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Brief communication

Parry–Romberg syndrome associated with borreliosis: could photochemotherapy halt the progression of the disease?

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Parry–Romberg syndrome (PRS) or progressive hemifacial atrophy is a rare entity characterized by unilateral atrophy of the skin, subcutaneous tissue and sometimes bone and cartilage. Although this syndrome has overlapping features of scleroderma 'en coup de sabre', it shows little or no sclerosis and may affect the entire distribution of the trigeminal nerve including the eye and tongue. As the pathogenesis is unknown, no effective therapy exists. We present here the third case of PRS associated with borreliosis and more interestingly two cases whose progressive course have been stabilized with gel PUVA therapy.

Key words: **borreliosis; Parry–Romberg syndrome; progressive hemifacial atrophy; PUVA.**

Parry–Romberg syndrome (PRS) is an acquired progressive hemifacial atrophy of the subcutaneous and bone structure of unknown pathogenesis (1). The tongue, palate and uvula are often involved.

It is not always possible to differentiate PRS from scleroderma 'en coup de sabre' (SCS). Although some clinical and histopathological features are proposed for differential diagnosis, whether this syndrome is a distinct entity or belong to the spectrum of scleroderma is a controversy (2).

As observed in some cases of localized scleroderma (LS), *Borrelia burgdorferi* has been related to PRS (3). We present here two cases of PRS one of which had positive serological tests for borreliosis and the progression of both was found to be halted with gel photochemotherapy.

Case 1

An 8-year-old girl presented with a hyperpigmented, atrophic and depressed patch of 1.5 cm in size on the right side of her chin and a linear and slight depression on the tip of her nasal ala with palatal hemiatrophy and significant ipsilateral atrophy of tongue and uvula which caused a facial asymmetry (Fig. 1). Soft tissue involvement of the face presented the areas of trigeminal nerve distribution. She had no history of any blunt trauma or a tick bite on the face. She was previously diagnosed as Borreliosis and her antibodies to *B. burgdorferi* were positive (immunoglobulin (Ig)M 1 : 16, IgG $>1:128$). She had recently completed her antibiotic therapy (60 mg/kg/day amoxicillin for 3 weeks) when she was consulted at our clinic.

Complete blood count, urine analysis, biochemical tests, VDRL testing, antinuclear and antihistone antibodies, rheumatoid factor and Ig levels were found to be normal or negative. Further evaluation of neurological and ocular systems did not reveal any abnormalities.

Punch biopsy of the chin demonstrated normal epidermis and minimal increase of mononuclear inflammatory cells in the dermis but subcutaneous adipose tissue was absent. Because PRS and LS had overlapping features, topical therapy with calcipotriol was commenced. Because no efficacy was obtained within 3 months of trial and new hyperpigmented macules developed, the therapy was switched to gel photochemotherapy. The patient was classified as skin type III (Fitzpatrick's classification) and was treated with gel PUVA (0.1% 8-methoxypsoralen containing gel). A thin layer of gel was applied only on hyperpigmented macules 1 h before UVA irradiation with an initial UVA dose of 2 J/cm^2 twice a week performed with a classic PUVA device (Spectra 311/350; Daavlin,

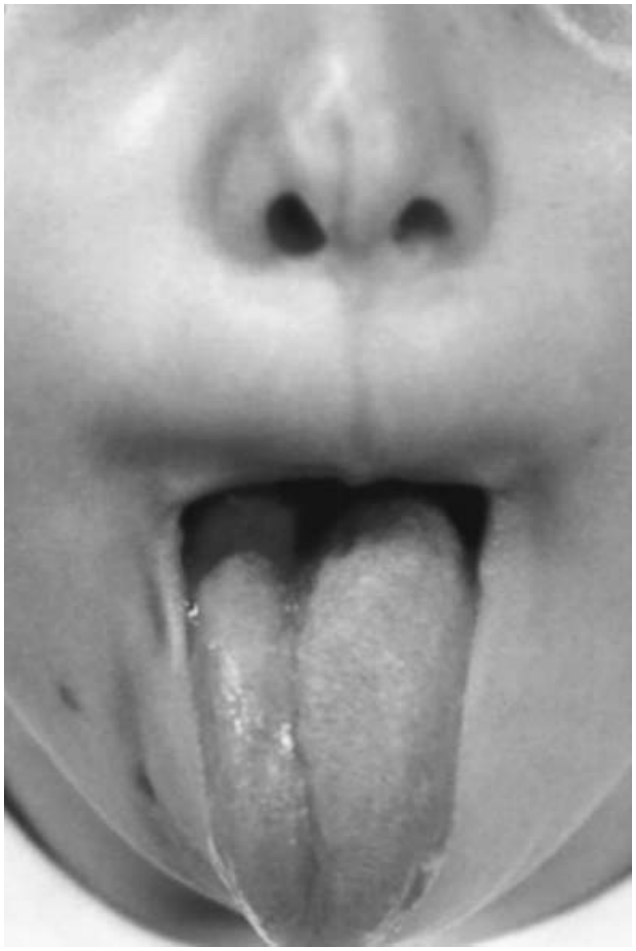

Fig. 1. Significant ipsilateral atrophy of the tongue.

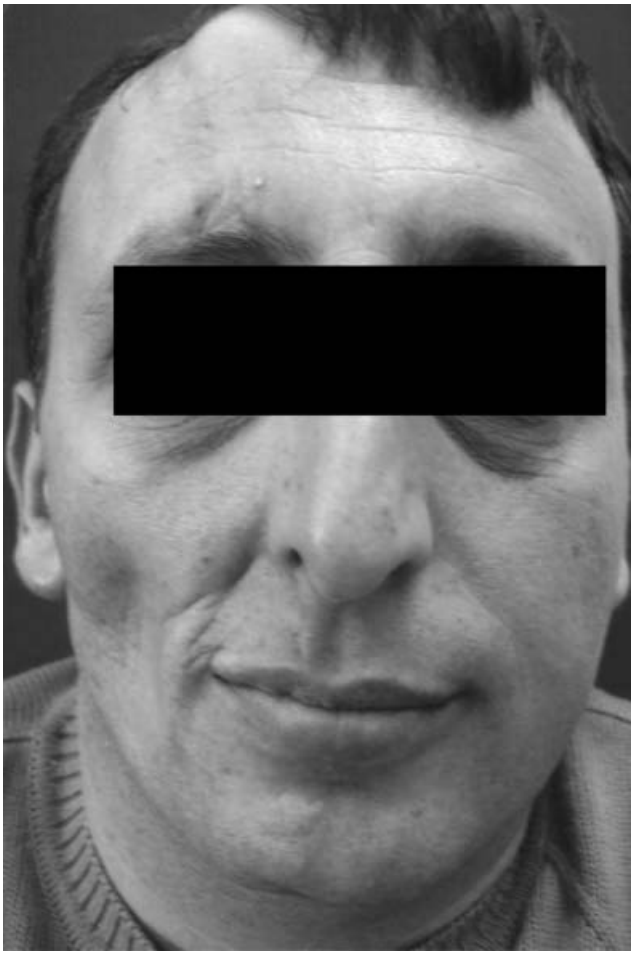

Fig. 2. Linear depressed, atrophic patch of 10 cm in size on right parietal scalp accompanied by the cicatricial alopecia of the affected area and on forehead. Significant ipsilateral atrophy of cheek.

Bryan, OH, USA). UVA dose was increased as 0.5 J/cm^2 after three sessions. At the end of 25th session, development of new lesions as well as deepening of previous lesions was halted and the color of two recent macules fainted away. The frequency of therapies was decreased to once a week. The patient currently receives the 39th session and treatment of gel PUVA therapy has resulted in a cumulative UVA dose of 137 J/cm^2 so far.

Case 2

A 35-year-old male patient presented with a firm, depressed, progressing linear atrophic patch of 10 cm in size with alopecia on right parietal area of his scalp, extending to forehead down to ipsilateral eyebrow and with a significant ipsilateral atrophy of his cheek of 12-year-duration (Fig. 2). His medical history was unremarkable with no history of any blunt trauma or a tick bite. Physical, neurological, ophthalmologic examinations and the laboratory investigations were normal similar to the first case.

Biopsy of the scalp demonstrated thinning and fibrosis of the underlying dermis and total loss of pilosebaseous units. Biopsy of the cheek showed chronic perifollicular and perivascular mononuclear infiltration of the dermis without atrophy in epidermis. Computerized tomography of the mandibula demonstrated a significant thinning of the ipsilateral subcutaneous tissue with normal thickness of underlying muscle tissues and no abnormalities of bone tissues.

The patient did not have a history of any topical or systemic medications. Based on our observations on the first case, gel photochemotherapy was commenced as described above. The patient was classified as skin type I (Fitzpatrick's classification). Initial UVA dose was 0.5 J/m^2 and it was increased as 0.5 J/m^2 per three sessions. Significant softness of the cutaneous induration on the scalp was observed at the end of 20th session. He currently receives the 35th session and the

current cumulative dose has been 63 J/m^2 so far. The patient was consulted with Plastic and Reconstructive Surgery for autologous fat transplantation to his cheek.

Discussion

The psychological burden of PRS on the patient is indisputable and as the pathogenesis is unknown, no satisfactory therapy exists either to stabilize or retrieve its course.

This syndrome has many features of LS but the atrophy is usually deeper and the overlying skin is less bound down. The features helpful for differential diagnosis are the presence of cutaneous sclerosis, antihistone antibodies and histopathological findings of fibrosis, adnexal atrophy and mononuclear cell infiltrates in the latter (2).

The coexistence of PRS and LS on trunk or extremities suggests the overlap of both conditions or different variations of the same disorder. Evidence that strengthens the link between two disorders is the presence of case reports of Borreliosis associated with PRS (3) similar to those of LS. Our first case is the third example of such an association in literature.

Therapy of PRS is still limited. Immunosuppressive drugs such as cyclophosphamide, methotrexate and prednisolone have been previously used in patients with cerebral involvement without significant benefit on clinical lesions (4).

In several studies, cream PUVA or systemic PUVA and UVA1 therapy were found to be successful in LS (5). Compared with other therapeutic options; topical PUVA is of advantage as the applied cumulative doses are usually very low. The immunologic, anti-inflammatory activities with the induction of collagenase may explain the therapeutic effects of this therapy in LS. Whether this therapy immunologically blocks or interrupts a common pathway or step in both LS and PRS remains to be elucidated but it appears that this therapy is especially efficient on sclerodermic lesions of PRS, its variant overlapping with LS, but does not improve the atrophy affecting the cheek area. If the hypothesis of the conversion of LS into PRS is accepted (2), the cessation of the disease process at sclerodermic stage could prevent the development of a deep atrophy.

The spontaneous 'burning out' of PRS could not be in fact excluded in these patients. However, based on our observation of cessation in disease progression in the first case and softening of old sclerodermic plaque with regression of cutaneous induration in the second, we conclude that the possibility of topical PUVA as a worthwhile therapeutic option in PRS should be considered.

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