

A Case of Pemphigus Vegetans in a Girl with Oculocutaneous Albinism

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Abstract

Oculocutaneous Albinism (OCA) is a group of inherited disorders affecting melanin pigment in the body. It is an autosomal recessive inherited disorder, characterized by hypopigmentation of skin, hair, and eyes which may cause serious problems over time. Pemphigus vegetans is an unusual localized form of pemphigus vulgaris. It is an autoimmune disease characterized by the production of IgG autoantibodies against intercellular adhesion protein desmoglein, forming acantholysis. In this case report, we've reported a very rare case of Pemphigus vegetans in a 24-year-old girl with Oculocutaneous albinism.

Keywords: Oculocutaneous albinism • Nystagmus • Diagnosis

Introduction

Oculocutaneous Albinism (OCA) is a group of inherited disorders affecting melanin pigment in the body. It is an autosomal recessive inherited disorder, characterized by hypopigmentation of skin, hair, and eyes with reduced visual acuity, iris transillumination, photophobia, nystagmus, and foveal hypoplasia. It has many types ranging from OCA Type I-VII [1-5]. Pemphigus vegetans is an unusual localized form of pemphigus vulgaris. It is an autoimmune disease characterized by the production of IgG autoantibodies against intercellular adhesion protein desmoglein, forming acantholysis. The eroded areas do not heal like usual in the case of Pemphigus vegetans instead form papillomatous growth and vegetation. It is commonly seen in middle-aged people and the lesions are usually seen on the flexors. However, vegetations can be seen at many sites [2,4].

Case Report

A 24-year-old female known case of oculocutaneous albinism presented with 3 weeks history of painful oral erosions and swellings at the genital area. The problem started 3 weeks ago with painful erosions at the lips, tongue, and oral cavity with the difficulty of swallowing. The patient has been seen by an internal medicine specialist who diagnosed her as having candida infection and prescribed her systemic antifungal agent. The erosions did not show any improvement and she started to develop wet swellings at the groin and around the anus with yellowish oozing. On examination at the dermatology clinic, the girl was white-skinned with blond hair and red eyes that has horizontal nystagmus. She has erosions at the lips, tongue, and buccal mucosa with areas of whitish, non-removable membranes [1-5].

Discussion

At the genitalia, there were two fungating, erythematous, and friable nodules at the right inguinal area and the intergluteal area. There were no

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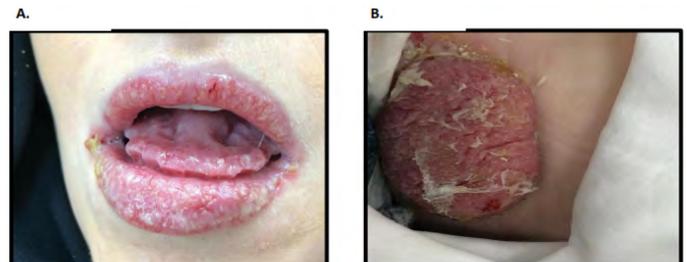


Figure 1. (A) Erosion at the lips and tongue. (B) Vegetative plaque at the groin.

constitutional symptoms or signs. Basic investigations including LFT, RFT, CBC, FBS, VDRL, HIV were within normal limits. Skin biopsies were taken for histopathology and DIF examination, which revealed supra-basilar epidermal cleft with acantholysis and inter-cellular IgG deposition. The clinicopathological correlate on was in favor of pemphigus vegetans. The patient was started on oral prednisolone along with topical tacrolimus and she showed dramatic improvement within two weeks of follow up (Figure 1).

Conclusion

We've reported a very rare case of Pemphigus vegetans in a young girl with oculocutaneous albinism. The diagnosis of such a case may be challenging, so we emphasize that physicians should keep this condition in mind in the clinical setting.

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