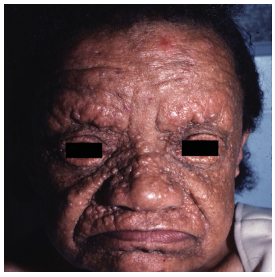

Brooke-Spiegler syndrome=multiple familial trichoepitheliomas and

cylindromassyndrome

Brooke-Spiegler syndrome is an uncommon disease with a predisposition to develop cutaneous adnexal neoplasms such as cylindromas, trichoepitheliomas, spiradenomas, trichoblastomas, basal-cell carcinomas, follicular cysts, organoid nevi, and malignant transformation of pre-existing tumors in the affected individuals [1]. Brooke-Spiegler syndrome is inherited in autosomal-dominant fashion, although expression and penetrance are variable. Lesions usually begin to appear in the second or third decades and gradually increase in number and size throughout adult life. Women are affected more frequently than are men. Mutations in the CYLD tumor-suppressor gene have been implicated in the phenotype diversity [2].

Cylindromas, trichoepitheliomas, and spiradenomas, which are the most commonly observed tumors, are typically located on the head and neck. Scalp cylindromas can become numerous and may eventually cover the entire scalp, which results in the so-called turban tumors and may result in partial or complete hair loss. Although cylindromas are usually benign neoplasms,

malignant transformation to cylindrocarcinomas is rare but well documented. The malignant cylindroma is locally aggressive, often metastasizes, and requires careful followup surveillance [3, 4]. Trichoepitheliomas are usually numerous and located on the face. In addition to neoplasms of the skin appendages, patients with Brooke-Spiegler syndrome also are at risk for developing tumors of the salivary glands, such as basal-cell adenomas and adenocarcinomas of the parotid glands and minor salivary glands [5].

Multiple trichoepitheliomas may be seen also in two other rare syndromes: Rombo syndrome (vermicular atrophoderma, milia, hypotrichosis, basal-cell carcinomas, trichoepitheliomas, and peripheral vasodilatation with cyanosis) and Bazex syndrome (follicular atrophoderma, hypotrichosis, occasional trichoepitheliomas, basal-cell carcinomas, and localized or generalized hypohidrosis) [6].

The histolopathologic spectrum of Brooke-Spiegler syndrome is broad and encompasses benign adnexal neoplasms of apocrine, follicular, and sebaceous differentiation, which can occur independently and conjointly. The most unusual findings are neoplasms with hybrid features, such as spiradenocylindromas, spiradenoma-trichoepitheliomas, cylindroma-trichoepitheliomas, and even the concurrence of all three adnexal tumors in one lesion [1, 7].

Treatment methods for adnexal neoplasms include excision, dermabrasion, electrodesiccation, cryotherapy, and radiotherapy. In addition, successful treatment with lasers such as the argon, CO2, and erbium:Yag plus CO2 have been reported [8]. Medical treatments for cylindromas that are currently being tested include sodium salicylate and prostaglandin A1, which are thought to restore growth control by inhibiting NF-B activity [9].