

Umbilicated Keratotic Papule on the Scalp

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A 72-year-old man was referred to our dermatology clinic for evaluation of a solitary papule on the scalp measuring 3.2 mm in diameter with a keratotic umbilicated center of 1 year's duration. His medical history included acute appendicitis. Treatment with fusidic acid ointment 2% was unsuccessful. The papule was hard without tenderness on palpation. An excisional biopsy was performed under local anesthesia.

WHAT'S YOUR DIAGNOSIS?

- a. Darier disease
- b. keratoacanthoma
- c. seborrheic keratosis
- d. verruca vulgaris
- e. warty dyskeratoma

PLEASE TURN TO **PAGE E29** FOR THE DIAGNOSIS

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The authors report no conflict of interest.

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THE DIAGNOSIS: Warty Dyskeratoma

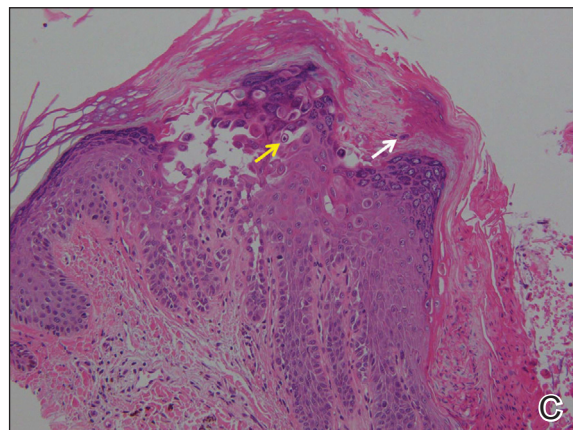
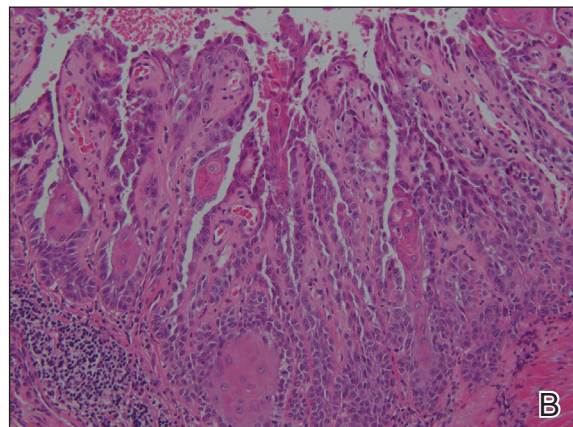
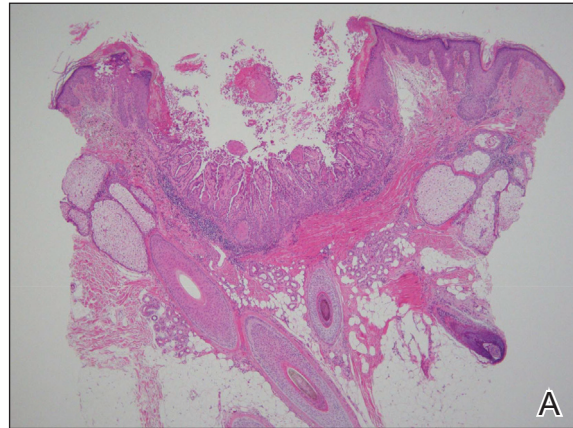
Warty dyskeratoma (WD) is a benign cutaneous tumor that was first described in 1954 as isolated Darier disease (DD). In 1957, Szymanski¹ renamed it *warty dyskeratoma* as a distinct condition from DD. Warty dyskeratoma typically presents as a flesh-colored to brownish, round, well-demarcated, and slightly elevated papule or nodule accompanied by an umbilical invagination at the center. It most commonly arises on the scalp, face, or neck.² In contrast to DD, familial occurrence is uncommon. It usually is difficult to distinguish WD from other conditions such as seborrheic keratosis, verruca vulgaris, or keratoacanthoma due to its macroscopic features. Therefore, histopathologic investigation is necessary for a precise diagnosis.

In our case, histologic investigation revealed a symmetric cup-shaped invagination filled with acantholytic and dyskeratotic keratinocytes with no atypia or mitotic figures (Figure, A). The bottom of the invagination was occupied by numerous villi covered by a single layer of basal cells (Figure, B). At the edge of the invagination, corps ronds and grains were observed in the granular layer and cornified layers, respectively (Figure, C).

The hallmark histopathologic findings are acantholysis and dyskeratosis just above the basal cell layer, called *focal acantholytic dyskeratosis*. The differential diagnosis includes other disorders associated with focal acantholytic dyskeratosis, such as DD and acantholytic squamous cell carcinoma.³ Distinguishing WD from DD may be difficult in rare cases with multiple lesions.⁴ In such cases, an autosomal-dominant inheritance pattern and younger age of onset should prompt clinicians to seek for mutations in the ATPase sarcoplasmic/endoplasmic reticulum Ca²⁺ transporting 2 gene, *ATP2A2*, for the diagnosis of DD.⁵ Additionally, the presence of atypia or mitotic figures will rule out malignant disorders such as squamous cell carcinoma.

Although the pathogenesis of WD is not fully understood, most clinicians consider it a follicular adnexal neoplasm because the lesions often are connected to the pilosebaceous unit on microscopic observation.⁶ Although WD-like lesions arising from the oral mucosa have been reported,⁷ their etiology may be different from WD because the oral mucosa lacks hair follicles.⁸ The term *warty* leads to speculation of the contribution of human papillomavirus to the pathogenesis of WD, but this has been questioned due to the negative result of viral DNA detection from WD lesions by polymerase chain reaction analysis.² Therefore, the term *follicular dyskeratoma* has been suggested as a novel denomination that reflects its etiology more precisely.²

The efficacy of topical treatment has not yet been established. Cryosurgery is another therapeutic option,



Warty dyskeratoma. A, A symmetric cup-shaped invagination filled with acantholytic and dyskeratotic keratinocytes (H&E, original magnification $\times 40$). B, Numerous villi covered by a single layer of basal cells at the bottom of the invagination (H&E, original magnification $\times 200$). C, At the edge of the invagination there were corps ronds (yellow arrow) in the granular layer and grains (white arrow) in the cornified layer (H&E, original magnification $\times 200$).

but it sometimes fails.⁹ As performed in our patient, excisional biopsy is the most reasonable treatment option to obtain both complete removal and precise diagnosis.

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