

Multiple minute digitate hyperkeratosis - a peculiar entity*

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Abstract: Multiple minute digitate hyperkeratosis is a rare, non-follicular dermatosis, with fewer than 30 cases described worldwide. It can be either acquired or inherited in an autosomal dominant pattern. We describe the case of an 83-year old patient with life-long, multiple, digitate, milimetric lesions, and a positive family history for the same dermatosis.

Keywords: Dermatology; Genes, dominant; Keratosis

INTRODUCTION

Multiple minute digitate hyperkeratosis (MMDH) is a rare, non-follicular, keratinization dermatosis of unknown etiology that can be either inherited (in what appears to be an autosomal dominant pattern) or acquired.¹ There are fewer than 30 cases described in international literature.²

We herein report the case of a female patient presenting with a life-long history of a spiny dermatosis.

CASE REPORT

An 83-year-old woman presented with multiple, asymptomatic, non-follicular, skin-colored finger-like projections sparing the face, palms and soles (Figure 1). In the anterior thoracic wall, the lesions were flattened, with a yellow hue, forming 1-3mm crateriform papules (Figure 2).

They had developed over a period of 50 years; she had no other complaints and her physical exam was otherwise unremarkable. Apart from hypertension, she had no other comorbidities. These lesions had also affected her father, sister and daughter, since adulthood.

Biopsy of one of the crateriform papules showed ortho- and parakeratotic hyperkeratosis overlying a slightly acanthotic epidermis with a preserved granular layer and a very mild perivascular lymphocytic infiltrate in the upper dermis (Figures 3 and 4). Ancillary tests, including complete blood count, kidney and liver function, serum protein electrophoresis and chest x-ray, were normal; furthermore, she was up-to-date with her age-appropriate cancer screening exams.

She was diagnosed with MMDH and began treatment with salicylic acid 8% in petrolatum ointment, resulting in transient improvement.

DISCUSSION

Since Goldstein's first description of MMDH, in 1967, at least 29 cases have been reported under different names, all comprising non-follicular digitate keratosis.^{1,2}

In a recent review, Caccetta et al. categorized digitate keratosis, proposing MMDH as a more accurate term to classify familial disseminated filiform hyperkeratosis, disseminated spiked keratosis, minute aggregate keratosis, parakeratotic horns, transient

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postinflammatory digitate keratosis and digitate keratosis, which they considered to be synonyms for the same keratinizing disorder.²

Typical MMDH lesions include white, yellow, brown or skin-colored, milimetric spicules and flat-topped, dome-shaped or crateriform papules, affecting mainly the trunk and limbs, sparing the facial and palmoplantar region.²



FIGURE 1: Multiple digitate filiform skin-colored hyperkeratosis occupying the inner aspect of the left thigh



FIGURE 2: Multiple, 2-3 mm, honey-colored, crateriform papules distributed over the right mammary region. These lesions were present bilaterally on the anterior thoracic wall

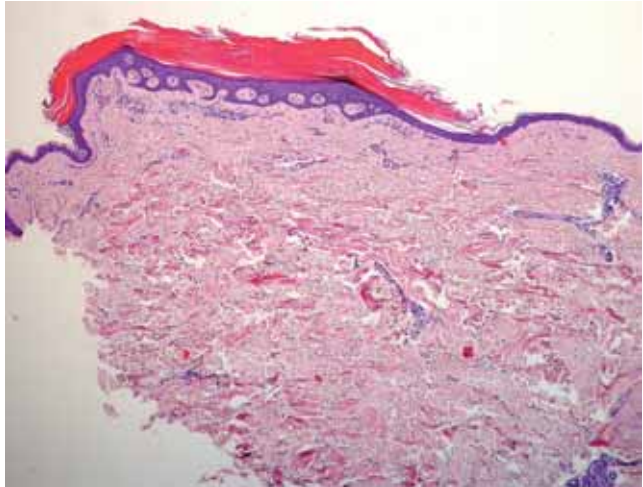


FIGURE 3: Biopsy of a crateriform papule, stained with HXE, displaying acanthosis, hyperkeratosis and a mild lymphocytic infiltrate in the upper dermis

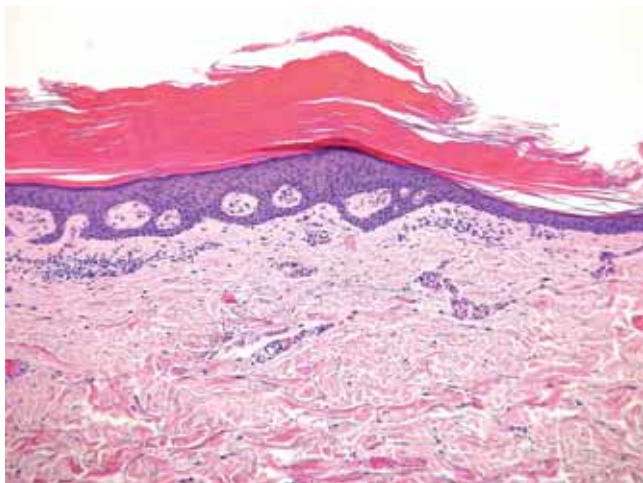


FIGURE 4: Detail of the crateriform papule's biopsy. Note the parakeratotic hyperkeratosis overlying a tented epidermis with acanthosis and a preserved granular layer

Histopathology presents focal orthokeratotic hyperkeratosis arising from a tented epidermis, and a stratum granulosum of variable thickness. Parakeratosis and epidermal invagination, as well as a superficial dermal lymphoplasmocytic infiltrate, have also been described, although more rarely.² A constant feature that is a common denominator throughout the reported cases, is the lack of follicular involvement.

Electron microscopy shows reduced keratohyaline granules with a variable number of Odland bodies.

MMDH can present as an inherited trait in the early twenties or thirties, or appear later in life in a sporadic acquired form.²⁻⁵ The latter has been reported

as a paraneoplastic syndrome, resolving after successful treatment of the accompanying laryngeal carcinoma.⁶ The former is most likely an autosomal dominant dermatosis, even though its genetic basis and pathophysiology remain unknown.

Other keratinization disorders can be entertained in the differential diagnosis of digitate keratosis, including: Kyrle's hyperkeratosis follicularis et parafollicularis, Flegel's hyperkeratosis lenticularis perstans, lichen spinulosus, phrynoderma, spiny keratoderma, arsenical keratosis, hyperkeratotic spicules and trichodysplasia spinulosa.

The first two entities can be separated from MMDH based on their distribution (predominantly acral), as well as their different histopathology.³

Lichen spinulosus occurs mostly in children and lesions tend to form plaques of follicular spines. Phrynoderma, due to vitamin A deficiency, also leads to generalized follicular spines, as well as xerosis.⁷ This follicular accentuation facilitates distinction from MMDH. Spiny keratoderma, either inherited or acquired, occurs only on the palms and soles, just like arsenic keratosis that also demonstrates a predilection for acral sites in the presence of arsenic exposure.⁸ This location is typically spared in MMDH.

Hyperkeratotic spicules are harder to separate from MMDH, since they can be follicular or non-follicular, and affect the entire integument, but these paraneoplastic keratoses contain keratin and paraprotein that appear as eosinophilic globules on histopathology.

In trichodysplasia spinulosa, which has recently been associated with polyomavirus in immunosuppressed individuals, follicular spines occupy the central face, allowing for distinction from MMDH.⁹

Systemic or topical retinoids and other topical keratolytic agents have reportedly led to temporary improvement but lesions inexorably return upon cessation of therapy.¹⁻⁵

Our case outlines not only the characteristic non-follicular, finger-like keratosis but also honey-colored crateriform papules that have seldom been described. This perhaps corresponds to the multiple aggregate keratosis that Shuttleworth differentiated from MMDH, based only on the semiology of the lesions. Our patient, however, exhibits both clinical features while maintaining typical histopathologic findings, thus strengthening the argument that MMDH and multiple aggregate keratosis are, in fact, the same entity.¹⁰ □

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