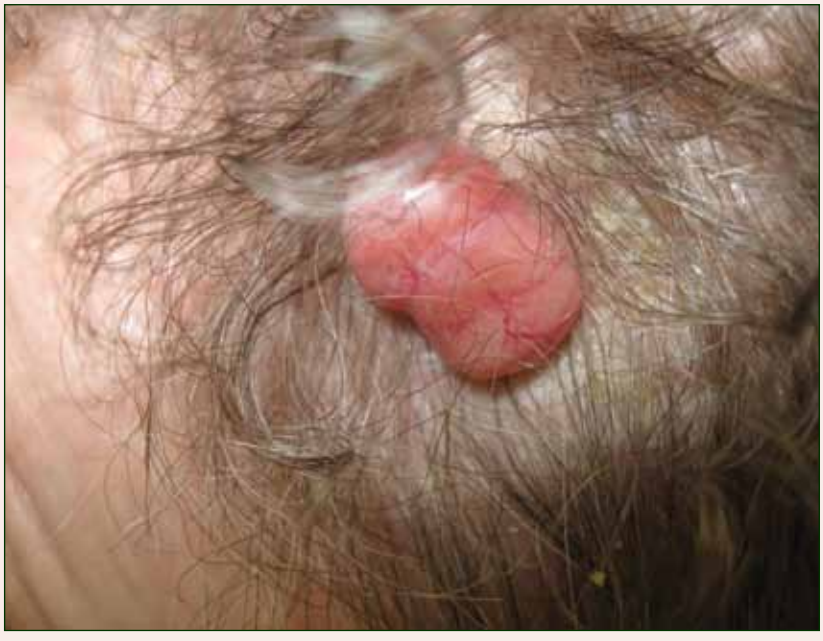
Lobular-Appearing Nodule on the Scalp

What's the diagnosis?



A 79-year-old woman presented with a lesion on the left side of the scalp of several years' duration that had slowly increased in size. Despite its growth, the lesion remained asymptomatic. Physical examination revealed an exophytic, lobular-appearing nodule on the left side of the temporoparietal scalp, measuring 1.5 cm in size.

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The authors report no conflict of interest.

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The Diagnosis: Dermal Cylindroma

Microsopic evaluation of a tangential biopsy revealed findings of a dermal process consisting of well-circumscribed islands of pale and darker blue cells with little cytoplasm outlined by a hyaline basement membrane (Figure). These cellular islands were arranged in a jigsawlike configuration. These findings were thought to be consistent with a diagnosis of cylindroma.

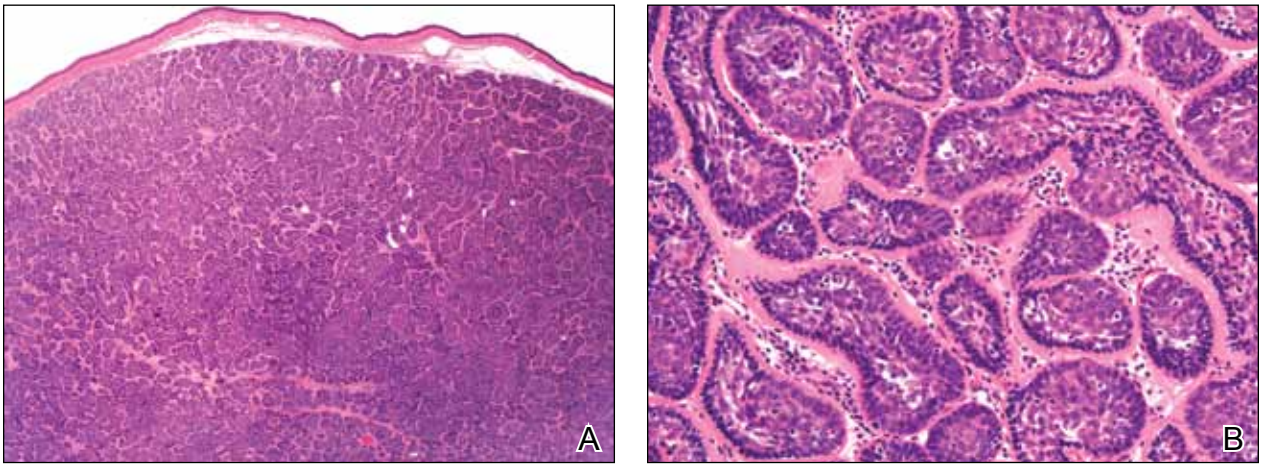
Cylindromas are benign appendageal neoplasms with a somewhat controversial histogenesis. Munger and colleagues[1] investigated the pattern of acid mucopolysaccharide secretion by these tumors in association with prosecretory vacuoles in proximity to the Golgi apparatus, which led to their impression that cylindromas most resemble eccrine rather than apocrine sweat glands. Other researchers, however, have concluded that cylindromas are of apocrine derivation.[2]

Clinically, cylindromas appear most often in 2 settings: isolated or as a manifestation of one of several inherited familial syndromes. One such syndrome is familial cylindromatosis, a rare autosomal-dominant disorder in which affected individuals develop multiple cylindromas, usually on the head and neck. The merging of multiple lesions gives rise to the often-employed term *turban tumor*.[3] This syndrome has been linked to mutations in the cylindromatosis gene, *CYLD*.[4] Brooke-Spiegler syndrome also has been associated with the development of multiple cylindromas. Similar to familial cylindromatosis, it is inherited in an autosomal-dominant fashion. Brooke-Spiegler syndrome is typified by the appearance of multiple cylindromas, trichoepitheliomas, and less commonly spiradenomas. Mutations in the *CYLD* gene also have been linked to Brooke-Spiegler syndrome in some cases.[5]

Although considered a benign entity, in rare cases cylindromas have shown evidence of malignant transformation to cylindrocarcinoma. This more aggressive tumor may occur in the setting of isolated cylindromas or more commonly in individuals with numerous lesions, as with both familial cylindromatosis and Brooke-Spiegler syndrome. These lesions may appear to grow rapidly, ulcerate, or bleed, traits that are not associated with their benign counterparts.

Diagnosis of cylindromas rests on histopathologic confirmation, which demonstrates well-defined dermal islands of epithelial cells comprised of dark- and pale-staining nuclei. These tumor islands are surrounded by a hyaline basement membrane and often take on the appearance of a jigsaw puzzle. Cylindrocarcinomas exhibit greater cellular pleomorphism and higher mitotic rates.

Dermal cylindromas require no further treatment but can be electively excised, while treatment of cylindrocarcinoma with excision is curative.[6] Definitive excision was offered to our patient, but she declined treatment.



Well-circumscribed dermal islands of both pale and darker blue cells outlined by a hyaline basement membrane. These cellular islands were arranged in a jigsawlike configuration (A and B)(both H&E, original magnifications ×20 and ×200).

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