Disseminated mucocutaneous herpes simplex in an atopic individual

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We report a case of keratoconjunctivitis associated with eczema herpeticum and highlight its resemblance to erythema multiforme major.

Case history

A 30-year-old Caucasian man was referred to eye casualty by his general practitioner (GP) with a two-day history of a widespread, rapidly progressing, blistering rash with ulceration, along with red painful eyes. It started with a ‘flu-like’ illness with symptoms of malaise and arthralgia for which the patient self-medicated with ibuprofen. After one day the rash began on the shaft of the penis and groin with painful vesicles that caused gross oedema. He had no genital mucosal involvement and denied dysuria or purulent discharge. Within several hours widespread painful, punched-out ulcers developed over his face and anterior thoracic wall. The individual was known to suffer from asthma and eczema. Past ophthalmic history revealed previous penetrating keratoplasty in the right eye and left cataract surgery without any lens implant in the left eye.

On examination he had an acute gingivostomatitis with blistering around the mouth and ulcerating lesions of the buccal mucosa, tongue and lips. Unaided visual acuities were 6/9 in both eyes. A punched-out rash was distributed over his sternum and he had a marked eczematous blepharitis with a papillary conjunctivitis (Figure 1). No symblephara formation was noted. Corneal punctate epithelial erosions were noted but anterior chamber examination did not reveal any abnormality. The intraocular pressures were normal. Fundoscopy was unremarkable. The individual had unprotected sexual intercourse with a new partner one week prior to the development of the rash. He denied any previous episodes of herpes simplex viral infection and had no previous high-risk behaviours such as intravenous drug use. Subsequent HIV testing was negative. Blood tests, including liver function tests, were normal. Polymerase chain reaction on a conjunctival swab was positive for herpes simplex virus (HSV) type 1.

The provisional diagnosis of Stevens-Johnson syndrome was made. He was treated initially with chloramphenicol ointment four times a day, hourly celluvisc 0.5% and two-hourly dexamethasone 0.1% eye drops. The patient was admitted under the medical team and subsequently reviewed by dermatology and the genitourinary medicine team the following day who clinically diagnosed disseminated mucocutaneous herpes simplex infection, also known as eczema herpeticum or Kaposi’s varicelliform eruption. His eye drops were subsequently changed to 3% aciclovir ointment five times a day, having received the above treatment for one day. He was treated with intravenous aciclovir 5 mg/kg three times a day for two days. Following a good response to the treatment he was discharged home with oral valaciclovir 1g PO for 10 days.

Discussion

Herpes simplex infection has a wide range of clinical manifestations, from minor localized oral or genital disease to disseminated systemic infection. Between 80–90% of the European population is infected with HSV type 1, with infection mainly being acquired in childhood.¹ Although dissemination of HSV infection is rare, widespread
mucocutaneous involvement can occur particularly in the immunocompromised, or in individuals with pre-existing atopic eczema. Dissemination in the latter, as in this case, is known as eczema herpeticum or Kaposi’s varicelliform eruption.² Disease severity varies, but it is usually cutaneous and mild and will respond well to treatment with antiviral agents such as aciclovir and valaciclovir.³ However, dissemination with visceral involvement can occur, causing complications such as a necrotizing encephalitis, hepatitis and mortality.¹ Disseminated mucocutaneous herpes simplex has also been reported widely in individuals with impaired immunity such as pregnancy (especially third trimester), corticosteroid therapy and other immunosuppressive drug use, haemopoietic disorders, bone marrow and solid organ transplants, neonates,³ cancer patients and burn victims.⁴ Acute ocular involvement beyond a blepharitis is uncommon⁵ but herpetic keratitis has been reported.⁶,⁷ Ocular manifestations reported in individuals with eczema herpeticum include a tendency for bilateral involvement, marked ocular discomfort, papillary conjunctivitis, and extensive corneal epithelial disease such as multiple dendritic ulcers and punctuate epitheliopathy.⁸ Other ocular conditions associated with eczema include blepharitis, atopic keratoconjunctivitis, cataracts and keratoconus,⁸ all of which have affected our patient previously.

Ocular complications, such as keratoconjunctivitis in disseminated herpes simpex have a low prevalence.⁵ The condition of disseminated mucocutaneous herpes simplex with ocular involvement can resemble to a certain degree erythema multiforme major and the ophthalmologist should consider it as a possible diagnosis in individuals presenting with an acute blistering dermatosis, especially in those with atopic eczema or other forms of immunocompromise. Any diagnostic error may result in inappropriate treatment. HSV-associated erythema multiforme in children has similarly previously been highlighted as a possible source of diagnostic error for Stevens-Johnson syndrome.⁹

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