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CASE REPORT

# An Unusual Case of Hydroa Vacciniforme with Ocular Involvement

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Hydroa vacciniforme (HV) is a sporadic, rare and idiopathic chronic photodermatosis characterized by recurrent vesicles and crust formation on a sun-exposed skin, typically resulting in vacciniform or varioliform scarring. Herein, we report on a 18-year-old boy who presented with rare ocular involvement in HV. **Key words:** hydroa vacciniforme, corneal opacities, conjunctival hypremia.

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## 1. INTRODUCTION

Hydroa vacciniforme (HV) is a very rare photodermatosis of unknown etiology that is principally seen in childhood. It is characterized by recurrent erythema and crops of papulovesicles and vesicles that appear on uncovered skin areas within 1 to 2 days after sun exposure (1, 2). Ocular manifestations of the disease are uncommon, but can include conjunctivitis, vesicular eruptions of the conjunctiva and cornea that resemble phlyctenular keratoconjunctivitis, corneal infiltration with vascularisation, and keratouveitis (3). In this report, we present a unique case of HV with the manifestations of decreased visual acuity and bilateral corneal opacities.

## 2. CASE REPORT

An 18-year-old male patient was consulted due to complaints of a 10-year history of progressively decreasing vision, and a 14-year history of recurrent wounds and scars over the sun exposed skin areas. His personal and family history was otherwise unremarkable. He was exposed to neither prior trauma nor ophthalmic surgery. The parents were non-consanguineous. Erythema, edema, and hemorrhagic crusting with scarring on the nose, lower lip, hand dorsa, fingers and ears were observed in dermatological examination (Figure 1a and 1b). There were significant corneal opacities and conjunctival hypremia (Figure 2a and 2b). The patient did not give informed consent for a skin punch biopsy of the lesions. No clinical evidence of hepatosplenomegaly and hemolysis was present. The results of laboratory studies were all within normal limits, including the complete blood cell with differential count, platelet count, liver and renal function tests, red blood cell porphyrin levels, fecal porphyrin levels and uroporphyrin levels. Ophtalmological examination revealed bilateral corneal opacities by evaluation with logMAR visual acuity, biomicroscopy, intraocular pressure measurement and fundoscopy. Best-corrected visual acuities (BCVAs) on the decimal scale were 0,1 for both eyes. Due to corneal opacities, optimal fundus examination could not be obtained. Several infectious (viral infections such as measles and other corneal infections) and non-infectious causes (trauma, vitamin A deficiency, surgical procedures, traditional practices, congenital, etc.) for bilateral corneal opacities were investigated; but no evidence of such etiologies were found. Serological, immunological and viral markers including CRP, ANA, Anti-dsDNA, ANCA, RF, Anti-Ro, Anti-La, HSV type 1 and 2 (Herpes simplex virus) IgM and IgG titers were performed; all results were negative or within normal limits. Finally, HV was considered as the major cause of corneal opacity. Keratoplasty was planned as a treatment strategy.

## 3. DISCUSSION

HV, initially described by Bazin in 1862, was overdiagnosed initially because of the terminological confusion and uncertainty concerning the role of porphyrin metabolism in the pathogenesis (4). At that point, some of the cases classified as HV were protoporphyria, which became evident once erythropoietic protoporphyria was clearly defined. HV is a very rare chronic photodermatosis of unknown etiology that principally starts in childhood. In the differential diagnosis; actinic prurigo, polymorphous light eruption, lupus erythematosus, porphyria cutanea tarda pseudoporphyria, herpes simplex, eczema herpeticum, erythema multiforme, and pemphigus erythematosus must be considered. Characteristic distribution of the lesions, age, and limitation of the recurrent attacks to late spring and

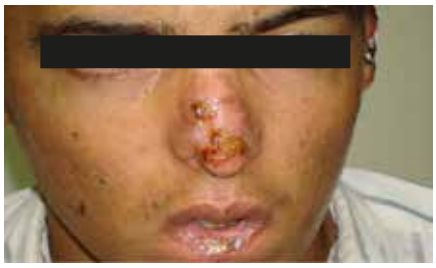

**Figure 1a.** Erythema, edema, and hemorrhagic crusting with scarring on the nose, ear, and lower lip.

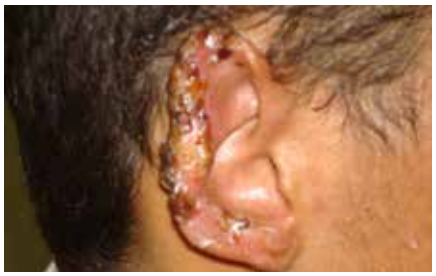

**Figure 1b.** Erythema, edema, and hemorrhagic crusting with scarring on ear.

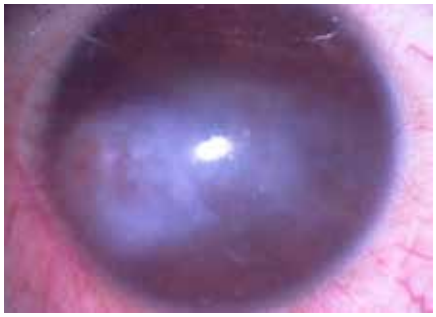

**Figure 2a.** Significant corneal opacity and conjunctival hyperemia (right eye).

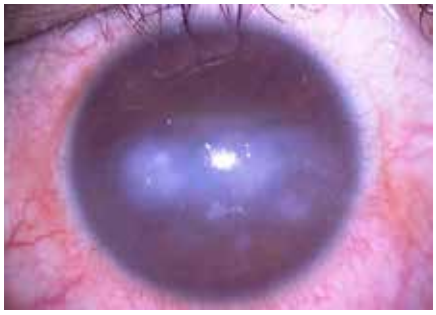

**Figure 2b.** Significant corneal opacity and conjunctival hyperemia (left eye).

summer should give the clinican clues for an easy diagnosis (5).

Ocular involvement occurs occasionally at a later time than cutaneous findings. Ocular complications are rarely observed in HV. In the English literature, ocular manifestations of the disease are uncommon but can include conjunctivitis, vesicular eruptions of the conjunctiva and cornea that resemble phlyctenular keratoconjunctivitis (6), corneal infiltration with vascularisation (7), keratouveitis (8, 9), and interstitial keratitis (10). The differential diagnosis of bilateral corneal opacities is broad and includes infectious and non-infectious causes. In the literature, viral eye infections (measles, etc.), corneal infections, etc. were demonstrated in the infectious etiology of bilateral corneal opacities. The non-infectious etiologic factors include vitamin A deficiency, congenital/neonatal disorders, couching/traditional practices and trauma. The lack of such etiologies in our case suggests HV as a non-infectious cause of bilateral corneal opacities.

In conclusion, it is crucial to consider HV in the differential diagnosis of patients presenting with decreased vision and bilateral corneal opacities, especially in the presence of cutaneous findings such as vesiculaton, crusting and scarring over the sun-exposed skin areas. Further, clinicians should be alert with regard to the risk of ocular involvement in HV and follow up patients closely with regular, fastidious ophthalmological examinations.

**Conflict of interest: none declared.**

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