Unusual association of diseases/symptoms

Anterior keratouveitis secondary to Hydroa vaccineforme: a role for ophthalmic slit-lamp examination in this condition?

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The authors report the case of an 8-year-old girl of Caucasian origin who attended the Emergency Eye Clinic with a 3-week history of a red, light sensitive left eye during the month of April. Her Snellen visual acuities were 6/5 right and 6/9 left. Examination revealed perilimbal injection and anterior chamber inflammation in the left eye consistent with an anterior uveitis. Inferior stromal haze and fine keratic precipitates were noted in the left cornea. Intraocular pressures were normal, with no vitritis and healthy looking discs, maculae and peripheral retina. Systemic examination revealed a papular rash over the face consistent with previously diagnosed Hydroa vaccineforme. With intensive topical steroids the inflammation gradually resolved.

BACKGROUND

- A rare dermatology case with ophthalmic sequelae.
- Of interest across both dermatology and ophthalmology disciplines.

CASE PRESENTATION

We report the case of an 8-year-old girl of Caucasian origin who attended the Emergency Eye Clinic with a 3-week history of a red, light sensitive left eye during the month of April. Her Snellen visual acuities were 6/5 right and 6/9

left. Examination revealed perilimbal injection and anterior chamber inflammation in the left eye consistent with an anterior uveitis. Inferior stromal haze and fine keratic precipitates were noted in the left cornea (figure 1). Intraocular pressures were normal, with no vitritis and healthy looking discs, maculae and peripheral retina. Systemic examination revealed a papular rash over the face (figure 2) consistent with previously diagnosed Hydroa vaccineforme (HV). With intensive topical steroids the inflammation gradually resolved.

Figure 1 Anterior segment photo demonstrating inferior stromal haze and very fine keratic precipitates.

Figure 2 Hydroa vaccine-forme: recurrent vesicles are evident on sun-exposed areas, healing with vaccine-forme scarring.

INVESTIGATIONS

Previous skin biopsies confirmed the diagnosis of HV.

TREATMENT

Topical steroids – uveitis resolved.

OUTCOME AND FOLLOW-UP

Anterior uveitis settled with topical steroid use. Currently under a 6-monthly review.

DISCUSSION

HV is an uncommon photodermatosis, primarily affecting areas of skin exposed to sunlight, particularly the face and ears.¹ It mainly affects Caucasians, with a male:female ratio of 2:1,¹ and prevalence has been estimated as approximately 0.34 per 100 000.²

The condition is characterised by an itchy vesiculopapular rash which heals with progressive keratinocyte necrosis and scarring.¹ Onset is during childhood and tends to recur, especially during summer, with remission usually by adolescence.

HV may rarely be associated with ocular findings, which are usually limited to mild photophobia and keratoconjunctivitis, as in this case. HV presenting initially with ocular signs has been documented only once previously, although in this case the diagnosis preceded the ocular pathology.³

HV is a result of photo-allergic sensitivity, whereby sunlight converts a proantigen present in tissues into an antigen which triggers antibody formation.⁴ While the exact pathophysiology remains unclear, activation of latent Epstein-Barr virus (EBV) has also been implicated.^{5 6} There was no evidence of EBV RNA in the sample of this patient.

This theory is supported by the fact that ocular signs usually occur after the initial skin presentation, as the

eyelids confer a degree of protection. The heterogeneity of HV presentation may be due to the variable distribution in body tissues of the proantigen, subsequently explaining such cases where ocular signs precede the dermatological manifestations of HV.

While consensus exists regarding the role of ultraviolet (UV) light in the pathogenesis of HV, both UVA and UVB have been implicated in having a role in the underlying aetiopathogenesis.^{7 8} Sunlight avoidance can be achieved with clothing which affords more protection and using sunscreen on areas which remain exposed. The use of sunglasses with 70% tint which filter out both UVA and UVB light will reduce the risk of ocular complications. Oral steroids and, in the case of EBV infection, ganciclovir have a suggested role in treatment.³

Although ocular pathology in HV is generally mild – corneal ulcers, conjunctivitis, corneal opacities, uveitis, severe chemosis and scleritis can occur.^{1 4 9 10} Furthermore, lesions on the eyelids can lead to cicatricial ectropion.⁴ As the ocular associations of HV are now well recognised, and with the extensive corneal clouding picked up on examination in this case, examination by an ophthalmologist in patients with HV may be appropriate to limit long-term sequelae.

Learning points

- ▶ HV is a rare dermatological condition.
- ▶ In such patients, an urgent ophthalmic referral is warranted to exclude anterior uveitis or corneal disease.
- ▶ The use of 70% tint glasses may be used to prevent recurrences of uveitis.

Competing interests None.

Patient consent Obtained.

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