JAMA Dermatology Clinicopathological Challenge

Punctate Keratotic Papules on the Palm

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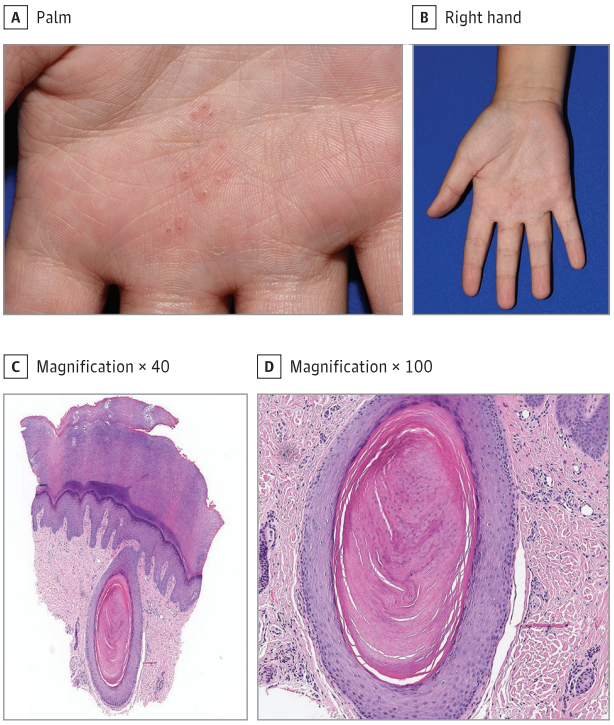

Figure. A and B, Grouped, dome-shaped, skin-colored, punctate papules with central keratotic plugging localized to the right palm and finger. C and D, Dilated follicular infundibulum with keratinous core and absent hair shaft (hematoxylin-eosin).

A girl in her teens presented for evaluation of asymptomatic, punctate, keratotic papules localized to the right palm and right third proximal finger that had been present since age 2 years. Prior treatments included cryotherapy, pulsed dye laser, and 40% salicylic acid, with partial improvement reported but never complete clearance. Medical history and family history were unremarkable. Physical examination revealed grouped and circumscribed, firm, skin-colored, 1- to 3-mm papules with a central punctate dark core (**Figure**, A and B). A 4-mm punch biopsy from a papule on the palm was performed (Figure, C and D).

WHAT IS YOUR DIAGNOSIS?

A. Porokeratotic eccrine ostial and dermal duct nevus

B. Punctate palmoplantar keratoderma

C. Familial comedonal Darier disease

D. Nevus comedonicus

Diagnosis

D. Nevus comedonicus

Discussion

Punch biopsy results revealed dilated cystic spaces with mild squamous hyperplasia and central laminated keratin resembling a dilated terminal hair follicle infundibulum (Figure, C and D). The clinical and pathologic findings were considered diagnostic of a nevus comedonicus.

Nevus comedonicus is a hamartomatous proliferation resulting from improper keratinization of the pilosebaceous unit, and it presents as punctate papules with a central keratotic core. Individual lesions may be present at birth or develop during childhood, but they are generally fully established by age 10 years.[1] Linear or blaschkoid presentations involving the face, neck, upper extremities, and trunk are most common, with a prevalence of 1 in 45 000 to 1 in 100 000 persons.[2] However, palmar-plantar presentations have been described despite these areas being devoid of pilosebaceous units.[3,4] Nevus comedonicus development is postulated to reflect abnormal filaggrin expression or overstimulation of fibroblast growth factor receptor 2, with high expression of interleukin 1 leading to improper pilosebaceous unit development.[2] However, there is still debate as to whether development of nevus comedonicus, particularly on the palms and soles where pilosebaceous units are absent, may be caused by abnormal epidermal invagination. Histopathology is characterized by keratin-filled, dilated, cystic spaces with infundibular differentiation in the absence of hair shafts, arrector pili muscle, or sebaceous glands with variable acanthosis.[2]

Nevus comedonicus most often occurs in isolation, but familial clustering and association with skeletal, ocular, and central nervous system involvement can occur in nevus comedonicus syndrome.[5] These syndromic presentations are variably associated with other cutaneous tumors such as trichoepithelioma, pilar sheath acanthoma, or syringocystadenoma papilliferum.[2]

Clinical history and histopathologic examination may be required to differentiate nevus comedonicus from other punctate palmar-plantar disorders. Porokeratotic eccrine ostial and dermal duct nevus and nevus comedonicus both present early in life and tend to be unilateral, but porokeratotic eccrine ostial and dermal duct nevus is characterized by a cornoid lamella overlying a dilated acrosyringium with loss of granular layer and focal dyskeratosis.[6] Punctate palmoplantar keratoderma generally has a later age of onset (teens to 20s), bilateral distribution, and mild epidermal invagination with central overlying orthohyperkeratosis on histopathologic analysis.[1] Familial comedonal Darier disease has comedonelike lesions but shows focal acantholytic dyskeratosis on histologic analysis and typically presents with a generalized seborrheic distribution rather than a linear or circumscribed distribution.[7] Lastly, nevus comedonicus can be confused with verruca vulgaris, but verruca vulgaris classically has interrupted skin lines and pinpoint hemorrhage on dermoscopy, bleeds on paring, and reveals papillomatosis with orthokeratosis and parakeratosis in addition to hypergranulosis and koilocytosis on histologic analysis.

Treatment is challenging. Destructive approaches using fractional carbon dioxide laser,[8] cryotherapy, surgical excision, and comedone extraction have been reported as well as medical approaches using topical or oral retinoids, topical antibiotics, calcipotriene, and keratolytics, with highly variable results described.[9] Because most cases are asymptomatic, patients can be reassured that treatment is not necessary aside from cosmetic concerns.

ARTICLE INFORMATION

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