Table 1 A comparison of ocular features of congenital varicella syndrome and herpes zoster ophthalmicus

Ocular features of congenital varicella syndrome ^{10 17 18}	Ocular features of herpes zoster ophthalmicus ²¹
Cicatrising dermatomal skin lesions	Cicatrising dermatomal skin lesions and ptosis
Chorioretinitis	Mucopurulent conjunctivitis
Optic atrophy	Episcleritis and scleritis
Anisocoria	Corneal anaesthesia
Nystagmus	Recurrent keratitis
Microphthalmia	Uveitis
Cataract	Iris atrophy
Corneal opacity	Glaucoma
Heterochromia	Ciliary ganglionitis causing abnormal pupil response
	Optic neuritis

Ophthalmologists will be interested in another distinct teratogenic effect of varicella zoster. The virus once acquired in pregnancy is able to cross the placenta causing systemic and ocular problems in the offspring. This association was first noted in 1947¹⁵ and has subsequently been recognised as the congenital varicella syndrome or the fetal varicella syndrome.^{10 16-18} The ocular associations of congenital varicella and shingles are depicted in Table 1. The case we have described does not show many of the ocular findings in congenital varicella syndrome but does illustrate many of the clinical features of adult herpes zoster ophthalmicus: a dermatomal skin lesion in the distribution of the ophthalmic division of the trigeminal nerve, absent corneal sensation, recurrent keratitis and uveitis, iris atrophy, and abnormal direct pupil response. Other conditions that may cause skin defects are direct pressure on the fetus in utero or cutaneous polyarteritis, a rare vasculitis in children.¹⁹ However, the dermatomal distribution and ocular features make these latter two conditions unlikely.

In conclusion, we suspect that the mother's varicella infection in pregnancy and the child's congenital conditions are causally related. The

varicella zoster virus probably crossed the placenta to lie dormant in the child's trigeminal ganglion. At some time before birth the virus was reactivated²⁰ and led to this unique case of 'presumed herpes zoster ophthalmicus' in utero which was present at birth. The clinical findings are so unusual and similar to adult herpes zoster ophthalmicus, that this seems the most likely explanation.

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Orbital multiple myeloma mimicking acquired angio-oedema

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Multiple myeloma is characterised by malignant plasmacytes in the bone marrow and excessive production of specific immunoglobulins or immunoglobulin components. Ocular involvement may result from myelomatous infiltration or may occur secondarily to haematological abnormalities. Virtually any ocular structure can be involved, including the conjunctiva, cornea, sclera, lens, retina, uveal tract, optic nerve,

lacrimal glands, and orbit.¹ Involvement of periorbital skin is unusual. To our knowledge this is the first reported case of metastatic multiple myeloma presenting as periorbital swelling with angio-oedema-like features.

Case report

A 58-year-old white woman presented to her

Figure 1 Angio-oedema of right upper eyelid. Note absence of involvement of right lower eyelid.

ophthalmologist with mild erythema and oedema of her upper right eyelid. A presumptive diagnosis of an allergic process was made during the initial examination and she was treated with oral antihistamines. Over the next 2 weeks the right upper eyelid oedema and erythema progressively increased. There were no associated symptoms of pain, epiphora, or pruritus. The medical history was significant for IgA κ multiple myeloma stage III, for which she was currently undergoing her third month of interferon alfa therapy. No history of trauma or infection was noted. Two weeks after the initial development of eyelid oedema, the patient was seen by her oncologist and immediately referred for dermatological and a separate ophthalmic examination.

On examination, the right upper eyelid was markedly oedematous with a yellow-orange translucent appearance (Fig 1). The best corrected visual acuity was 20/400 in the right eye and 20/20 in the left eye. The right pupil was 3 mm, sluggishly reactive, and had a 2+ afferent pupillary defect. Motility of the right eye was decreased in all fields of gaze (Fig 2). The conjunctiva had clear chemosis on the right side. The remainder of the anterior segment examination was normal. Examination of the fundus showed multiple flame-shaped retinal haemorrhages without disc oedema. Hertel exophthalmometry showed 6 mm of proptosis on the right. The right globe was inferiorly displaced by 8 mm. Examination of the left eye and periorbital tissue was normal. Computed tomography (CT) examination of the orbits demonstrated an intraconal and extraconal infiltrating mass (Fig 3).

Figure 2 Pitting oedema of right upper eyelid and right hypotropia.

Figure 3 Axial CT scan demonstrating diffuse infiltration of the intraconal space, extraconal space, and eyelid on the right side.

There was no evidence of orbital bone involvement. Innumerable small lytic foci in the calvarium were consistent with diffuse involvement by multiple myeloma.

A diagnosis of metastatic multiple myeloma was made based on the radiological studies and the clinical history. The patient received radiation therapy (2500 Gy in 10 treatments over 14 days). One month after radiation therapy, the visual acuity of the right eye improved to 20/40 with marked regression of eyelid oedema and proptosis (Fig 4).

Comment

Specific cutaneous manifestations of multiple myeloma include extramedullary plasmacytomas of the skin and mucous membranes or, more commonly, may result from direct extension of primary bone lesions. Non-specific cutaneous manifestations include primary amyloidosis, changes secondary to cryoglobulins, changes secondary to anaemia, leucopenia, or thrombocytopenia, toxic eruptions, and changes secondary to internal organ involvement.^{2,3}

Ocular manifestations may also be due to direct infiltration of the neoplasm (primary or metastatic) or from associated haematological changes. Progressive proptosis is occasionally the initial presentation of multiple myeloma, and is a common symptom when there is any involvement of the eye. Other symptoms include diplopia, pain, and visual loss. Signs of ocular involvement consist of large dilated fundal veins with flame-shaped and punctate haemorrhages. Cysts of the ciliary body and pars plana are also

Figure 4 One month post-radiation treatment to the right orbit. Note the marked reduction in angio-oedema of the right eyelid and partial resolution of the right hypotropia.

seen.^{4,6} Although the periorbital skin is rarely involved, Gudas reported a case presenting as periorbital cellulitis.⁷ The infection was presumed to be secondary to the patient's compromised immunological status.

Our patient presented with metastatic multiple myeloma and periorbital findings consistent with angio-oedema. This has not been described in the literature previously. Acquired forms of angio-oedema are known to be induced by physical agents, C1 esterase inhibitor autoantibodies, excessive consumption of C1 esterase inhibitor in lymphoproliferative disorders, and drug-induced inhibition of degradation of kinin-like products of C1 esterase.⁸ Acquired angio-oedema has not been reported as a side effect of interferon alfa therapy and is unlikely to be the aetiology in this case. We presume the angio-

oedema-like features in our patient were secondary to the mass effect of the retro-orbital tumour causing obstruction of venous and lymphatic drainage from the periorbital skin.

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