

Localized hyperkeratotic scabies in a patient with Down syndrome simulating verrucous carcinoma*

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Resumo: É apresentado um paciente de Salvador, Bahia, com síndrome de Down e escabiose hiperqueratósica localizada.

Palavras-chave: Escabiose; síndrome de Down.

Summary: A patient from Salvador, Bahia, with Down syndrome having localized hyperkeratotic scabies is presented.

Key words: Scabies; Down syndrome.

INTRODUCTION

Scabies is caused by the anthropophil mite *Sarcoptes scabiei*, which is transmitted by close physical contact. In spite of the fact that there is a single pathogenic agent, a clinical spectrum is found in both humans and animal species.¹ Infrequently, an infestation becomes widespread and the patients acquire thick crusted lesions containing numerous burrows and mites. This aspect of the disease is called hyperkeratotic scabies (HyS), crusted scabies or Norwegian scabies because it was first reported in patients from Norway by Danielssen and Boeck in 1848. This condition is highly contagious² and usually responsible for the outbreaks of local epidemics.³ HyS is rarely found in otherwise healthy individuals. It has traditionally been associated with mentally retarded and physically debilitated patients. Recent reports have increasingly linked this variant of classical scabies with immunosuppression, as in acquired immunodeficiency syndrome (Aids), leprosy and in transplant recipients.^{4,5,6} Rarely this form of scabies have a localized appearance.⁷

Herein we report a case of localized HyS, on a old burn scar of a patient with Down syndrome, in which verrucous squamous cell carcinoma was strongly considered

in the clinical differential diagnosis. Ultimately, the histopathology established the correct diagnosis.

REPORT OF CASE

A 34-year-old black woman with Down syndrome was burned on her thorax, right arm and right anterior thigh at the age of 16 years. She underwent skin grafting to the right thigh at that time, resulting in pigmented scars with some achromyc areas. Since two years interned in a comprehensive care residential psychiatric unit of the Santo Antonio Hospital, presenting an asymptomatic patchily pigmented warty lesion 20cm in diameter, causing fissures and repeated secondary infections, solely within the confines of the thigh burn scar, simulating a verrucous carcinoma (Figure 1, 2). No definite diagnosis was made at first.

Ordinary treatment at regularly intervals was made with topical scabicides alone, which failed to clear her lesions. Nosocomial outbreak of scabies in the care unit where she was interned occurred periodically.

The histological examination revealed a markedly thickened epidermis with hyperkeratosis. The mean deep of the epidermis was 2.640mm; the stratum corneum alone had an average thickness of 0.9mm. There was also crusting and the presence of multitudes of pearl-shaped mites, located in channels in the stratum corneum and within the subcorneal region (Figure 3). In the upper dermis, the perivascular areas and the interstitium had infiltrates composed of a moderate number of lymphocytes and eosinophils.

For successful cure of HyS it was necessary to proceed isolation of the patient. She was treated for two weeks with 20 percent Benzyl Benzoate lotion over the body and

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Figure 1: Clinical picture. Appearance of the right anterior thigh at the time of diagnosis. Note scaly plaques and crusts

three percent sulfur ointment occlusive over the lesion for 12 hours alternated with 0.1 percent retinoic acid for 12 hours, which resulted in the clearing of the skin lesions. After the applications, there was complete resolution of the parasitic infestation. No further nosocomial transmission occurred.

DISCUSSION

HyS is uncommon in clinical practice and may present as papulosquamous dermatosis.⁸ The localized skin lesions in this case simulated those of verrucous squamous

cell carcinoma and because its development in chronic scars is a well known phenomenon, such a diagnosis was strongly considered clinically at first. The infection is frequently overlooked because of its atypical presentations,⁹ as was the case for the patient under discussion. Consequently, she became the source of epidemic scabies in the care unit.

Patients with cognitive deficiency or an immunodeficiency disorder (including immunosuppressive therapy) are predisposed to developing HyS.⁹ Down's syndrome is a frequent association. The reason for this combination with mental abnormality is not completely understood, but lack of appreciation of pruritus may be important.¹⁰ The infestation is related to the cutaneous immune response and thus HyS should be considered an opportunistic infestation.⁷ Having a common physiopathological mechanism with the ordinary scabies, the host reaction process and epidermal hyperplasia in HyS has been considered the consequence of a delayed type hypersensitivity reaction which is abnormally modulated. The immune response is unable to reject the invading parasites, being defective or at least deviated from the more usual reaction pattern.¹ On the other hand, immunologic consequences of scabies may depend on mast cell stimulation and since mediators of inflammation will be released due to degranulation of mast cells, this could play a role in the pathogenesis of the clinical and histologic picture of scabies.¹¹

The transmission in hospitals of ordinary scabies from a patient with HyS to other patients, health care workers, etc., could be expected. Patients with HyS demand strict barrier nursing if nosocomial transmission is to be avoided. Therapy needs the sequential use of scabicides, usually over a longer period than is required to clear an

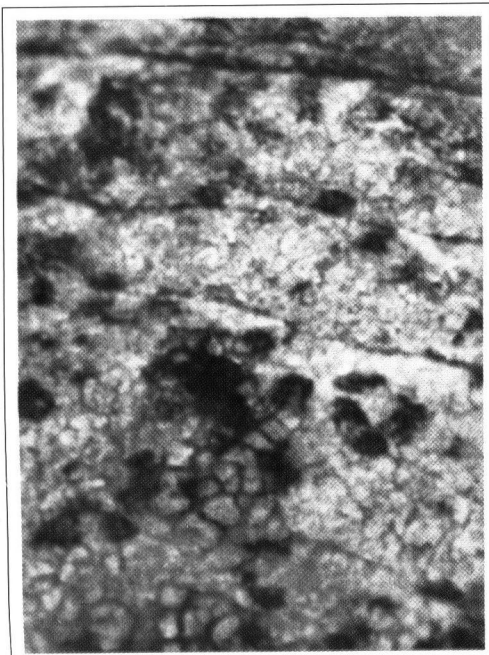


Figure 2: Close-up view of the lesion to show irregularly shaped verrucous aspect

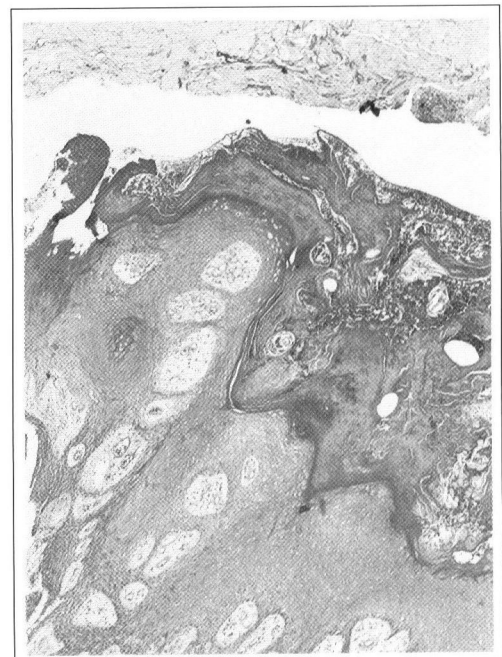


Figure 3: Skin from the crusted lesion. Sections of many burrows and mites of Sarcoptes scabiei are in the hyperkeratotic horny layer (HE, 100x)

ordinary case of scabies. After isolation, our patient responded to the scabicide Benzyl Benzoate associated with keratolytic agents.

This case illustrates the pitfalls in recognition of HyS and the importance of early diagnosis and therapy. □

REFERENCES

1. Van Neste D. Critical commentary to "Coexistence of an Unusual Form of Scabies and Lepromatous Leprosy". *Path Res Pract* 1996;192:92-3.
2. Feinsod FM. Arthropods directly causing human injury. In: Warren KS, Mahmoud AAF, eds. *Tropical and Geographical Medicine*. New York: McGraw-Hill, 1984:499-516.
3. Moberg SAW, Lowhagen GBE, Hersle KS. An epidemic of scabies with unusual features and treatment resistance in a nursing home. *J Am Acad Dermatol* 1984;11:242-4.
4. Lang E, Humphreys DW, Jaqua Stewart MJ. Crusted scabies: a case report and review of the literature. *S D J Med* 1989;42(4):15-7.
5. Inserra DW, Bickley LK. Crusted scabies in acquired immunodeficiency syndrome. *Int J Dermatol* 1990;29(4):287-9.
6. Barbosa Jr. AA, Silva TMC, Santos MIR, et al. Coexistence of an unusual form of scabies and lepromatous leprosy. *Path Res Pract* 1996;192:88-90.
7. Arico M, Noto G, La Rocca E, Pravata G, Bivona A. Localized crusted scabies in the acquired immunodeficiency syndrome. *Clin Exp Dermatol* 1992;17(5):339-41.
8. Chen DY, Lan JL. Crusted scabies in systemic lupus erythematosus: a case report. *Chung Hua Min Kuo Wei Sheng Wu Chi Mien I Hsueh Tsa Chih* 1993;26(1):44-50.
9. Kolar KA, Rapini RP. Crusted (Norwegian) scabies. *Am Fam Physician* 1991;44(4):1317-21.
10. Burns DA. Diseases caused by arthropods and other noxious animals. Crusted scabies (Norwegian scabies). In: Champion RH, Burton JL, Ebling FJG, eds. *Textbook of Dermatology*. 5th ed. Oxford: Blackwell, 1992:1305-7.
11. Mostafa FF, Nasr AN, El-Harras M. The role of mast cells in treatment of scabies. *Int J Dermatol* 1995;34(3):186-9.

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