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COSMETIC COMMENTARY

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A case of bilateral Parry-Romberg syndrome successfully treated with hyaluronic acid filler augmentation

Jacqueline Watchmaker MD | Dana Saadeh MD | Christina Lam MD | Neelam A. Vashi MD

Department of Dermatology, Boston University School of Medicine, Boston, Massachusetts

Correspondence
Neelam A. Vashi, Department of Dermatology, Boston, MA.
Email: nvashi@bu.edu

Abstract

Parry-Romberg syndrome is a rare acquired neurocutaneous disorder typically characterized by hemifacial atrophy. Few cases of bilateral facial involvement have been reported. We report a case of a 60-year-old female with a 20-year history of progressive bilateral facial atrophy. Although an initial diagnosis of acquired partial lipodystrophy was considered, a diagnosis of bilateral Parry-Romberg was favored for three main reasons: (a) lipoatrophy was confined to the face (b) imaging findings of enophthalmos and underlying white matter changes (c) laboratories showing absence of C3 nephritic factor and normal complement levels. The patient was treated with hyaluronic acid fillers with dramatic improvement in cosmetic appearance.

KEYWORDS
hyaluronic acid filler, lipoatrophy, Parry-Romberg

1 | INTRODUCTION

Parry-Romberg syndrome (PRS) is an uncommon variant of morphea typically characterized by a slowly progressive hemifacial atrophy that can involve the skin, subcutaneous tissues, muscles, and underlying bony structures. Clinical and radiographic evidence of underlying central nervous systemic involvement is also frequent in PRS.[1,2] The majority of cases reported describe unilateral involvement with only few reported cases of bilateral involvement.[3,4] We report a case of a 60-year-old female with a 20-year history of progressive bilateral facial atrophy diagnosed with bilateral PRS.

2 | CASE REPORT

A 60-year-old Haitian woman with past medical history significant for latent tuberculosis, hepatitis B, type II diabetes mellitus, and essential hypertension initially presented to an outside hospital with a 20-year history of progressive facial atrophy. She denied any significant illness or trauma that preceded the atrophy, and her family history was negative for any type of similar condition. Two punch biopsies of the left cheek were performed which showed mild superficial perivascular and perifollicular lymphocytic infiltrate with rare eosinophils, telangiectasia, and eccrine coils with loss of peri-eccrine adipose tissue. Biopsy from the face in an area with clinical preservation of subcutaneous fat showed presence of subcutaneous fat with no significant pathologic changes.

Laboratory work-up including complete blood count (CBC), basic metabolic panel, lipid panel, hepatic function panel, urinalysis, C-reactive protein (CRP), erythrocyte sedimentation rate (ESR), rapid plasma regain (RPR), anti-neutrophil antibody (ANA), anti-Ro antibody, anti-La antibody, anti-double stranded DNA, SCL-70 antibody, anti-centromere antibody, rheumatoid factor antibody, anti-neutrophil cytoplasmic antibody (ANCA), proteinase-3 antibody, myeloperoxidase antibody, complement levels, and human immunodeficiency virus (HIV) were all unremarkable. Computed tomography (CT) scan showed hypodensities within the right periventricular white matter and asymmetric volume loss. Magnetic resonance imaging (MRI) showed moderate cerebral volume loss with asymmetric right periventricular white matter changes and marked thinning of the skin and loss

of subcutaneous fat in the face and frontal region with enophthalmos secondary to volume loss of the orbital fat. Based on these findings, the differential diagnosis included acquired partial lipodystrophy vs bilateral Parry-Romberg syndrome.

The patient then presented to our medical center in January of 2018. Outside records were reviewed. Repeat CBC and complete metabolic panel were unchanged from outside hospital laboratories, and C3 nephritic factor was found to be negative. A diagnosis of bilateral Parry-Romberg was favored for three main reasons: (a) lipoatrophy was confined to the face (b) imaging findings of enophthalmos and underlying white matter changes (c) laboratories showing absence of C3 nephritic factor and normal complement levels. Given her enophthalmos, ophthalmology evaluation was included as part of the patient's multidisciplinary care. She was found to have a normal eye examination; however, she was diagnosed with trigeminal neuralgia and started on carbamazepine.

Given severity of involvement and possible progression of disease, the patient was started on low-dose methotrexate (10 mg weekly) after initiating treatment for latent tuberculosis with concomitant cosmetic intervention. A trial of hyaluronic acid (HA) fillers was instituted in a staged approach. Significant improvement was seen with HA fillers and there was no apparent progression of disease process at 2-month follow-up (Figures 1, 2).

3 | COMMENT

Parry-Romberg syndrome is a rare disease of unknown etiology characterized by a slow and progressive atrophy typically of one side of the face. Only 2%-10% of cases are described as being bilateral.[3-5]

Parry-Romberg syndrome can be associated with epilepsy, migraines, ophthalmologic issues, trigeminal neuralgia, depression, and anxiety.[1-5] Accordingly, our patient was diagnosed with trigeminal neuralgia. Stress, surgery, and pregnancy have been reported to possibly accelerate the disease process.[4] Our patient denied any significant life stressors, illness, or surgery prior to the onset of her condition.

The main differential diagnosis of bilateral PRS is acquired partial lipodystrophy (APL), which was considered in our patient. APL typically presents with a gradual onset of symmetrical bilateral subcutaneous fat loss from the face and upper trunk with sparing of the lower extremities.[6] Most patients with APL have C3 nephritic factor and low C3. C3 nephritic factor is postulated to contribute to the degradation of adipose tissue.[7] Our patient had normal complement levels, lacked C3 nephritic factor, and her fat loss spared her upper trunk leading us to favor a diagnosis of bilateral PRS over APL. In addition, our patient's underlying white matter changes and enophthalmos are not typical of APL.

Treatment for Parry-Romberg syndrome can be challenging with methotrexate being the most commonly used medication for active disease. Methotrexate was started on our patient given concern that her disease was still slowly progressive and to prevent any possible cosmetic related acceleration of this rare and etiologic unknown disease. Treatment of the esthetic deficit of PRS is important to many patients and usage of autologous fat transfer, hyaluronic acid fillers, silicone injections, flaps, and prosthesis have all been reported with variable success.[4,8] Initially, autologous fat transfer was considered with plastic surgery; however, literature shows that only 19% of patients with Parry-Romberg syndrome treated with fat transfer have had a successful outcome.[4] It has been hypothesized that disturbances in fat metabolism may contribute to PRS and therefore the authors felt a hyaluronic acid-based filler would be less subject to degradation than autologous fat. In our patient, significant improvement was seen with HA fillers and there was no apparent progression of disease process at 2-month follow-up.

In conclusion, despite the rarity of the disease, dermatologists should have a high index of suspicion for bilateral Parry-Romberg syndrome when evaluating a patient with progressive bilateral facial atrophy, underlying white matter changes, and unremarkable laboratory work-up. The investigators would recommend initiating systemic treatment prior to cosmetic intervention and using hyaluronic acid fillers over autologous fat transfer.

ORCID

Jacqueline Watchmaker https://orcid.org/0000-0002-9830-6352

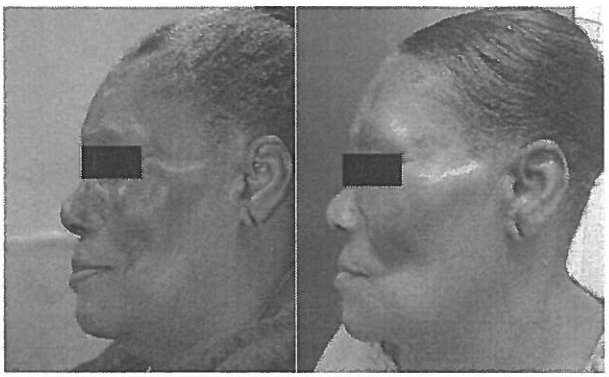

FIGURE 1 Left face before (left) and after (right) hyaluronic acid filler injections to malar cheeks and temples

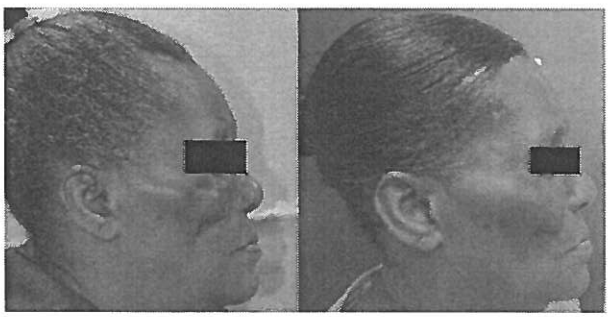

FIGURE 2 Right face before (left) and after (right) hyaluronic acid filler injections to malar cheeks and temples

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