



AB011. A Gottron's papule mimicker: an unusual presentation of porokeratosis plantaris palmaris et disseminata

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Abstract: A 52-year-old woman with a past medical history of breast cancer presented for evaluation of a scaly eruption on bilateral dorsal and palmar hands for the preceding seven months. The patient also noted scaling of her right foot, hallux, and Achilles tendon. Her physical exam was notable for nail cuticle dystrophy with normal nail fold capillaries, scattered hyperkeratotic erythematous coalescing papules and plaques overlying her metacarpophalangeal and distal interphalangeal joints. The right palm was notable for tender, firm, subcutaneous nodules. The patient reported neither joint pain, muscle pain, nor weakness. The clinical appearance of Gottron's papules and presence of palmar papules, raises suspicion for dermatomyositis; therefore, a biopsy of a papule present on her right Achilles and a punch biopsy of a papule on her right fourth proximal interphalangeal joint were performed. The histopathology of both biopsies demonstrated interface alteration with columns of parakeratosis. There were also areas of epidermal invagination and dells with plugs of broad parakeratosis resembling cornoid lamellae and underlying interface alteration, suggesting a rare

diagnosis of porokeratosis plantaris palmaris et disseminata (PPPD). In follow-up, the patient returned with new distinctly annular palmar lesions with peripheral scale, resembling porokeratosis. Of note, poikilodermatous skin in dermatomyositis may histologically display an interface dermatitis, seen in our patient, with epidermal atrophy and necrotic, dyskeratotic keratinocytes with basal layer vacuolization, increased dermal mucin deposition, and vascular damage. The histology of Gottron's papules is similar, and often shows a lymphocytic infiltrate, epidermal hyperkeratosis, papillomatosis, acanthosis and less epidermal atrophy. However, with the patient's developing clinical picture of palmar annular lesions and her pathology results showing cornoid lamellae (a hallmark of porokeratosis), dermatomyositis was considered less likely and PPPD more likely. Age appropriate malignancy screening was performed and normal. The patient is being followed closely and doing well. Teaching Point: biopsy can help confirm or refute a diagnosis of Gottron's papules which is often considered a pathognomonic sign of dermatomyositis. In this patient the hand lesions were a mimicker, and dermatomyositis was not diagnosed.

Keywords: Porokeratosis plantaris palmaris et disseminate (PPPD); Gottron's papules

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