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Parry–Romberg Syndrome in Two Young Afghan Patients: First Case Report

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ABSTRACT

Parry–Romberg syndrome (PRS), or progressive hemifacial atrophy, is a rare neurocutaneous disorder characterized by unilateral, slowly progressive atrophy of facial soft tissues and, in some cases, underlying bone. Reports from South-Central Asia are scarce, and no confirmed cases have previously been documented from Afghanistan. We report two young Afghan patients—an 18-year-old male and a 22-year-old female—who presented with insidious, progressive left-sided facial atrophy without prior trauma, infection, or autoimmune disease. Clinical examination revealed unilateral soft-tissue wasting, skin thinning, and facial asymmetry, with normal neurological findings. Computed tomography demonstrated atrophy of subcutaneous fat and facial musculature with subtle osseous thinning. Follow-up over 6 and 4 months showed no further progression. Quantitative imaging revealed an 18–22% reduction in soft-tissue thickness in one patient and 15–17% in the other. The left-sided predominance observed is consistent with international reports. Although the etiology of PRS remains unclear, early recognition is essential due to its functional, aesthetic, and psychosocial impact. These cases highlight potential underdiagnosis in low-resource settings and emphasize the importance of reporting PRS from underrepresented regions.

Keywords: Parry–Romberg syndrome, Progressive hemifacial atrophy, Neurocutaneous disorder, Facial asymmetry, Afghanistan

Introduction

Parry–Romberg syndrome (PRS), also known as progressive hemifacial atrophy (PHA), is a rare neurocutaneous disorder characterized by unilateral, slowly progressive atrophy of the skin, subcutaneous fat, and facial musculature, with occasional involvement of cartilage and bone (1). It typically begins in childhood or adolescence and progresses over several years before reaching a stable phase (1,3). The pathogenesis of PRS remains unclear, with proposed mechanisms including autoimmune dysregulation, vascular abnormalities, and neurogenic

disturbances (2,4). Although PRS may overlap clinically with localized scleroderma-particularly morphea en coup de sabre-current evidence suggests they are distinct but occasionally coexisting entities (1).

Clinically, PRS presents with progressive unilateral facial thinning due to loss of soft tissue and, in some cases, underlying bone (3,5). Additional features may include dental malalignment, ocular changes such as enophthalmos, and neurological manifestations including headache or trigeminal neuralgia (4,6). Diagnosis is primarily clinical but is typically supported by CT or MRI to assess soft-tissue and bony involvement (4). The estimated global incidence is approximately 1 per 700,000 individuals, though this figure may underestimate the true prevalence due to under recognition and diagnostic challenges (3). One report suggests a female predominance and a higher frequency of left-sided facial involvement (1).

Despite its global distribution, PRS is significantly underreported in South-Central Asia. No confirmed cases have previously been published from Afghanistan (1). Under recognition, limited access to specialized imaging, and low clinical awareness likely contribute to this gap. In this report, we present 2 young Afghan patients with left-sided PRS, providing concise clinical and radiological documentation. These cases represent the first formally reported PRS diagnoses from Afghanistan and help address a notable regional knowledge gap.

Case Presentation

An 18-year-old male presented with a three-year history of progressive left-sided facial atrophy that began insidiously during adolescence. The condition gradually involved the cheek, periorbital region, and lips, without any preceding trauma, infection, or evidence of systemic autoimmune disease. His family history was unremarkable for similar disorders.

Clinical examination revealed marked facial asymmetry with skin thinning, loss of subcutaneous fat, mild hyperpigmentation, and slight deviation of the mouth and nose toward the affected side. Neurological evaluation was normal, while ophthalmologic assessment identified mild enophthalmos. Dental examination demonstrated mild malocclusion corresponding to the atrophic region. Computed tomography (CT) confirmed significant atrophy of the subcutaneous tissues and facial musculature, along with mild hypoplasia of the maxilla and mandible; no intracranial abnormalities were detected. Autoimmune workup-including ANA, ENA panel, RF, ESR, and CRP- was entirely unremarkable. Based on clinical progression and stability over the past 6 months, the disease was classified as being in the late/ stable stage.

The patient underwent autologous fat grafting in the Department of Plastic and Burn Surgery at Esteqlal Hospital, Kabul, Afghanistan, with the aim of restoring facial symmetry and soft tissue volume. Postoperative outcomes were satisfactory, showing appreciable improvement in facial contour and symmetry, maintained graft viability. Clinical and radiologic stability were maintained during 6-month follow-up period (Fig. 1,2).

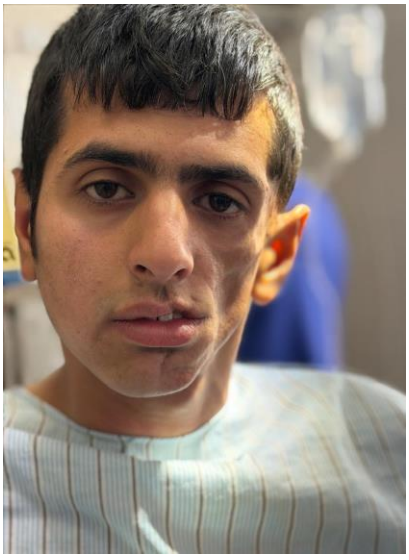

Fig. 1: Frontal view of the first patient before treatment, showing left-sided facial atrophy and asymmetry

Fig. 2: Frontal view of the first patient after fat grafting, demonstrating improved facial symmetry and soft tissue volume

Case 2

A 22-year-old female presented with a five-year history of progressive left-sided hemifacial atrophy, affecting the cheek, periorbital region, lips, and jawline. She reported mild functional discomfort during mastication and expressed concerns regarding facial aesthetics. There was no history of systemic illness or prior trauma.

On clinical examination, significant soft tissue loss was noted on the left side of the face, accompanied by mild enophthalmos, lip deviation, and dental malocclusion corresponding to the atrophic region. Neurological evaluation was unremarkable. CT revealed atrophy of the subcutaneous tissues and masticatory muscles, along with mild bony thinning of the maxilla and mandible, while intracranial structures appeared normal. Autoimmune laboratory testing (ANA, RF, ESR/CRP) was negative. Given the absence of progression over the last 4 months, the disease was categorized as stable/inactive. She underwent autologous fat grafting in the Department of Plastic and Burn Surgery at Esteqlal Hospital, Kabul, Afghanistan, with the goal of restoring facial contour and soft tissue volume. Postoperative outcomes were satisfactory, showing marked improvement in facial symmetry. The patient remained clinically stable with sustained graft volume over 4-month follow-up period (Fig. 3,4).

Fig. 3: Frontal view of the second patient before treatment, illustrating left-sided hemifacial atrophy

Fig. 4: Frontal view of the second patient after fat grafting, showing restoration of facial contour and enhanced symmetry

Discussion

The PRS remains a complex and incompletely understood condition, characterized by variability in clinical presentation, disease progression, and systemic involvement. Our two cases—young Afghan patients with left-sided hemifacial atrophy treated with fat grafting—provide valuable data from an underreported geographic region.

PRS pathogenesis is thought to be multifactorial, involving autoimmune dysregulation, neurogenic mechanisms and microvascular abnormalities inflammatory processes, neurovascular or sympathetic dysfunction, infection, trauma, and genetic factors (1, 6, 16). Evidence of small-vessel neurovasculitis has been proposed as a unifying mechanism explaining progressive soft-tissue and bone atrophy (1, 5, 15). Although recent studies have suggested possible genetic susceptibility, genetic findings remain inconclusive and likely interact with environmental factors rather than representing a primary cause (6, 11, 16). Emerging hypotheses highlight chronic lymphocytic neurovasculitis of small vessels supplying facial tissues, leading to progressive ischemia, tissue loss, and eventual fat and soft tissue atrophy (1,15). Such vascular insufficiency may partially explain deeper structure involvement (muscle, bone) and the variable patterns of disease progression (5, 15).

Isolated reports have suggested potential triggers, including immune-mediated events; for example, PRS reactivation after COVID-19 vaccination has been reported, raising the possibility of immune flares in predisposed individuals (2). Although anecdotal, these observations underscore the importance of clinical vigilance for environmental or immunologic triggers. Classically, PRS manifests as unilateral facial atrophy—most commonly on the left side (1, 6)—with onset in the first or second decade of life (11,12). Both of our patients showed the typical pattern of slow unilateral progression and soft-tissue loss with mild dental malalignment.

The disease typically progresses slowly over 2–20 years before reaching a plateau (6,11). Both of our cases, left-sided and presenting in early adulthood, align with this classical pattern. Beyond superficial soft tissue atrophy, PRS may involve deeper structures including muscle wasting, bone or cartilage hypoplasia (15). Our cases showed mild maxillomandibular hypoplasia consistent with late/stable –stage disease. Dental and occlusal abnormalities, ocular changes such as enophthalmos, and neurologic or intracranial involvement have been reported (4,7,10,13). Advanced neuroimaging has documented white-matter lesions, hemispheric atrophy, leptomeningeal enhancement, and microhemorrhages, particularly in patients with neurologic symptoms such as epilepsy (10,13). In a recent MRI series of 80 patients, 60% exhibited brain abnormalities, and 20% had epilepsy; all patients in the epilepsy subgroup demonstrated ipsilateral MRI lesions (13). Neither of our patients showed neurologic manifestations or intracranial abnormalities, which is consistent with the milder phenotypic spectrum of stable-stage PRS.

Phenotypic severity may vary with age of onset: early-onset cases often show more pronounced deformity and bone involvement, whereas later-onset cases tend to have milder soft tissue changes (15), emphasizing the importance of early recognition and longitudinal follow-up. Currently, there is no definitive cure for PRS. Management is primarily reconstructive and symptomatic, typically delayed until disease stabilization (“burn-out”) to reduce the risk of graft resorption or recurrence (6, 11). Autologous fat grafting remains the most commonly employed technique for restoring facial volume and symmetry, although partial graft resorption is common, necessitating repeat procedures or overcorrection, fat grafting continues to offer reliable aesthetic improvement, even in

resource-limited settings (1,11). Adjunctive use of platelet-rich plasma with fat grafts to enhance graft survival, angiogenesis, and skin quality, though standardized protocols and long-term outcomes remain unclear (9).

Our patients underwent fat grafting achieving satisfactory cosmetic outcomes with maintained symmetry during follow-up. This success demonstrates that reconstructive treatment is feasible even in resource-limited settings, provided surgical expertise is available.

Most published PRS cases originate from Europe, North America, and parts of Asia, with occasional unusual presentations reported from other regions (1, 6, 12, 17). This report provides the first clinically and radiologically confirmed PRS cases from Afghanistan, addressing a significant geographic and epidemiologic gap. Documenting cases from underrepresented regions enhances global understanding of PRS and highlights the need for increased awareness and diagnostic vigilance among clinical in South-Central Asia.

Conclusion

Parry–Romberg syndrome is a rare, progressive disorder with variable clinical expression, making timely diagnosis and appropriate management essential. Early recognition of disease progression and thorough baseline evaluation—including neurologic, ophthalmologic, dental, and radiologic assessment—are critical for guiding treatment decisions and anticipating potential complications.

Given the multisystem nature of the condition, a multidisciplinary approach involving plastic surgery, neurology, ophthalmology, dentistry, and radiology ensure comprehensive care and allows for tailored intervention strategies. Autologous fat grafting remains an effective and accessible technique for restoring facial contour once the disease stabilizes, as demonstrated in our patients who achieved favorable aesthetic outcomes. By reporting two cases from an underrepresented region, this study underscores the importance of broader geographic documentation and reinforces the need for collaborative, multidisciplinary management to optimize functional and cosmetic results in patients with Parry–Romberg syndrome.

Ethical Considerations

Written informed consent was obtained from both patients for participation in this report and for the publication of their clinical data and images. All ethical principles regarding patient confidentiality were strictly observed. This case report was prepared in accordance with the CARE (checklist to ensure completeness, transparency, and standardized reporting quality.

Conflict of interest

The authors declare no conflict of interests.

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