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SHORT COMMUNICATION

Case report

Bilateral Parry-Romberg syndrome associated with retinal vasculitis

C. BELLUSCI[1], R. LIGUORI[2], A. PAZZAGLIA[1], L. BADIALI[3], C. SCHIAVI[1], E.C. CAMPOS[1]

[1]Ophthalmology Unit
[2]Department of Neurological Sciences
[3]Pathology Unit, University of Bologna, School of Medicine, Bologna - Italy

PURPOSE. *To describe an unusual case of bilateral progressive facial hemiatrophy (Parry-Romberg syndrome (PRS)) associated with retinal vasculitis.*
METHODS. *In a 37-year-old man with bilateral PRS, retinal vasculitis of the right eye was evident on fundus examination and fluorescein angiography. Right temporalis muscle biopsy and needle electromyography of the masseter muscles were performed. The patient underwent immunosuppressive therapy and retinal laser photocoagulation.*
RESULTS. *Biopsy specimens showed large fibrosis with focal lymphohistiocytic infiltration of the muscle fibers. Electromyographic findings are consistent with a primary muscle disease. Visual acuity improved from 20/25 to 20/20 in the right eye with a follow-up of one year.*
CONCLUSIONS. *The evidence of retinal vasculitis and the histologic findings of facial changes observed in this PRS case could support the pathogenetic model of a chronic inflammatory process as a plausible explanation for progressive facial hemiatrophy. (Eur J Ophthalmol 2003; 13: 803-6)*

KEY WORDS. *Electromyography, Parry-Romberg syndrome, Retinal vasculitis, Suppressive therapy*

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INTRODUCTION

Progressive facial hemiatrophy (also known as Parry-Romberg syndrome (PRS)) is a sporadic entity of unknown origin characterized by slowly progressive atrophy of one side of the face, primarily involving subcutaneous and fat tissues (1).

Extension of the atrophic process to the contralateral side of the face is rare but may occur (2). Ophthalmic involvement is common and the most frequent finding is progressive enophthalmos secondary to atrophy of orbital fat (3).

We report an unusual case of bilateral PRS associated with retinal vasculitis.

Case report

A 37-year-old white man presented with a history of progressive atrophy of the left side of his face starting before 18 years of age. His best-corrected visual acuity was 20/25 in the right eye (RE) and 20/20 in the left eye (LE). The face examination revealed extensive atrophy of subcutaneous tissue, fat, and muscles, more severe on the left side (Fig. 1). Extensive areas of alopecia were present in the midline of the scalp.

He had bilateral, asymmetric (LE > RE) enophthalmos and ptosis. A large-angle left hypo-exotropia was present. Ocular motility of the LE was almost com-

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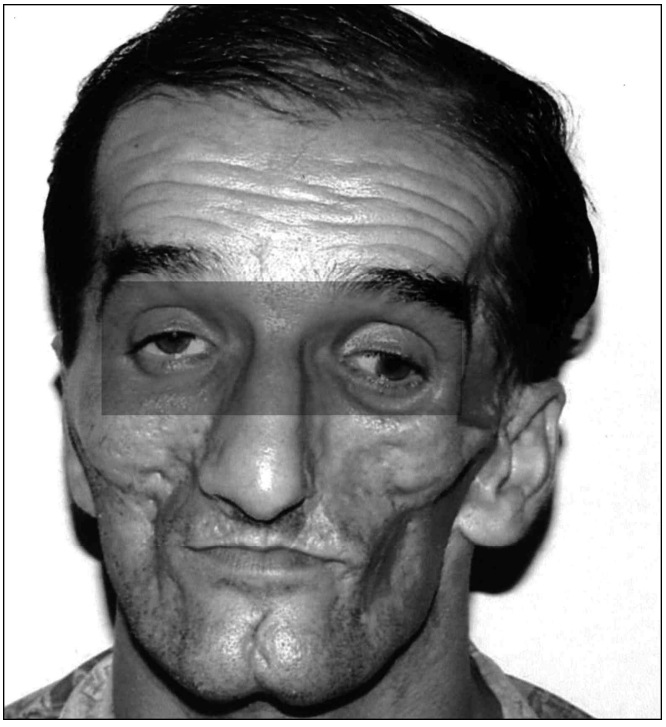

Fig. 1 - *A 37-year-old patient with a history of bilateral progressive facial hemiatrophy. Note the severe atrophy of the subcutaneous tissues of the left side of the face with restrictive strabismus.*

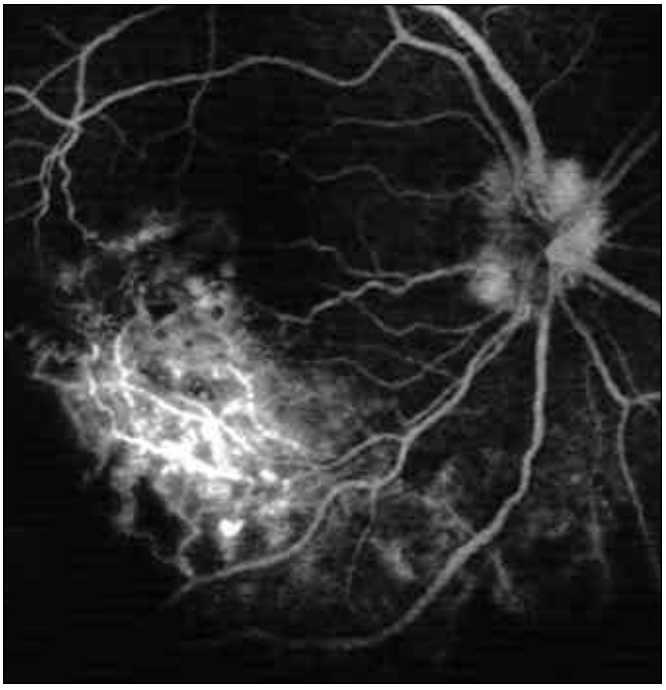

Fig. 2 - *Fluorescein angiography of the right eye reveals macular edema due to sectoral angiopathy of vessels in the perimacular region. An apparent nodular pattern of the optic disc due to leakage of papillary capillaries is evident.*

pletely absent; the elevation was severely limited in both eyes. Forced duction test performed in all directions was unequivocally positive for mechanical restriction of ocular motility in both eyes.

Slit-lamp examination results were normal. Fundus examination of the RE showed marked signs of retinal vasculitis with macular edema. Fluorescein angiography of the RE (Fig. 2) revealed a sectoral angiopathy of vessels in the perimacular region and an apparent nodular pattern of the optic disc due to leakage of papillary capillaries. Extensive areas of capillary dropout and distal small retinal hemorrhages were present in the inferior midperiphery. The left fundus examination showed several zones of choroidal atrophy extending to the ora serrata but sparing the posterior pole.

Routine laboratory tests were normal, as were antinuclear antibodies and rheumatoid factor.

Right temporalis muscle biopsy revealed diffuse fiber loss with large fibrosis and focal lymphohistiocytosis (Fig. 3). Quantitative analysis of the motor unit potentials with needle electromyography of the masseter muscles showed decreased mean potentials duration and increased incidence of polyphasic potentials.

Orbital magnetic resonance established the asymmetric (LE > RE) presence of fibrous tissue with replacement of the retrobulbar fat and thinning of the extraocular muscles. The brain magnetic resonance scans were normal.

These findings were consistent with the diagnosis of bilateral PRS associated with retinal vasculitis.

We performed laser photocoagulation of ischemic retinal areas. Moreover, the patient underwent immunosuppressive therapy with A-cyclosporine (4 mg/kg daily). Visual acuity improved to 20/20 in the RE with a follow-up of 1 year. Retinal angiography confirmed the improvement of vascular leaking with macular edema decrease (Fig. 4).

DISCUSSION

PRS is a sporadic but not extremely rare syndrome characterized by progressive shrinking and deformation of one side of the face (1). The onset of PRS usually occurs during the first two decades and the atrophic process involves skin, subcutaneous fat, connective tissue, and muscles of one side of the face (4).

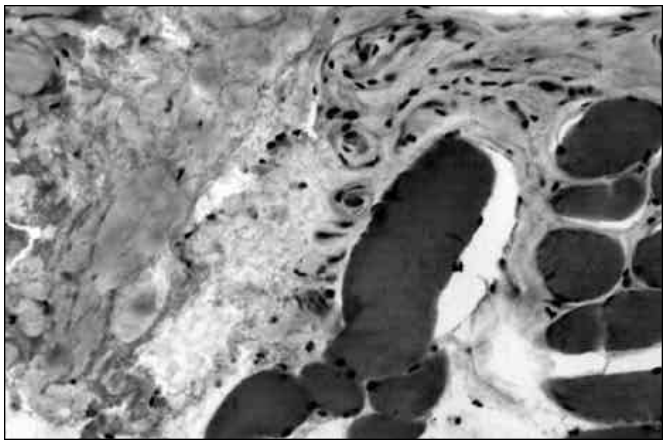

Fig. 3 - *Temporalis muscle biopsy shows large fibrosis with mild lymphohistiocytic infiltration. Several lymphocytes were recognizable between the atrophic-dystrophic muscle fibers (hematoxylin-eosin, original magnification x 40).*

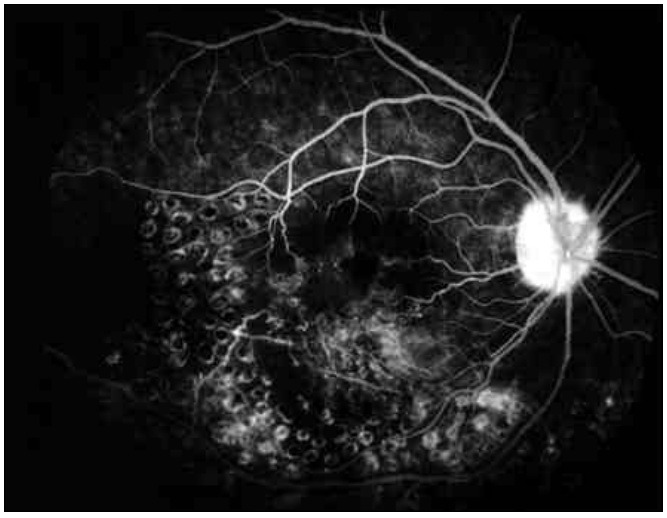

Fig. 4 - *Improvement of vascular leaking from perimacular vessels as well as from papillary capillaries is evident at the end of follow-up. Note the laser photocoagulation of ischemic retinal areas.*

Extension of the atrophy to the opposite side of the face was found to occur in 47 of the 400 cases reviewed by Archambault and Fromm (2). Ocular involvement in PRS is well recognized and occurs in up to 40% of cases; enophthalmos has been reported as the commonest ocular manifestation (5).

Our patient with bilateral PRS showed the unusual association with retinal vasculitis. Biopsy specimens of the right temporalis muscle revealed extensive fibrosis with focal lymphohistiocytic infiltration. Similar pathologic findings were found in the biopsy specimen from the lateral rectus muscle, in the retrobulbar fat, and in the lacrimal gland of a patient with intracranial vascular malformations (6). Moreover, the decreased mean potentials duration together with the increased incidence of polyphasic potentials found on electromyography of the masseter muscle are consistent with a primary muscle disease.

All these elements are in favor of an inflammatory process at the base of PRS. A chronic inflammatory process may be responsible for the subsequent atrophic changes, including enophthalmos secondary to shrinkage of the eyeball and thinning of the extraocular muscles (7).

The cause of progressive facial hemiatrophy is unknown. Two different pathogenetic models have been presented to explain PRS (8). The first model suggests that a dysfunction of trophic fibers of peripheral nerves may lead to the facial atrophy. The second model is based on an inflammatory process primarily localized in facial tissues and sometimes, by local invasion, in parts of the brain.

The evidence of retinal vasculitis and the histologic findings of facial changes observed in this PRS case could suggest the presence of a chronic inflammatory process leading to atrophy of various tissues of the face. Cyclosporine treatment may represent the beginning of a new trend in treatment of this syndrome (9). In this very slowly progressive disease, therapeutic efficacy is difficult to evaluate; therefore, a case with active retinal vasculitis where changes are clearly visible could potentially be ideal.

Reprint requests to:
Prof. Emilio C. Campos
Clinica Oculistica dell'Università
via Massarenti 9
40138 Bologna, Italy
campos@alma.unibo.it

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