

Hydroa vacciniforme with ocular involvement: a rare clinical presentation

Hidroa vacciniforme com envolvimento ocular: uma apresentação clínica rara

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Abstract

Hydroa vacciniforme (HV) is a rare photodermatosis in children, characterized by recurrent vesiculopapular eruptions on sun-exposed areas, leaving small, varioliform atrophic scars. Ocular involvement is rare and poorly documented. We describe the case of a 13-year-old boy presenting with bilateral ocular redness and decreased visual acuity associated with photosensitive skin lesions present since the age of 6. Examination revealed numerous atrophic facial scars as well as papulovesicular and crusted lesions on sun-exposed skin. Ophthalmological examination revealed superficial punctate keratitis and keratouveitis with a corneal epithelial ulcer. Epstein-Barr virus serology revealed positive immunoglobulin G (IgG) antibodies and negative IgM antibodies. The diagnosis of HV was confirmed by skin biopsy. This case highlights a rare complication of HV and underscores the importance of a multidisciplinary approach involving dermatologists and ophthalmologists. Early detection of ocular involvement can prevent long-term visual impairment and ensure optimal patient management.

Keywords: Hydroa vacciniforme. Kerato-uveitis. Ocular inflammation. Pediatric. Photodermatosis. Photosensitivity.

Resumo

A hidroa vacciniforme (HV) é uma fotodermatose rara em crianças, caracterizada por erupções vesiculopapulares recorrentes em zonas expostas ao sol, deixando pequenas cicatrizes atróficas varioliformes. O envolvimento ocular é raro e pouco documentado. Descrevemos o caso de um rapaz de 13 anos que apresentava vermelhidão ocular bilateral e diminuição da acuidade visual associadas a lesões cutâneas fotossensíveis presentes desde os 6 anos de idade. O exame revelou numerosas cicatrizes atróficas na face, bem como lesões papulovesiculares e crostosas na pele exposta ao sol. O exame oftalmológico revelou queratite puntata superficial e queratouveíte com úlcera epitelial da córnea. A serologia para o vírus Epstein-Barr (EBV) revelou anticorpos IgG positivos e anticorpos IgM negativos. O diagnóstico de hidroa vacciniforme foi confirmado por biópsia cutânea. Este caso evidencia uma complicação rara da hidroa vacciniforme e sublinha a importância de uma abordagem multidisciplinar que envolva dermatologistas e oftalmologistas. A deteção precoce do envolvimento ocular pode prevenir a deficiência visual a longo prazo e garantir uma gestão ideal do doente.

Palavras-chave: Hidroa vaciniforme. Ceratouveíte. Inflamação ocular. Pediátrico. Fotodermatose. Fotossensibilidade.

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Received: 23-02-2026

Accepted: 06-03-2026

DOI: 10.24875/PJDV.26000027

Available online: 09-04-2026

Port J Dermatol and Venereol. (ahead of print)

www.portuguesejournalofdermatology.com

Introduction

Hydroa vacciniforme (HV) is a rare childhood photosensitivity disorder characterized by pruritic or painful vesicles in photo-distributed areas. It is thought to be a disease of early childhood that disappears spontaneously in adolescence. These lesions usually occur after sun exposure and typically heal with permanent varioliform scarring. Ocular manifestations are uncommon, but may include conjunctivitis, vesicular eruptions of the conjunctiva and cornea that resemble phlyctenular keratoconjunctivitis, corneal infiltration with vascularization, and kerato-uveitis^{1,2}. Epstein-Barr virus (EBV) infection may be linked to HV, according to recent studies. However, the role of EBV in the pathogenesis is uncertain, but it is thought that EBV-induced immune dysregulation could contribute to the development of the disease³. In this report, we present a rare case of HV with the manifestations of decreased visual acuity and bilateral ocular redness, highlighting the importance of collaboration between dermatologists, ophthalmologists, and pathologists to establish a rapid diagnosis, which can help minimize esthetic and functional damage by initiating appropriate treatment early.

Observation

We report the case of a 13-year-old boy with no medical history, followed in ophthalmology for kerato-uveitis of unknown etiology, referred to our dermatology department for pruritic skin lesions on the face and hands, which appeared at the age of six. His mother reported marked photosensitivity with remission during the winter months. Clinical examination revealed multiple papulovesicular lesions, some umbilicated and others crusted, located on the back of the hands (Fig. 1A). Numerous varioliform atrophic scars were observed on the face (Fig. 1B). The patient also presented with conjunctival hyperemia and decreased visual acuity. The other mucous membranes were spared, and there was no lymphadenopathy, systemic signs, erythrodontia, hypertrichosis, or family history of similar cases.

Ophthalmological examination revealed a 2-mm epithelial corneal ulcer, corneal edema, and diffuse superficial punctate keratitis (Fig. 2).

Complete blood count, autoimmune panel, and porphyrin levels were all within normal limits, and the serological tests requested by the ophthalmologists were negative. EBV serology revealed positive immunoglobulin G (IgG) antibodies and negative IgM; however,

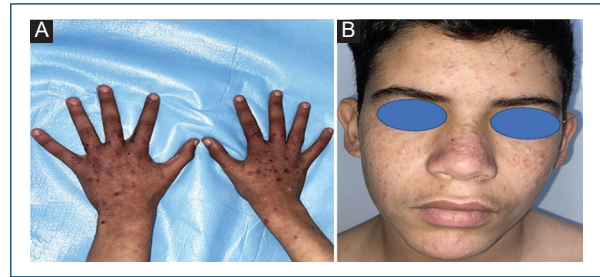


Figure 1. Clinical images **A:** clinical presentation of hydroa vacciniforme showing papulovesicles with an umbilicated surface and hemorrhagic crusts of the hands. **B:** varioliform atrophic scars of the cheeks and nose.

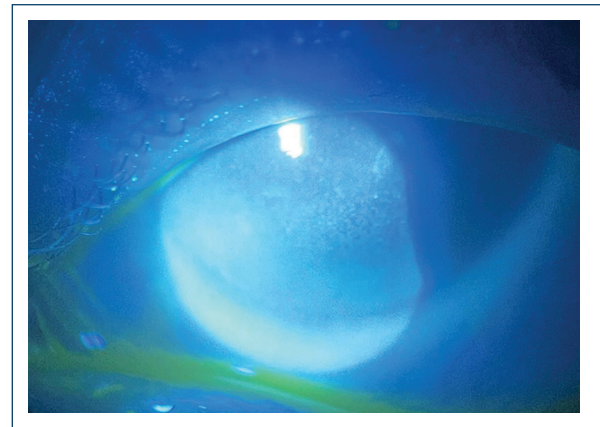


Figure 2. Fluorescein staining under cobalt blue light reveals a 2 mm epithelial corneal ulcer with diffuse superficial punctate keratitis.

polymerase chain reaction was not performed. A skin biopsy showed an acanthotic epidermis with spongiosis and a vesicle containing degenerated neutrophils. The dermis showed an inflammatory infiltrate composed of small lymphocytes with foci of suppuration (Fig. 3). Immunohistochemistry revealed a predominance of T cells (CD7-positive), with rare B-cell infiltrates (PAX5-positive). The T-cell infiltrate expressed both CD4 and CD8, with a slight predominance of cytotoxic CD8+ lymphocytes. These findings were consistent with HV, ruling out NK/T-cell lymphoma, discoid lupus erythematosus, and porphyria cutanea tarda (PCT). Unfortunately, due to financial limitations, chromogenic *in situ* hybridization analysis for EBV DNA could not be performed.

The patient was treated with a potent topical corticosteroid and strict photoprotection, combining both

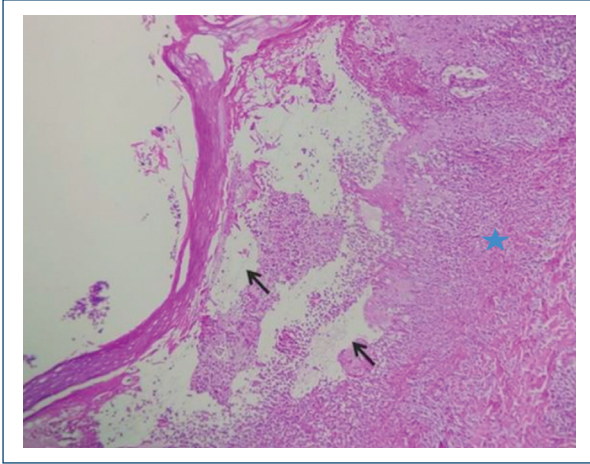


Figure 3. Spongiosis with a vesicle containing altered neutrophils (arrows), inflammatory infiltrate of the dermis composed of small lymphocytes with foci of suppuration (star) (H&E, $\times 100$ magnification).

chemical and physical measures, in addition to topical corticosteroid eye drops, oral acyclovir, and ophthalmologic follow-up. After 3 months, clinical reassessment showed resolution of the cutaneous lesions and improvement of visual acuity.

Discussion

HV is an extremely rare photosensitivity dermatosis first described by Bazin in 1862. It is considered a childhood disorder, occurring more frequently in males than in females with fair skin phototypes, with two age peaks: early childhood (1-7 years) and post-puberty (12-16 years). Its onset is typically seasonal, appearing in spring or summer. The prevalence is estimated at 0.1-0.5 cases/100,000 inhabitants/year^{2,3}.

The pathophysiology of HV remains poorly understood. Ultraviolet A (UVA) radiation, a major component of sunlight, appears to play a critical role, as the characteristic lesions of HV can be experimentally reproduced following artificial UVA exposure⁴. Furthermore, several studies have demonstrated the presence of EBV within the lymphocytic infiltrate of HV lesions in both pediatric and adult patients. High levels of EBV DNA in peripheral blood, along with the detection of EBV-encoded small RNAs in skin lesions, further support the implication of EBV in the pathogenesis of this photodermatosis⁵.

Classic HV typically presents as recurrent erythematous papules and vesicles associated with pruritus,

and occasionally accompanied by a tingling sensation⁶. These eruptions appear within hours to days following sun exposure, predominantly affecting sun-exposed areas, such as the face and the dorsal surfaces of the hands in a bilateral and symmetrical distribution. The lesions often evolve into ulcerations covered with necrotic crusts and subsequently heal over a period of 1-6 weeks, leaving behind varioliform, atrophic scars⁷.

Among the principal differential diagnoses of HV, discoid lupus erythematosus should be considered. This condition typically affects young to middle-aged adults, with predominance in women, and may be associated with systemic manifestations. Clinically, it presents as erythematous plaques with atrophic centers and adherent scales on photo-exposed sites, particularly the nose, cheeks, and ears. Histopathological findings commonly include hyperkeratosis, follicular plugging, basal vacuolar degeneration, and peri-adnexal as well as perivascular inflammatory infiltrates³.

PCT is another important differential diagnosis. It is characterized by photosensitivity and cutaneous fragility, predominantly involving the dorsum of the hands and the face. The lesions manifest as vesicles and bullae that progress to erosions, followed by dystrophic scarring with skin atrophy and the formation of milia⁸. Additional signs may include malar hypertrichosis, heterogeneous hyperpigmentation, and scleroderma-like changes, although these are rarely seen in children. The biochemical diagnosis of PCT is established by demonstrating elevated urinary levels of uro- and coproporphyrins⁹.

Ocular involvement in HV is uncommon and generally develops later than the cutaneous manifestations. Reported ophthalmic complications are rare, with the English literature describing only a few cases. Documented findings include conjunctivitis, vesicular eruptions of the conjunctiva and cornea mimicking phlyctenular keratoconjunctivitis, corneal infiltration with vascularization, and kerato-uveitis, as we found in our case¹⁰.

Indeed, based on the characteristic anamnestic, clinical, and histological features of HV, and after excluding any infectious or autoimmune cause of the ocular involvement, as confirmed by the ophthalmologists, the diagnosis of HV complicated by kerato-uveitis was established.

Photoprotection is the cornerstone of HV management. It involves avoiding sun exposure during peak hours (11:00 AM-3:00 PM) and adopting protective clothing measures, such as wearing thick fabrics, long sleeves, long trousers, a wide-brimmed hat, and sunglasses with 70% tint, which filter out both UVA and UVB

light, to reduce the risk of ocular complications. Using a broad-spectrum sunscreen with an SPF of 30-50 that provides effective UVA protection is also essential. A generous amount of sunscreen should be applied 15-30 min before sun exposure, then reapplied every 2 h, and immediately after swimming or towel drying. Vitamin D supplementation is also recommended due to the risk of deficiency associated with strict photoprotection^{11,12}.

In moderate to severe forms, prophylactic desensitization phototherapy with TL01 at the beginning of spring may be considered for cases that do not respond adequately to broad-spectrum sunscreens and behavioral sun avoidance¹³.

Other systemic treatments may be proposed for resistant cases, including azathioprine, thalidomide, cyclosporine, and hydroxychloroquine, whose use is restricted due to the ocular risk².

In addition, several case reports have highlighted that oral treatment with acyclovir or valacyclovir may have a beneficial effect on HV symptoms¹⁴.

Conclusion

HV is a rare chronic photodermatosis that typically affects children and adolescents. Although it usually presents with vesiculopapular eruptions on sun-exposed skin, ocular involvement may occur and compromise visual prognosis. This case highlights the critical need for a multidisciplinary approach, particularly the collaboration between dermatologists and ophthalmologists, to ensure early detection of ocular involvement, prevent complications, and provide comprehensive management tailored to both the cutaneous and ocular manifestations of the disease.

Funding

None.

Conflicts of interest

None.

Ethical considerations

Protection of human subjects and animals. The authors declare that no experiments on humans or animals were performed for this research.

Confidentiality, informed consent, and ethical approval. The authors have followed their institution's confidentiality protocols, obtained informed consent from all patients, and secured approval from the Ethics Committee. SAGER guidelines have been followed as applicable to the nature of the study.

Declaration on the use of artificial intelligence (AI). The authors declare that no generative artificial intelligence was used in the writing or creation of the content of this manuscript.

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