



## ORIGINAL RESEARCH

# Dowling-Degos Associated to Verneuil Disease: A New Case Report

Meryem Khalidi\*, Hasna Kerrouch, Mohammed El amraoui, Naoufal Hjira and Mohammed Boui



Department of Dermatology-Venerology, Mohammed V Military Hospital of Instruction, Rabat, Morocco

\*Corresponding author: Meryem Khalidi, Department of Dermatology-Venerology, Mohammed V Military Hospital of Instruction, Rabat, Morocco

## Introduction

Dowling-Degos disease (MDD) is a rare autosomal dominant genodermatosis belong to the epidermal differentiation disorders. Its association with Verneuil's disease (MV) has only recently been described, and we are reporting a new case.

## Case Report

It's about a 50-year-old woman, with no notable medical history, who consulted for skin hyperpigmentation of the folds and face that had appeared for 6 years before. The clinical examination showed hyperpigmented reticulated macules of the lateral faces of the neck and face, the nape of the neck, axillary, under breast and inguinal folds associated with cysts and comedones and suppurative nodulocystic and fistulous formations under breast and inguinal resulting in deep irregular scars. The patient reported the same lesions in her paternal aunt. A skin biopsy taken from the hyperpigmented lesions showed hyperpigmented epidermal tabs without cytological atypia, connected to an epidermis of normal thickness. The dermis was normal.

## Discussion

Dowling-Degos disease (MDD) is a rare autosomal dominantly inherited genodermatosis of variable penetrance, secondary to a mutation in the keratin 5 gene, known to be involved in cell adhesion and transfer of melanosomes 123 [1]. It occurs in the 3<sup>rd</sup>-4<sup>th</sup> decade of life with a predilection for the female sex. It is characterized by a progressive evolution of acquired reticu-

lated skin hyperpigmentation which initially affects the flexion regions: The armpits, the folds of the groin and subsequently the subgluteal and submammary regions, the neck and the trunk. Subsequently, pseudo-comedones appear on the back and neck [2]. Perioral cribriform scars as well as follicular cysts are also observed. MDD and MV association has already been described and even seems relatively frequent, ranging up to 38% MV in series of MDD [3]. It is postulated that the abnormal epithelial proliferation which forms the bed for DCS and results in follicular occlusion, would promote the genesis of VM [2]. Microscopic examination shows epidermal buds forming thin, threadlike, pigmented and interconnected, epithelial tabs of epidermal and follicular origin. The hair follicles are sometimes dilated making horny cysts [4]. The differential diagnosis is mainly made with acanthosis nigricans and seborrheic keratosis [2]. But the clinical context makes it easy to distinguish them. Unfortunately therapies are very poor, and just a few cases had tried cyclins or isotretinoin but without significative improvement. Patients claim specially the high social psychological impact of the disease [2].

## Conclusion

The association of Dowling Degos disease and verneuil disease is increasingly described around the world, it is a rare association to look for in any hereditary context of a reticulated pigmentary pathology.

## References

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