

(CASE REPORT)



Ketorash: An underdiagnosed entity

Duarte Flor ^{1,*}, Joana Xará ¹, Francisco Martins ¹, Joana Calvão ¹, Inês Coutinho ¹ and Ricardo Vieira ^{1,2}

¹ Dermatology Department, Coimbra Local Health Unit, Portugal Praceta Prof. Mota Pinto 3004-561 Coimbra.

² Dermatology Department, Faculty of Medicine, University of Coimbra, Azinhaga de Santa Comba, Celas 3000-548 Coimbra.

World Journal of Biology Pharmacy and Health Sciences, 2024, 19(03), 069–073

Publication history: Received on 26 July 2024; revised on 02 September 2023; accepted on 05 September 2024

Article DOI: <https://doi.org/10.30574/wjbphs.2024.19.3.0596>

Abstract

Ketoacidosis-Induced Prurigo Pigmentosa is a rare, inflammatory disease characterized by pruritic, erythematous papules with a reticulate pattern on the torso and neck. It affects predominantly young adult Asian women with risk factors for metabolic ketosis, such as Diabetes Mellitus, anorexia nervosa, bariatric surgery and ketogenic diet.

A 26-year-old female being treated for refractory focal frontal and generalized epilepsy with ketogenic diet for four months was consulted due to a three-month-long dermatosis which started in the anterior and posterior cervical region and progressed towards the dorsal midline area. Skin lesions were papular and erythematous, often merging with a vaguely reticulated pattern. These lesions were highly pruriginous and worsened during periods of higher ketonemia.

Histological examination of a cutaneous biopsy sample was compatible with Ketorash. The patient suspended ketogenic diet and was medicated with topical erythromycin, with full resolution of the lesions and no recurrences after 8 months.

Keywords: Ketorash; Ketosis; Ketogenic diet; Prurigo Pigmentosa; Pruritus.

1. Introduction

Ketoacidosis-Induced Prurigo Pigmentosa (also known as “Ketorash” or Nagashima disease) is a rare, inflammatory disease characterized by pruritic, erythematous papules with a reticulate pattern on the torso and neck. It affects predominantly young adult Asian women with risk factors for metabolic ketosis, such as Diabetes Mellitus, anorexia nervosa, bariatric surgery and ketogenic diet¹.

We present an extremely rare occurrence of ketorash induced by ketogenic diet in a caucasian patient.

2. Clinical Case

We report the case of a 26-year-old female diagnosed with generalized focal frontal epilepsy since childhood. Her condition was resistant to medical therapy, and therefore she had been treated with ketogenic diet for the last four months. She did not take any chronic medication, nor had any history of allergies.

She was referred to the Dermatology department due to a three-month-long dermatosis which started in the anterior and posterior cervical region and progressed towards the dorsal area, predominantly in the midline. Skin lesions were papular and erythematous, often merging with a vaguely reticulated pattern, with superimposed punctiform excoriations (figure 1). These lesions were highly pruriginous and worsened during periods of higher ketonemia.

* Corresponding author: Duarte Flor



figure 1a Cervical and dorsal cutaneous lesions



Figure 1b Cervical papular and erythematous lesions, merging with a vaguely reticulated pattern



Figure 1c Dorsal papular and erythematous lesions with superimposed punctiform excoriations

Histological examination revealed moderate acanthosis of the epidermis, with foci of parakeratosis, diffuse spongiosis, focal exocytosis of small lymphocytes and scarce apoptotic keratinocytes. In the superficial dermis a lymphomononuclear perivascular infiltrate was predominant, with a few scattered extra and intra-vascular neutrophils. Hematoxylin and eosin staining is shown in figure 2.

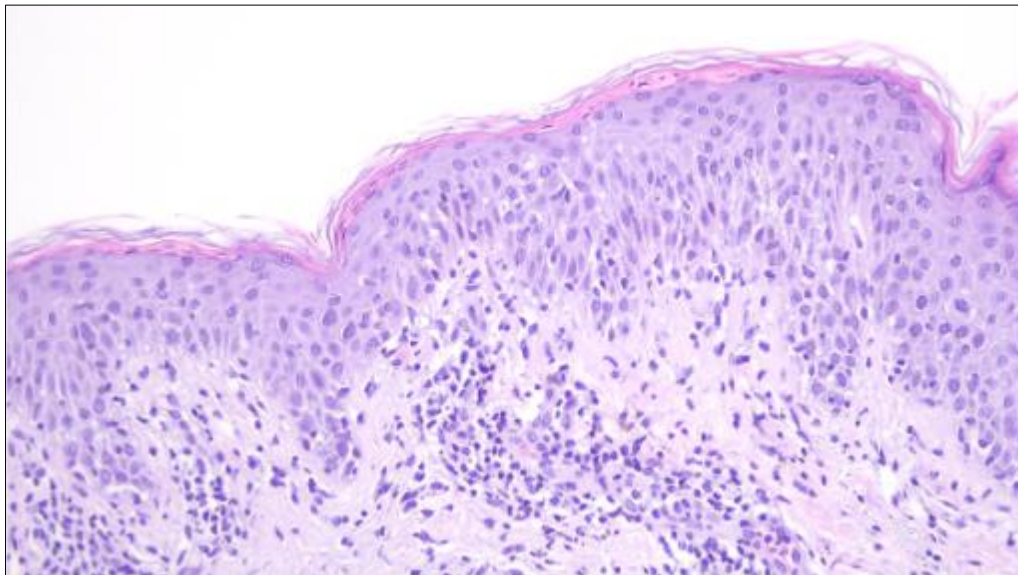


Figure 2a Hematoxylin and eosin staining of lesion biopsy

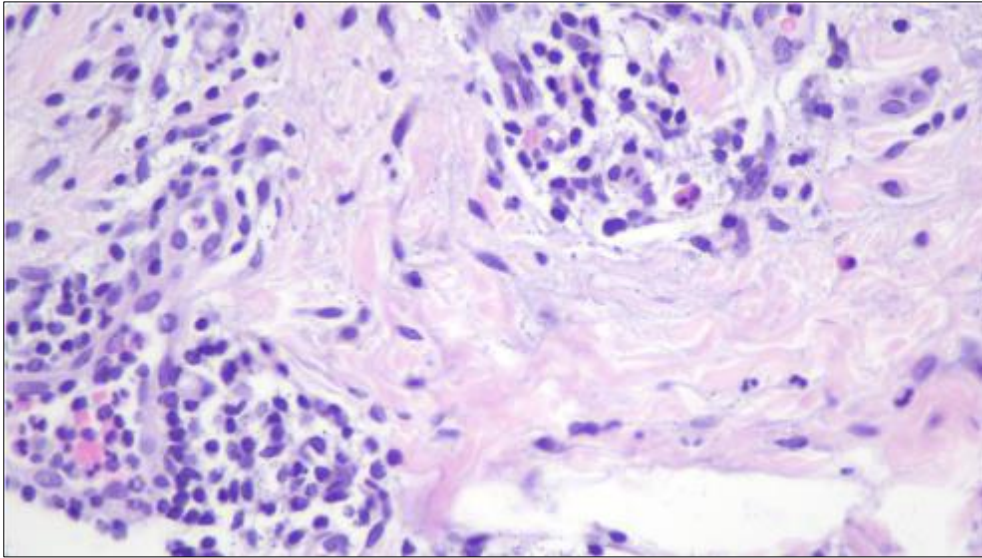


Figure 2b Same HE staining showing lympho-mononuclear perivascular infiltrate

The patient was diagnosed with Ketoacidosis-Induced Prurigo Pigmentosa and treated with topical erythromycin, as well as suspension of the ketogenic diet, and presented complete resolution and no recurrences in 8 months, confirming the diagnosis.

3. Discussion

Ketoacidosis-Induced Prurigo Pigmentosa (also known as “Keto rash” or Nagashima disease) is a rare, inflammatory disease affecting predominantly young adult Asian women and typically characterized by pruritic, erythematous papules on the torso and neck that evolve into a reticulate pattern and resolve with hyperpigmentation, although diverse presentations may develop¹.

Risk factors include Diabetes Mellitus, anorexia nervosa, bariatric surgery and ketogenic diet, which can all induce a state of metabolic ketosis, resulting in rash onset. The pathogenesis is unknown, but it is theorized that ketone bodies may induce perivascular inflammatory infiltrate. Local factors, such as sweating and pre-existing atopic dermatitis, may play a role². Differential diagnosis includes Grover’s disease, Darier disease and foliaceous pemphigus.

Histological examination is essential to the diagnosis. Early stages present superficial perivascular neutrophilic infiltrate, while more advanced lesions show spongiosis and necrotic keratinocytes, followed by lymphocytic infiltrate with melanophages in the papillary dermis^{2,3}.

Treatment is based on reduction of ketosis, as well as adjuvant topical antibiotics⁴. Oral tetracyclines, especially minocycline, have been used with success. Dapsone may be effective in refractory cases. The disease responds poorly to topical corticosteroids and antihistamines. Most patients completely recover after a few weeks, apart from residual hyperpigmentation of the affected area.

4. Conclusion

We report a rare case of Ketoacidosis-Induced Prurigo Pigmentosa in a Caucasian patient, an inflammatory, underdiagnosed condition that must be considered in the differential diagnosis of similar dermatosis in patients with known risk factors for ketosis.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of ethical approval

The present research work does not contain any studies performed on animals/humans subjects by any of the authors.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

References

- [1] Zeng X, Li L, Cui BN. Prurigo pigmentosa: a clinical and histopathological study of nine Chinese cases. *J Eur Acad Dermatol Venereol*. 2016 Oct;30(10):1794-1798. doi: 10.1111/jdv.13659. Epub 2016 Jul 21. PMID: 27440732.
- [2] Xiao A, Kopelman H, Shitabata P, Nami N. Ketogenic Diet-induced Prurigo Pigmentosa (the "Keto Rash"): A Case Report and Literature Review. *J Clin Aesthet Dermatol*. 2021 Dec;14(12 Suppl 1):S29-S32. PMID: 35291259; PMCID: PMC8903224.
- [3] Shahrigharahkoshan S, AlHalees Z, Pehr K. Ketogenic diet-induced prurigo pigmentosa: a rising association. *Int J Dermatol*. 2022 Jul;61(7):779-782. doi: 10.1111/ijd.15991. Epub 2021 Nov 26. PMID: 34826138.
- [4] Almaani N, Al-Tarawneh AH, Msallam H. Prurigo Pigmentosa: A Clinicopathological Report of Three Middle Eastern Patients. *Case Rep Dermatol Med*. 2018 Jul 9; 2018:9406797. doi: 10.1155/2018/9406797. PMID: 30105102; PMCID: PMC6076956.