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Mal de Meleda - description of a case report

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Background and Aim: Palmoplantar keratodermas (PPKs) consist of a heterogeneous group of disorders characterized by thickening of the palms and soles. The condition may be subdivided into hereditary keratodermas, acquired forms, and conditions in which PPK is an associated feature of a specific dermatosis. Mal de meleda (MdM), a rare autosomal recessive form of PPKs is characterized by erythema and hyperkeratosis of the palms and soles with a sharp demarcation and that progress with age and extend to the dorsal aspects of the hands and feet, accompanied by unguis and digital constrictions changes (Pseudoainhum).

Case description: 23 years old caucasian woman. Only child of a healthy couple in the first degree consanguineous (direct cousins). Without relevant pathological antecedentes. She presents since birth, yellowish transgressive palmoplantar keratoderma, with paraffin aspect, that was aggravated with growth. Currently, the lesions are accompanied by erythema, which extends to the back of the hands and feet, nail dystrophy and pseudoainhum. It also presents excessive sweating and foul odor of the feet and hands. Without comorbidities or other systemic changes. She was followed in dermatology consultation and treated with retinoids without great benefit, so at the moment is only with keratolytic and emollient in continuous use. The patient is unemployed and she attributes this to the disease, for its location with great exposure and negative aspect.

Discussion: MdM is a chronic disease with great therapeutic difficulty. It has high morbidity, negative impairment in the patient's quality of life and interpersonal relationships. This case report draws attention to the importance of family assessment, particularly the family genogram, to evaluate the transmission of hereditary diseases, especially in cases of consanguinity. The GP plays a key role in monitoring and support these patients in both prescription comfort therapy, as in addressing its consequences and psychological rebate.