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Sir,

Bilateral periocular swelling in Sweet's syndrome
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Sweet's syndrome is an uncommon skin disorder of unkown aetiology characterised by painful erythematous plaques, and associated with pyrexia, marked leucocytosis and elevated erythrocyte sedimentation rate (ESR). Although originally described as affecting the arms and legs, it has been reported on the head and neck. Periocular presentations are unusual and particularly associated with escharotic lesions. We report a case with marked bilateral periocular swelling.

Case report

A 72-year-old man developed bilateral periocular swelling overnight. He gave no history of facial trauma, sinus disease, or allergy. He denied diplopia, or visual loss. His past medical history included ischaemic heart disease for which he took oral diltiazem, bumetanide, elantan LA, aspirin, and sublingual glyceryl trinitrate. On admission, the patient was pyrexial with a temperature of 38.8°C and dyspnoeic with ankle swelling. He had gross bilateral periocular swelling and vesicular exudative lesions of the eyelids (Figure 1). There was no lymphadenopathy, or skin involvement other than the periocular area.

The corrected near vision was N8 right and left. Pupil reactions were brisk and normal. Motility was restricted particularly on the left and in all positions of gaze with mild proptosis. The anterior and posterior segments were healthy.

Clinically he was treated for probable Herpes Zoster cellulitis with secondary bacterial infection. In view of his pyrexia and likely cardiac failure, a medical opinion was sought and the following investigations were requested: ESR 100 mm/h (<20), elevated white cell count (WCC) 13.9 × 10/l (4–16), C-reactive protein 638 mg/l (<10), random glucose 25.6 mmol/l (4.5–5.6), urea 17 mg/dl (3–7), creatinine 172 μ/l (60–110), sodium 124 mmol/l (135–146), potassium 4.6 mmol/l (3.5–5.0), blood cultures that subsequently were negative and a chest X-ray (CXR) showed an enlarged heart and pulmonary oedema.

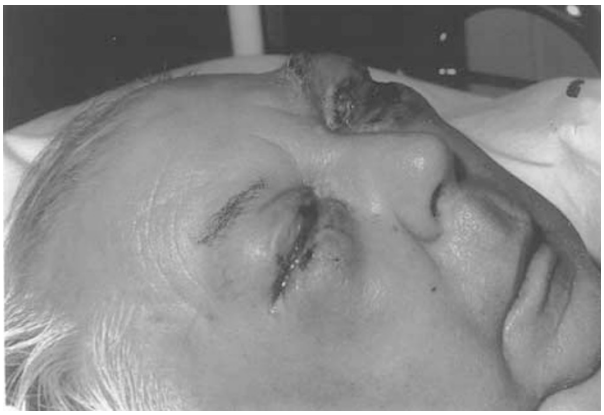

Figure 1 Bilateral periocular swelling with exudtive vesicles more marked on the left.

Intravenous insulin and frusemide were commenced to control his diabetes and heart failure. His cellulitis was treated with intravenous acyclovir and benzylpenicillin with flucloxacillin.

After 24 h, computed tomography (CT) was arranged of the orbits, paranasal sinuses, and brain in order to exclude orbital cellulitis, subperiosteal abscess, cavernous sinus thrombosis, or mucormycosis. The CT demonstrated only preseptal ocular and facial swelling (Figure 2).

Over the next 48 h the diffuse swelling subsided but the crusting became haemorrhagic and necrotic particularly over the left upper lid (Figure 3). A dermatology opinion suggested a dermatosis, and a skin biopsy was taken of the left upper lid.

Pathologic examination demonstrated that the epidermis was uninvolved but there was a dense polymorph infiltration of the entire thickness of the dermis (Figure 4). The blood vessels demonstrated swelling of the endothelial cells but no evidence of a true vasculitis (Figure 5, arrow). This suggested a diagnosis of Sweet's syndrome, or acute neutrophilic dermatosis.

He was commenced on oral prednisolone 30 mg daily and made a rapid recovery.

Comment

Sweet's syndrome, also known as acute neutrophilic dermatosis, was first described by Dr Robert D Sweet in 1964.[1] It is characterised by abrupt onset fever, painful erythematous skin papules and plaques, and

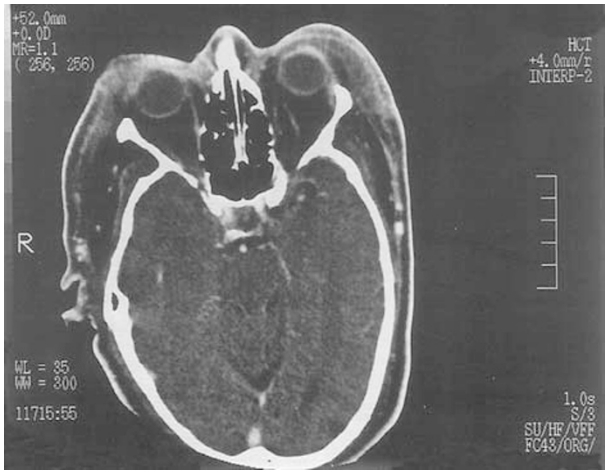

Figure 2 CT demonstrates preseptal and facial swelling.

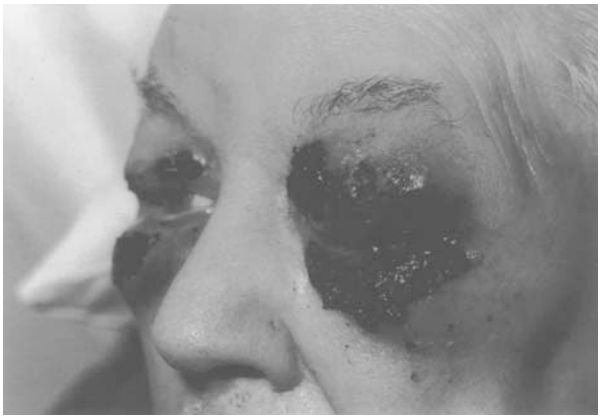

Figure 3 Crusting escharotic appearance especially on the left.

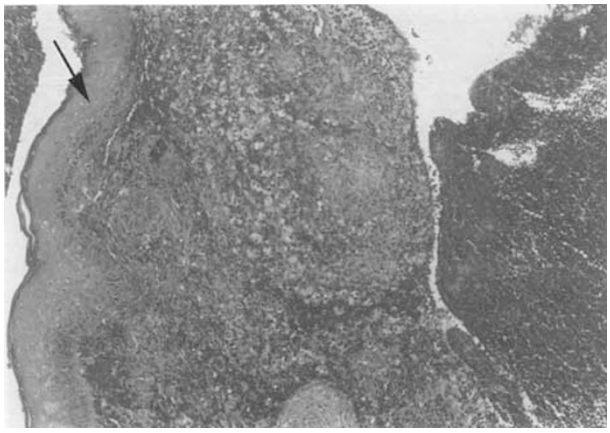

Figure 4 Unremarkable epidermis (black arrow) with haemorrhagic dermis infiltrated by polymorphs.

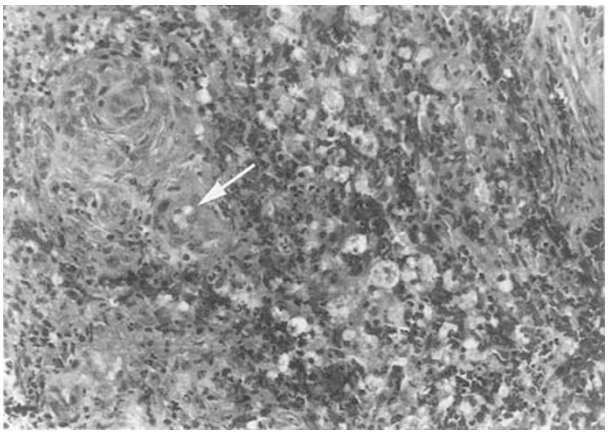

Figure 5 Arrow shows swollen endothelial lining, but lack of inflammatory changes of blood vessels denoting absence of a true vascilities.

neutrophilic leucocytosis. It may be idiopathic, drug induced,[2] associated with haematological malignancy or immunologic conditions such as rheumatoid arthritis, and inflammatory bowel disease.

The main histological feature is the infiltration of the dermis by mature neutrophils and absence of vasculitis although in lesions of longer duration a secondary vasculitis may develop.[3]

It is important to note that the diagnosis of Sweet's syndrome is based on clinical suspicion. von den Driesch[4] has proposed diagnostic criteria which must include typical skin lesions and histology accompanied by other features. In atypical cases, the diagnostic features include a markedly raised ESR, leucocytosis, and pyrexia but no evidence of infection. Pathologic investigation is invaluable in making the diagnosis since up to a third of cases will have recurrent episodes.

Various case reports of ocular manifestations have been described such as scleritis.[5] One report is of a 40- year- old man presenting with an erythematous vesiculopustular facial eruption that had prominent black escharotic crust particularly over the periorbital area.[6] This was identical to our patient except that marked preseptal swelling was absent and in our patient the escharotic appearance developed during the course of the disease.

Two interesting ocular manifestations have been described in patients with systemic diseases presenting with Sweet's syndrome. Wilson *et al*[7] describe a patient with rheumatoid arthritis who had separate episodes of Sweet's syndrome, pyoderma gangrenosum, pustular vasculitis, and peripheral ulcerative keratitis (PUK). While PUK is a recognised entity in association with rheumatoid arthritis alone, they comment on the

inflammatory spectrum of these conditions and the effective response to cyclosporine.

Morgan and Callen[8] report a 38-year-old man with acute myelogenous leukaemia presenting with a left cellulitis and later with haemorrhagic pustules on his hands. Biopsy revealed a neutrophil infiltrate of the dermis.

The pathogenesis of Sweet' syndrome is unknown. Gassuddin *et al*[9] propose an immunologic mechanism in which there is an imbalance between T helper cell types and cytokine secretion. The release of cytokines occurs in response to an unidentified trigger that results in chemoattraction of leucocytes. This may explain the rapid response to immunosuppressive therapy.

This unusual skin condition should be considered in all cases of periorbital oedema of unknown cause.

References

1 Sweet RD. An acute febrile neutrophilic dermatosis. *Br J Dermatol* 1964; **76**: 349–356.
2 Gilmour E, Chalmers RJ, Rowlands DJ. Drug-induced Sweet's syndrome (acute febrile neutrophilic dermatosis) associated with hydralazine. *Br J Dermatol* 1995; **133**: 490–491.
3 Malone JC, Slone SP, Wills-Frank LA *et al*. Vascular inflammation (vasculitis) in Sweet syndrome: a clinicopathologic study of 28 biopsy specimens from 21 patients. *Arch Dermatol* 2002; **138**: 400–403.
4 von den Driesch P. Sweet's syndrome (acute febrile neutrophilic dermatosis). *J Am Acad Dermatol* 1994; **31**: 535–535.
5 Chen TC, Goldstein DA, Tessler HH *et al*. Scleritis associated with acute neutrophilic dermatosis (Sweet's syndrome). *Br J Ophthalmol* 1998; **82**: 328–329.
6 Bulengo-Ransby SM, Brown MD, Dubin HV *et al*. Sweet's syndrome presenting as an unusual periorbital eruption. *J Am Acad Dermatol* 1991; **24**: 140–141.
7 Wilson DM, John GR, Callen JP. Peripheral ulcerative keratitis—an extracutaneous neutrophilic disoder: report of a patient with rheumatoid arthritis, pustular vasculitis, pyoderma gangrenosum, and Sweet's syndrome with excellent response to cyclosporine therapy. *J Am Acad Dermatol* 1999; **40**: 331–334.
8 Morgan KW, Callen JP. Sweet's syndrome in acute myelogenous leukaemia presenting as periorbital cellulites with an infiltrate of leukaemic cells. *J Am Acad Dermatol* 2001; **45**: 590–595.
9 Gasuddin ASM, El Orfi AHAM, Ziu MM *et al*. Sweet's syndrome: is the pathogenesis mediated by helper T cell type 1 cytokines?. *J Am Acad Dermatol* 1998; **39**: 940–943.

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Sir,

Optic disc haemorrhage following frontal head trauma
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Optic neuropathy following head trauma usually occurs secondary to contusion of the optic nerve sheath, impaled bony fragments in the optic canal, or optic nerve oedema with intracanalicular compression and orbital deformity. Rarely direct ocular trauma can produce a sufficient contrecoup or shearing force to avulse the optic nerve from the globe.[1] We present a case of post-traumatic optic disc haemorrhage following trivial head trauma.

Case report

A 66-year-old lady presented to the eye casualty complaining of decrease in visual acuity in her right eye following head trauma. The night before her presentation she banged her right forehead on the doorframe on her way indoors from the garden. She immediately developed a right periorbital bruise and was unable to open the eye. There was no associated headache or symptoms suggestive of subarachnoid haemorrhage. Next morning on waking when she tried opening her right eye, she noticed blurred vision in it. Past medical history did not suggest any systemic or ocular predisposing factors for occurrence of optic disc haemorrhage.

On ocular examination, the visual acuity in her right eye was 6/18 not improving with pin hole and 6/6 in the left eye. Right eye examination revealed a nontense periorbital haematoma, relative afferent pupillary defect, and reduced colour vision on Ishihara plates. The ocular movements were full. Rest anterior segment examination including intraocular pressures was normal. On fundus examination, there was incomplete posterior vitreous detachment and a haemorrhage covering temporal half of right optic disc (Figure 1), which was subhyaloid in location. Visual fields of right eye revealed a superior arcuate and early inferior arcuate field loss. The visual field of the left eye was within normal limits. Fluorescein angiography was normal except for blocked fluorescence at the site of optic disc haemorrhage (Figure 2). CT scan showed no evidence of intracanalicular fracture or optic nerve compression. A diagnosis of right traumatic optic