Panuveitis revealing Melkersson Rosenthal syndrome: a case report

Panuvéite révélant un syndrome de Melkersson Rosenthal : à propos d'un cas

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RÉSUMÉ

But : Décrire un cas de panuvéite bilatérale révélant un syndrome de Melkersson Rosenthal (SMR).
Observation : Patiente âgée de 35 ans, suivie pour panuvéite bilatérale chronique idiopathique, adressée pour exploration. A l'examen, l'acuité visuelle était chiffrée à 3/10 au niveau de l'œil droit et limitée à voir bouger la main du côté gauche. L'examen à la lampe à fente a mis en évidence un segment antérieur calme, des synéchies irido-cristalliniennes rompues et une hyalite plus dense à gauche. L'examen du fond d'œil a objectivé des engainements vasculaires périphériques et l'absence de foyers chorio-rétiniens. L'angiographie à la fluorescéine a mis en évidence une vasculite rétinienne occlusive bilatérale. Le bilan étiologique a éliminé une pathologie infectieuse sous-jacente, notamment une tuberculose oculaire. Une revue des systèmes avec examen de l'interniste a révélé une chéilite avec plicature de la langue et la notion de deux épisodes de paralysie faciale avec œdème palpébral. Le diagnostic de SMR a été retenu, et la patiente a été mise sous corticoïdes par voie systémique associé à une photocoagulation sectorielle au laser des territoires d'ischémie rétinienne.
Conclusion : Le SMR est une entité rare. L'atteinte oculaire, notamment l'uvéite, est exceptionnelle. Ce syndrome doit être reconnu par les cliniciens pour éviter des erreurs diagnostiques et des explorations inutiles.
Mots clés : syndrome de Melkersson Rosenthal, uvéite, paralysie faciale, œdème palpébral

SUMMARY

Aim: To describe a case of panuveitis revealing Melkersson Rosenthal syndrome (MRS).
Case report: A 35-year-old female with a history of bilateral chronic idiopathic panuveitis was referred for work up. On examination, her visual acuity was 3/10 in the right eye and limited to hand motion in the left eye. Slit lamp examination revealed posterior synechiae and vitritis more severe in the left eye. Fundus examination showed bilateral vascular sheathing with no retinitis or choroiditis. Fluorescein angiography revealed bilateral occlusive retinal vasculitis. A careful questioning of the patient revealed the history of relapsing facial paralysis with palpebral edema. The work up ruled out an underlying infectious disease, especially ocular tuberculosis. Examination of the internist revealed a cheilitis and a fissured tongue. The diagnosis of MRS was made and the patient was treated with systemic corticosteroids and scatter laser treatment of retinal nonperfusion areas.
Conclusion: MRS is a rare disorder. Ocular involvement, especially uveitis, is uncommon. Clinicians should be aware of this syndrome to avoid misdiagnosis and extensive work up.
Key words: Melkersson Rosenthal syndrome, uveitis, facial paralysis, eyelid edema

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INTRODUCTION

Melkersson–Rosenthal syndrome (MRS) is a granulomatous disease, of unknown cause, characterized by recurrent orofacial edema, facial palsy, and a fissured tongue (1**)**. Ocular involvement in MRS is rare, and it most often affects the periocular area (1,2)**.** Herein we describe a case of panuveitis revealing MRS.

CASE REPORT

A 35-year-old female patient, with a history of chronic panuveitis was referred for further work up. She was initially diagnosed with bilateral idiopathic panuveitis 8 years ago and treated with systemic corticosteroids.

On examination, her visual acuity (VA) was 3/10 in the right eye and limited to hand motion in the left eye. Slit lamp examination revealed a clear cornea, a quiet anterior chamber, ruptured posterior synechias, a moderately subcapsular cataract in both eyes, and vitritis more severe in the left eye. Fundus examination showed bilateral vascular sheathing with no retinitis or chroiditis (Figure 1). Fluorescein angiography revealed occlusive retinal vasculitis with peripheral nonperfusion areas (Figure 2). Optical coherence tomography showed no macular edema in both eyes. The diagnosis of bilateral panuveitis with occlusive retinal vasculitis was made.

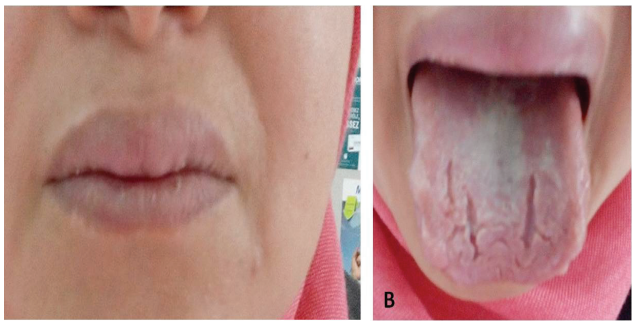

Figure 1. (A) Fundus photography of the right eye showing vascular sheating with retinal hemorrhages nasally to the optic disc (black arrow). **(B)** Fundus photography of the left eye showing dense vitritis.

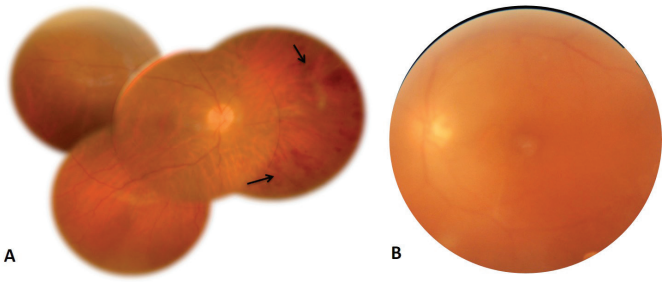

Figure 2. **(A, B)** Fluorescein angiography showing bilateral occlusive retinal vasculitis with peripheral non perfusion areas (yellow stars).

A careful questioning of the patient revealed the history of relapsing facial paralysis with palpebral edema. The work up including complete blood count, erythrocyte sedimentation rate, syphilis serology, conversion enzyme assay, and a chest radiograph revealed no abnormalities. Tuberculin skin testing and Quantiferon were negative and thus ruled out ocular tuberculosis. Examination of the internist revealed a cheilitis and a fissured tongue (Figure3). The diagnosis of panuveitis associated with MRS was made and the patient was treated with systemic corticosteroids initiated at a dose of 1mg/Kg/day then gradually tapered. Peripheral scatter photocoagulation of retinal nonperfusion areas was also performed. After a follow-up of 12 month, her final VA was 7/10 in both eyes with no recurrences.

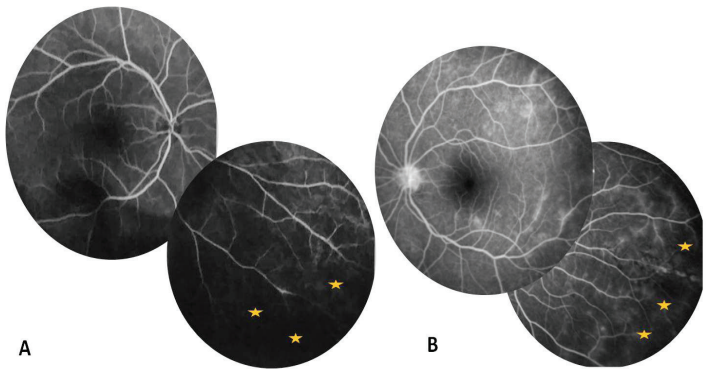

Figure 3. Photographs of the patient face showing a cheilitis **(A)** and a fissured tongue **(B)**.

DISCUSSION

MRS is a rare disorder with an unknown etiology (1). It often occurs in the first two decades of life. It is characterized by persistent or recurrent orofacial edema, relapsing facial paralysis and fissured tongue (1,2). The complete triad is very rare and the presence of at least two of the clinical criteria is generally considered sufficient to make the diagnosis (1,2).

An array of ocular manifestations in MRS are described, including blepharospasm, epiphora, granulomatous blepharitis, anisocoria, Claude Bernard Horner's syndrome, lagophthalmia (by facial palsy), dry keratitis, peripheral corneal opacities, recurrent conjunctivitis, and exophthalmos (3,4). Optic neuritis, papilloedema, and trigeminal nerve palsy are less common (5).

Uveitis is a less common finding in MRS. To our knowledge; only two cases of isolated anterior uveitis related to MRS have been reported in the literature (6,7). Ocular involvement was unilateral in the first case and bilateral in the second one. Babu et al reported a case of MRS in a patient diagnosed and treated for tubercular panuveitis (8). The diagnosis of ocular tuberculosis was made on the basis of a positive Mantoux test, a calcified nodule on chest X-ray and demonstration of Mycobacterium tuberculosis genome by polymerase chain reaction on the vitreous sample. The authors concluded that the association of MRS and tubercular uveitis may be casual (8).

Our case is the first report of panuveitis with retinal vasculitis associated with MRS. In our patient, the diagnosis of MRS was made on the basis of the classic triad of symptoms. The laboratory work up ruled out an infectious or systemic cause of uveitis. The clinical findings were not suggestive of a specific ocular entity. The diagnosis of panuveitis related to MRS was made and the patient was successfully treated with systemic corticosteroids.

CONCLUSION

MRS is a rare disorder, often overlooked. Ocular involvement, especially uveitis, is uncommon. Clinicians should be aware of this syndrome to avoid misdiagnosis and extensive and inefficient work up.

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