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Sir,

Reply to MA Elgohary, DYL Leung and DSC Lam

We would like to thank Dr Elgohary for the interest in and comments on our paper.[1] The evidence for the relative importance of the immunomodulatory effects of oestrogen and progesterone remains inconclusive. In specifying the exact date of onset of acute uveitis, we were sensitive to the problem that there may be a short delay between immune 'trigger' and onset of symptoms. In view of this, it would be difficult to draw firm conclusions about the hormonal influence on particular days within the late phase of the menstrual cycle, at which time hormonal levels change precipitately. We can confirm that no patient was using concurrent oral steroid or immunosuppression at presentation.

We are also grateful for the comments on our paper[1] by Leung and Lam. While we understand and appreciate the points made, we disagree with most. Almost all regularly menstruating women have excellent recall of the date of last menstrual period (LMP). Where there was doubt, the patient was not included in our study; there was therefore no recall bias on this parameter. We commented that it may be interesting to measure oestrogen and progesterone levels in such patients; however, the absence of these data does not affect the validity of our comments. Our proposition is not that uveitis commences at a particular hormone level, but that it may be precipitated by hormone withdrawal in the predisposed. It is not necessary to measure hormone levels to prove that a woman is in the late phase of the menstrual cycle, if the LMP date is known.

We strongly disagree that it is 'not unusual' for anterior uveitis to be relatively silent in the early phase of disease; all our patients had acute-onset anterior uveitis with clear memory of the date of onset of symptoms, and rapid attendance at our ophthalmic emergency department, usually within 24 h. No patient had acute-on-chronic disease and no patient had recently discontinued topical steroid. Where there was doubt, the patient was excluded. We disagree that the aetiology of uveitis could affect presentation; while uveitis such as that related to juvenile idiopathic arthritis (JIA) can indeed be asymptomatic, that is not relevant to acute-onset symptomatic uveitis presenting in adulthood. We can confirm that no patient had JIA-related uveitis. We disagree that measurement of objective signs of severity might be a more reliable indicator of onset than reported symptoms; there is a wide variation in the severity of inflammation in those presenting with acute anterior uveitis; there is no evidence that this has a bearing on the duration of inflammation before presentation. On the contrary, it is not unknown for patients with recurrent anterior uveitis to be aware of recurrence (and to present with symptoms) before cells are detected in the anterior chamber. Finally, although the incidence of uveitis in the premenstrual phase did not quite reach statistical significance, the incidence in the whole postovulatory phase did; having carefully stated potential sources of error, we believe that our conclusions remain valid.

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Sir,

Eyelash poliosis in association with sarcoidosis

Sarcoidosis is a multisystem granulomatous disease. It frequently affects young adults, with a female

preponderance and commonly presents with respiratory, dermatological, or eye symptoms. One of the commonest presentations to the Eye department in patients with sarcoidosis is of acute anterior uveitis. We present a case of lash poliosis and anterior uveitis in a patient with sarcoidosis. We believe it to be the first reported case of poliosis associated with ocular sarcoidosis.

Case report

A 50-year-old Asian woman presented to Rheumatology department with an itching left calf and swelling in the right calf. An incision biopsy demonstrated granulomatous inflammation on histology. A chest radiograph showed appearances consistent with sarcoidosis. Her serum ACE was elevated at 119 IU/l and a diagnosis of sarcoidosis was made.

She was commenced on oral prednisolone 30 mg o.d. and referred to Ophthalmology department to screen for eye involvement.

At the Ophthalmic outpatients department she complained of itchy eyes. There were no other associated symptoms. Examination revealed no lacrimal gland swelling or lid margin granulomata. She had eyelash poliosis bilaterally (Figure 1). The tear breakup times were reduced at 2 s bilaterally. The right eye demonstrated keratic precipitates and cells in the anterior chamber. Her intraocular pressures were normal. There was mild inflammatory activity in the right anterior vitreous; however, both retinae were normal.

She was diagnosed with dry eyes and right anterior uveitis, and commenced on topical steroids and artificial lubricants.

Discussion

Poliosis is the depigmentation of hair and has been described previously in association with several inflammatory conditions including idiopathic uveitis, Vogt–Koyanagi–Harada (V–K–H) syndrome, vitiligo, Marfan's syndrome and tuberous sclerosis.[1–3]

Figure 1 Eyelash poliosis.

The ocular manifestations of sarcoidosis are well documented and include lid granulomata, lacrimal gland involvement, conjunctival follicles, episcleritis and scleritis, anterior uveitis, cataract, vitritis, choroiditis, periphlebitis, and retinal granulomata among others.

Sarcoidosis is also known to be associated with vitiligo, as are other granulomatous inflammatory diseases such as V–K–H syndrome.[1–3] There is considerable overlap between various conditions involving ocular inflammation and depigmentation of both skin and hair.

It has been postulated that depigmentation is the result of an autoimmune mechanism. In sarcoidosis and V–K–H syndrome, there is overactivity of CD3 and increased expression of cytokines IL-1β, IL-2, IL-6, and TNF-α. This leads to T-helper cell differentiation and subsequent activation of T-cytotoxic cells and B cells, the latter producing circulating antibodies that attack melanocytes, thereby causing failure of pigmentation.[4,5]

To the best of our knowledge there have been no prior reports of an association between sarcoidosis and poliosis in the literature. We suggest that in cases of uveitis seen with poliosis, immediate investigations should specifically include those directed to elicit the presence of sarcoidosis, which may require systemic therapy.

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Sir,

Anti-TNF alpha therapy in chronic necrotizing scleritis resistant to standard immunomodulatory therapy in a patient with Wegener's granulomatosis

Wegener's granulomatosis is characterized by a necrotizing granulomatous vasculitis. Ocular complications may include uveitis, scleritis, and retinal vasculitis and occur in up to 45% of patients.[1,2]

Case report

A 75-year-old man was referred to our clinic for peripheral ulcerative keratitis and anterior uveitis in both eyes. High serum proteinase-3 antibody levels in addition to a biopsy of the nasal mucosa revealing a necrotizing granulomatous vasculitis confirmed the diagnosis of Wegener's granulomatosis (WG). His visual acuity (VA) was 20/50 in the right eye (OD) and 20/100 in the left eye (OS). Systemic immunomodulatory therapy with oral cyclophosphamide (3 mg/kg/BW) with 75 mg glucocorticoids was initiated; however, despite adjustment of cyclophosphamide (4 mg/kg/BW), the scleritis remained active over the following 5 months (Figure 1a). His VA dropped to 20/200 OD and counting fingers OS. Owing to ongoing inflammation and positive reports on infliximab in systemic Wegener's granulomatosis,[3–5] infliximab was started at 5 mg/kg/BW on day 0, and repeated at weeks 2, 6, thereafter every 8 weeks. Cyclophosphamide was discontinued after week 6. After the second infliximab infusion, the anterior chamber (a/c) cell counts as well as the necrotizing scleritis decreased. At week 8, his scleritis was in remission. His VA increased to 20/60 OD and remained at counting fingers OS. Prednisolone was tapered to 12.5 mg/day (Figure 1b). After 5 months, infliximab was discontinued due to an acute herpes zoster infection, resulting in a flare-up 6 weeks later with 4+ a/c cells OD (Figure 1c) and an almost total melting of the sclera OS. His VA was counting fingers OD and light perception OS.

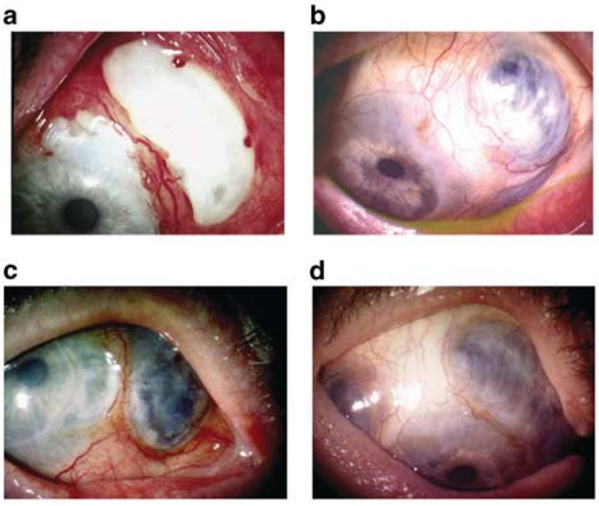

Figure 1 (a) Photograph demonstrates the active necrotizing scleritis with inflammation of the eye. (b) At 5 months after infliximab therapy has been started. The scleritis is almost completely in remission. (c) Reactivation of the scleritis after discontinuation of the scleritis. (d) At 3 months into reinstallation of combined infliximab and azathioprine therapy.

Therefore, infliximab (5 mg/kg/BW) was restarted and azathioprine at 1.5 mg/kg BW and prednisolone 50 mg/day were added. For the last 8 months, infliximab has been administered every 4–5 weeks keeping the scleritis under remission (Figure 1a). His VA increased to 20/400 in the right eye. The corticosteroid dose was tapered to 12.5 mg/day.

The classic regimen consists of cyclophosphamide and prednisone.[1,2] However, as not all patients sufficiently respond to standard therapy, and due to its toxicity alternative treatment should be considered. This is just a single case report. Still the fact that our case, in addition to previous reports, shows the effectiveness of specific TNFα-blockers in systemic WG[3–5] suggests that selective TNFα inhibition is an option in ocular manifestations of this disease.

Acknowledgements

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