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Sir,

Re 'Isolated eyelid edema in Melkersson–Rosenthal syndrome: a case series'

Rawlings *et al*[1] have reported on a series of five patients with isolated eyelid edema and have made the diagnosis of Melkersson–Rosenthal syndrome on the basis of granulomatous inflammation. Melkersson–Rosenthal syndrome is described as a granulomatous disease with the triad of facial palsy, facial edema, and a fissured tongue, although the complete triad is reported to be seen in only 25% of cases.[2]

We have recently reported on a series of 15 patients with chronic eyelid edema, and in 9 of these cases (60%) there was an associated diagnosis of acne rosacea.[3] Granulomatous inflammation was present in some of these patients, and this has been reported before in the presence of acne rosacea.[4] Indeed, acne rosacea and Melkersson–Rosenthal syndrome have some overlap in their clinical and pathological features and both are classified as granulomatous dermopathies. The illustrations of Cases 1 and 2 in the series of Rawlings *et al*[1] show facial features that would be consistent with acne rosacea, with rhinophymatous change and thickened glabellar skin. I suspect these two illustrated patients do indeed have acne rosacea rather than Melkersson–Rosenthal syndrome, and it would be of interest to know whether any of the other three patients in the series also showed features of rosacea.

It is likely that Melkersson–Rosenthal syndrome is over diagnosed when the other features of the syndrome are absent, and many of the reported cases of eyelid edema as the only feature of the syndrome are more likely to have acne rosacea as the underlying cause of their eyelid edema. Such phymatous change in the eyelid was certainly the commonest cause in our series, which to date is the largest published series of chronic eyelid lymphedema.

Conflict of interest

The author declares no conflict of interest.

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Sir,

Response to Dr McNab

We thank Dr McNab[1] for his comments on our report of isolated eyelid edema in Melkersson–Rosenthal Syndrome (MRS).[2] He suggests that our cases are better considered examples of acne rosacea, and cites his own study[3] and the case report of Lai *et al*[4] in support of this.

In Dr McNab's case series, histopathological examination of eyelid skin from five patients with a clinical diagnosis of acne rosacea and chronic eyelid edema showed some degree of granulomatous inflammation in three specimens.[3] The granulomas were not illustrated but were described as 'poorly formed' (case 4), 'single' (case 11) and 'surrounding dilated lymphatics' (case 15). Lai *et al*[4] also referred to the presence of 'ill-defined perivascular granulomas' but did not illustrate them. Other studies of chronic eyelid edema in rosacea did not mention dermal granulomas.[5,6] In none of our cases was rosacea felt to be the primary underlying cause, either clinically or histopathologically.

In our practice, we do not regard poorly defined granulomas as indicative of any specific diagnosis. Granulomatous rosacea is typically characterised by a tuberculoid (necrotising) or sarcoid-like response, possibly to the contents of hair follicles. In our series, the granulomas were neither tuberculoid nor sarcoid-like but were sharply defined, perivascular, and perilymphatic, often with an intralymphatic component. In addition, and illustrated in our paper, discrete granulomas were identified in orbicularis muscle and anterior orbital soft tissue, which does not appear to have been described in rosacea.

Dr McNab may be correct in saying that MRS is over diagnosed in cases of isolated eyelid edema.[1] Nevertheless, the clinico-pathological pattern that we and other authors have ascribed to monosymptomatic MRS appears quite distinct. Until our understanding of the etiology and pathogenesis of oro-facial granulomatosis, of which MRS is one part, increases, we see no justification for regarding these cases as a form of granulomatous rosacea.

Conflict of interest

The authors declare no conflict of interest.

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Sir,

Lipomatosis of the Orbits: possibly a form of Madelung's disease

Madelung's disease is a systemic disorder, which is typically characterized by symmetrical deposits of adipose tissue in the neck and upper back. In 1888, Madelung collected a series of cases outlining the macroscopic structure and clinical features of this disease. It is more common in males and associated with high alcohol consumption.[1] In this report, we present the clinical and radiological manifestations of Madelung's disease in the orbits, which to our knowledge has not been described before.

Case report

A 49-year-old Russian male was referred to the orbital clinic with prominent eyes. He had no previous ocular history. Three months before this he was diagnosed with Madelung's disease when he presented with neck swelling secondary to fatty infiltration. He underwent bilateral neck dissections with histology revealing well-differentiated adipose tissue. He denied excessive alcohol consumption although his gamma-glutamyl transpeptidase was raised at 202 (normal range 2–50).

On examination, his visual acuity was 6/6 with normal colour vision on Ishihara test plates for both eyes. Hertel exophthalmometry revealed proptosis of 23 mm bilaterally and marked periorbital soft-tissue fullness (Figure 1). He had full range of eye movements and his remaining eye examination was normal. His thyroid function tests were normal. A computerized tomography scan of the orbits demonstrated symmetrical orbital fat deposition with straightening of the optic nerves (Figure 2). In view of the worsening nature of the disease and straightening of the optic nerves indicative of risk of compromise, he underwent bilateral medial orbital wall decompression and fat excision. Post operatively, he achieved a 2 mm reduction in proptosis bilaterally. Histology of excised fat demonstrated mature adipose tissue consistent with Madelung's disease.

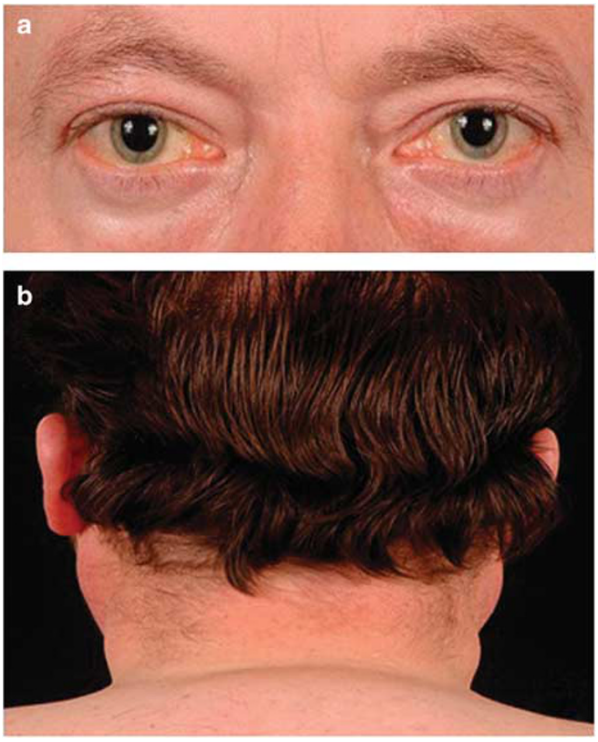

Figure 1 (a) Photograph of patient's eyes in the primary position, demonstrating bilateral proptosis and periorbital fullness secondary to adipose deposition. (b) Photograph of patient's neck with pathological fat deposition.

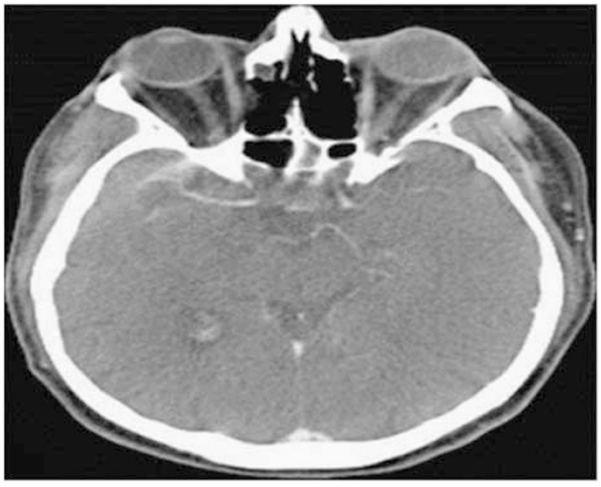

Figure 2 Axial computer tomography scan of the brain and orbits, demonstrating symmetrical straightening of the optic nerves secondary to orbital fat deposition and proptosis.