

## Progressive Hemifacial Atrophy With Contralateral Uveitis: A Case Report

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**Introduction:** Progressive hemifacial atrophy, known as Parry-Romberg syndrome (PRS), was first described by Parry in 1825. There is a progressive atrophy of facial tissues including skin, bones and muscles. Ophthalmic disorders are common and include keratitis, uveitis, cataract, ipsilateral enophthalmos, optic neuritis, retinal vasculitis and scleral melting.

**Case Presentation:** We describe a patient with progressive hemifacial atrophy at right facial side who developed granulomatous uveitis and periferic retinal vasculitis in his left eye. We started topical and systemic steroid therapy. Uveitic reaction had regressed almost entirely after a 3-month steroid treatment.

**Conclusions:** The individuals should have multidisciplinary approach for the variety of disorders to maintain the appropriate treatment for a better appearance of the patients.

**Keywords:** Progressive Hemifacial Atrophy; Parry Romberg Syndrome; Uveitis

### 1. Introduction

Progressive hemifacial atrophy, known as Parry-Romberg Syndrome (PRS), was first described by Parry in 1825 (1). It was well-defined by Romberg in 1846 (2). It is a craniofacial disorder with onset in the first or second decade of life in individuals who are morphologically normal at birth (3). Hemifacial atrophy is a rare condition of unknown etiology, characterized by a slowly progressive atrophy of one side of the face, primarily involving the skin, subcutaneous fat, muscles and bones (4, 5). In addition to the progressive hemifacial atrophy, multiple accompanying disorders have been reported; ocular and neurologic changes and ipsilateral progressive body atrophy (4). Ocular involvement is commonly as fat loss around orbit leading to enophthalmos (6).

We describe a patient with progressive hemifacial atrophy who developed granulomatous uveitis at the opposite site. Also, there was posterior segment involvement in the form of periferic retinal vasculitis with optic neuritis.

### 2. Case Presentation

A 28-year-old male patient was referred to ophthalmology department of Gulhane Military Medical Academy in Ankara, Turkey, with blurred vision in his left eye in May 2010. His complaint developed 3 month ago. Visual acu-

ity was 20/20 in both of his eyes. Biomicroscopy and funduscopy findings were normal in the right eye. In his left eye, there was mutton-fat keratic precipitate, + 2 anterior chamber cell and + 2 vitreous cell in biomicroscopy and funduscopy showed minimal optic disc hyperemia. Intraocular pressures, ocular motilities and pupil reactions of both eyes were normal (Table 1).

At the right side, there was madarosis of lateral part of upper and lower eyelid (Figure 1). There was an alopecia area at left eyebrow and a linear sclerotic lesion placed at his left forehead. Also, there was hemifacial asymmetry at his right part of face (Figure 2). Fundus fluorescein angiography (FFA) (HRA, Heidelberg Retinal Angiogram; Heidelberg Engineering GmbH, Dossenheim, Germany) revealed periferic retinal vasculitis and late phase optic disc hyperfluorescence in his left eye (Figure 3). Foveal thickness and retinal nerve fiber layer analysis in ocular coherence tomography (Stratus III OCT™; Carl Zeiss Meditec AG, Jena, Germany) was in normal ranges. Laboratory tests including blood count, erythrocyte sedimentation rate, routine chemistries, antinuclear antibody (ANA), antineutrophil cytoplasmic antibody (ANCA), anti-dsDNA, anticardiolipin and antiphospholipid antibody, fluorescent treponemal antibody absorption (FTA-ABS), anti-TORCH antibodies, C3, IgG and IgM, romatoid factor, C-reactive protein,

serum angiotensin converting enzyme (ACE) levels, tuberculosis, Lyme and Brucella screening were normal. Dental consultation revealed no disorder in mouth, tongue and teeth. After physical examination and dental consultation, the patient was diagnosed as PRS with hemifacial atrophy at right facial side and after a

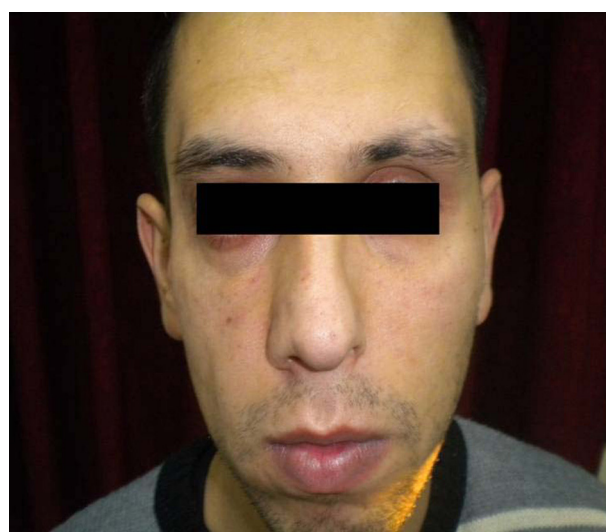
detailed questionnaire the patient noted a subcutaneous fat injection under his right cheek. We diagnosed idiopathic granulomatous panuveitis in his left eye and started the topical and systemic steroid therapy. Uveitic reaction had regressed almost entirely after a 3-month steroid treatment.

**Table 1.** Patient Characteristics

| Variables                                    | Characteristics                                                                                                                                                                                                                                        |
|----------------------------------------------|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| <b>Demographic data</b>                      | A 28-year-old male patient                                                                                                                                                                                                                             |
| <b>Symptom</b>                               | blurred vision in his left eye                                                                                                                                                                                                                         |
| <b>Duration of symptom</b>                   | 3 months ago                                                                                                                                                                                                                                           |
| <b>Visual acuity</b>                         | 20/20 in both of his eyes                                                                                                                                                                                                                              |
| <b>Biomicroscopy</b>                         | Right eye: normal, left eye: there was mutton-fat keratic precipitate, + 2 anterior chamber cell and + 2 vitreous cell                                                                                                                                 |
| <b>Fundoscopy</b>                            | Right eye: normal, left eye: minimal optic disc hyperemia                                                                                                                                                                                              |
| <b>Intraocular pressure</b>                  | Normal in both of his eyes                                                                                                                                                                                                                             |
| <b>Ocular motilities and pupil reactions</b> | Normal in both of his eyes                                                                                                                                                                                                                             |
| <b>Periocular examination</b>                | At the right side there was madarosis of lateral part of upper and lower eyelid. There was an alopecia area at left eyebrow and a linear sclerotic lesion placed at his left forehead. Also, there was hemifacial asymmetry at his right part of face. |
| <b>Fundus fluorescein angiography</b>        | Periferic retinal vasculitis and late phase optic disc hyperfluorescence in his left eye                                                                                                                                                               |
| <b>Ocular coherence tomography</b>           | Foveal thickness and retinal nerve fiber layer analysis was in normal ranges                                                                                                                                                                           |
| <b>Dental consultation</b>                   | Hemifacial atrophy of right facial side and no disorder in mouth, tongue and teeth                                                                                                                                                                     |
| <b>Laboratory tests</b>                      | Normal                                                                                                                                                                                                                                                 |
| <b>Previous treatment</b>                    | Subcutaneous fat injection under his right cheek                                                                                                                                                                                                       |



**Figure 1.** Madarosis of Upper and Lower Eyelid



**Figure 2.** Hemifacial Asymmetry at Right Side of the Face and Alopecia at Left Eyebrow



**Figure 3.** Periferic Retinal Vasculitis and Late Phase Optic Disc Hyperfluorescence

### 3. Discussion

The etiology of PRS is unknown and its pathology is rare. There are many hypotheses explaining the etiology; a relationship with either localized scleroderma, trauma, infection, an autoimmune disorder, a trigeminal neurovasculitis, heredodegenerative factors or a defect of the sympathetic nervous system have been suggested as possible causes (4, 5). There is a progressive atrophy of facial tissues including skin, bones and muscles (4, 7). The initial lesion in PRS is commonly a small area of abnormal pigmentation immediately superior to the eyebrow, then this lesion enlarges and becomes atrophic (8). There are patients presenting with a demarcation line between normal and abnormal skin, reminding a linear scar, known as 'coup de sabre', as we reported in this case (4, 9). Vitiligo, alopecia of the eyebrow and alopecic patches of the scalp were also reported (10). The most important features of this pathology are the enophthalmos, the deviation of mouth and nose to the affected side and unilateral exposition of teeth, among others (4, 9). Ophthalmic disorders are common and include keratitis, uveitis, cataract, ipsilateral enophthalmos, optic neuritis and atrophy, pupil anomalies ranging tonic to Horner pupil, heterochromia iridis, retinal pigmentary changes, progressive refractive changes, contralateral ocular muscle involvement, glaucoma, hypotony, retinal vasculitis, and scleral melting (3). Granulomatous uveitis with posterior

segment involvement is a rare condition as we reported in our case. Also, we think it is an interesting condition that although hemifacial atrophy was at right side, the uveitis was in the left eye. Parry-Romberg syndrome is an auto-limitable condition and there is no certain cure (9). The presence of specific signs of PRS in this case and performing the appropriate examinations are the positive points of the present study. Our study limitation is the lack of previous history of the patient. The individuals should have multidisciplinary approach of physicians, dentists, phonoaudiologists and psychologists for the variety of disorders to maintain the appropriate treatment for a better appearance to the patients (6, 9).

### Authors' Contributions

Study concept and design: Onder Ayyildiz, and Simel Ayyildiz. Drafting of the manuscript: Onder Ayyildiz. Critical revision of the manuscript for important intellectual content: Ali Hakan Durukan, and Gungor Sobaci.

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