Progressive hemifacial atrophy with ciliary body atrophy and ocular hypotony

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Progressive hemifacial atrophy (PHA) is a disease of unknown etiology affecting one-half of the face. Ocular involvement is uncommon. Atrophy of iris is rare, with only a few cases of partial atrophy being reported in the literature. We report a case of total atrophy of iris and ciliary body with associated ocular hypotony in a 16-year-old girl with PHA. We believe this is the first reported case of complete atrophy of iris and ciliary body in PHA. Ocular hypotony in PHA was thought to be due to intra-ocular inflammation. However in our case it appears to be secondary to severe atrophy of the ciliary body.

Key words: Ciliary body atrophy, iris atrophy, ocular hypotony, progressive hemifacial atrophy

Parry-Romberg syndrome or progressive hemifacial atrophy (PHA) affects the skin, soft tissue and skeletal structures of the face. Ocular involvement occurs in 10–35%, ranging from uveitis to enophtalmos.\(^1\,\(^2\)\) Ocular hypotony in PHA has been attributed to intra-ocular inflammation. Atrophy of iris in PHA is very rare, with only a few cases of partial iris atrophy being reported previously. We report a case of PHA with complete atrophy of iris and ciliary body. We hypothesize that the accompanying severe ocular hypotony may be secondary to atrophy of the ciliary body.

Case Report

A 16-year-old girl presented with painless progressive diminution of vision in right eye since 2 years, associated with glare. Left eye was asymptomatic. She noticed asymmetry of her face at 13 years of age and was diagnosed with PHA. The asymmetry was progressive, and she received fat grafting on the right side of her face. There were no systemic manifestations like seizures, migraine, intellectual deterioration, trigeminal neuralgia or neuropsychiatric disturbances.

General physical examination showed atrophy of the right half of the face involving subcutaneous tissue, muscle and bone [Fig. 1]. There was no neurologic deficit or oral involvement. Visual acuity was 20/60 in right eye and 20/20...
in left eye. Best-corrected visual acuity was 20/20 with +2.25 cylinder at 90° in right eye.

Right eye had enophthalmos and severe hypotony with intraocular pressure of 4 mm of Hg by Goldmann’s applanation tonometer. Intra-ocular pressure of her left eye was 16 mm of Hg. Slit lamp examination of right eye revealed marked iris atrophy amounting to aniridia. The pupil was fully dilated and nonreactive to light and accommodation [Fig. 2]. Lens showed partial subluxation inferiorly. Fundus examination showed hyotonic maculopathy with internal limiting membrane folds radiating across the macula [Fig. 3]. Gonioscopy showed open angles with broad ciliary body band. Ultrasound biomicroscopy revealed atrophic iris and ciliary body [Fig. 4]. Lids, adnexa and ocular motility were normal. There was no evidence of iridocyclitis or retinal vasculitis in this patient.

At 6 months follow-up, the intraocular pressure in her right eye has further reduced to 2 mm of Hg. However, best-corrected visual acuity remains 6/6 with the same correction. Fundus and ultrasound bio-microscopy findings remained the same.

**Discussion**

The clinical features of PHA were initially observed by Caleb Hillier Parry in 1825 and subsequently described by Moritz Heinrich Romberg in 1846. Although the exact etiology is unknown it is believed to be related to localized facial scleroderma. The pathological process consists of chronic lymphocytic infiltration around neurovascular bundles. PHA has a slight female preponderance with age of onset around 5–15 years. It is usually unilateral. There are progressive atrophy and deformation of one side of the face, resulting in unilateral facial atrophy, deviation of mouth and nose toward affected side. PHA has been linked with various neurologic, ophthalmic, cardiac, rheumatologic, infectious, endocrine, maxillofacial and orthodontic manifestations.

Involvement of the eye is uncommon and reported to be in the range of 10–35%. Various ophthalmic associations so far reported are listed in Table 1. A few cases of partial iris atrophy have been reported in the literature. Preechawat et al. reported a case of partial iris atrophy with sluggishly reacting 3 mm-pupil. Two cases of partial iris atrophy with poor light reflex were reported by Miller and Spencer. Dawczynski

![Figure 1: Hemifacial atrophy involving the right side of the face](image1)

![Figure 2: Slit lamp diffuse illumination showing near total iris atrophy with dilated pupil](image2)

![Figure 3: Fundus photo showing hypotonic maculopathy](image3)

![Figure 4: Ultrasound biomicroscopy of both eyes showing iris with ciliary body atrophy on the right side](image4)
et al. reported a patient with pupillary abnormalities with enophthalmos. However, the condition of the ciliary body was not mentioned. All these cases reported partial atrophy of the iris. In our case, there was total atrophy of iris and ciliary body and with ocular hypotonia. Ocular hypotony has been attributed as being secondary to intraocular inflammation. But in our case we believe ocular hypotony occurred secondary to ciliary body atrophy. To our best knowledge, such a combination of features has not been described in the literature before. Atrophy of iris and ciliary body may be a part of generalized facial atrophy.

No specific treatment has been mentioned for ocular involvement PHA. Hypotony has been managed by silicone oil injections in some cases. Antiinflammatory drugs might be given in patients with active inflammatory disease. Facial and oculoplastic surgery may be done for cosmetic purposes.

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Cite this article as: Kini TA, Prakash VS, Puthalath S, Bhandari PL. Progressive hemifacial atrophy with ciliary body atrophy and ocular hypotony. Indian J Ophthalmol 2015;63:61-3.

Source of Support: Nil. Conflict of Interest: None declared.

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**Table 1: Various ocular manifestations associated with Parry Romberg syndrome**

| Orbit | Lids and adnexa | Extraocular muscles | Anterior segment | Posterior segment |
|-------|----------------|--------------------|-----------------|-----------------|
| Enophthalmos | Lid atrophy | Exotropia | Decreased corneal sensitivity | Vitritis |
| Phthisis bulbi | Lid retraction | Esotropia | Band keratopathy | Papillitis |
| Nerve paresis | Loss of cilia | Hypotropia | Episcleritis | Retinal telangiectasis |
| Blepharoptosis | Restrictive strabismus | Extraocular muscle thinning and impairment | Uveitis | Retinal edema |

**Orbit**

- Enophthalmos
- Phthisis bulbi
- Nerve paresis

**Lids and adnexa**

- Lid atrophy
- Lid retraction
- Loss of cilia
- Blepharoptosis

**Extraocular muscles**

- Exotropia
- Esotropia
- Hypotropia
- Restrictive strabismus

**Anterior segment**

- Decreased corneal sensitivity
- Band keratopathy
- Episcleritis
- Uveitis

**Posterior segment**

- Vitritis
- Papillitis
- Retinal telangiectasis
- Retinal edema
- Retinal pigment lesions