

Progressive Hemi-facial Atrophy and Keratoconus

Progressive hemifacial atrophy (Parry-Romberg syndrome, PHA) is characterised by slowly progressive atrophy, usually involving one side of the face, and may be associated with ocular manifestations which include enophthalmos, restrictive strabismus and hypotony. We report a case of keratoconus seen in a patient with PHA after obtaining approval from our Institutional Review Board. We are unaware of previous reports in the literature describing an association between the two.

A 40-year-old female with a known diagnosis of left-sided PHA was referred to the Cornea Services for progressive blurring of vision in both eyes. On examination, her visual acuity in the right eye (RE) was 6/120 correctable to 6/18 and in the left eye (LE) was 6/60 correctable to 6/30. She had left-sided facial hemiatrophy with an enophthalmos (Fig. 1) of 5 mm. Her ocular movements were full and she was orthophoric. Intra-ocular pressure was 7 mmHg in the RE and 5 mmHg in the LE. Anterior segment examination revealed Vogt striae, Fleischer's ring and distorted mires on Placido's disc in both eyes. Posterior segment examination was normal. A corneal topography was done which confirmed the diagnosis of keratoconus (Figs. 2). Her manifest refraction in the RE was -4.00/-7.00 x 170 and in the LE was +3.00/-11.00 x 180.

The patient was offered a trial of rigid gas permeable lenses (RGP). With Toric RGP lenses, her best corrected distant acuity was 6/7.5 in the RE and 6/6 in the LE.

On follow-up, she complained of excessive lens movement in her LE and was noted to have a nasally displaced contact lens. She was offered a trial of scleral lens and piggy back lens, but a proper contact lens fit could not be obtained. She was counselled for penetrating keratoplasty in view of contact lens intolerance, which she is currently considering.

PHA has been previously associated with enophthalmos,¹ motility problems,² refractive errors, hypotony, glaucoma and retinal vasculitis but not with keratoconus.²⁻⁴ Besides

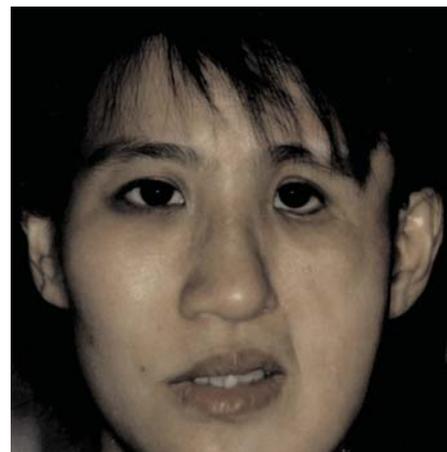


Fig. 1. Photograph depicting left sided facial atrophy and enophthalmos in a patient with Parry Romberg Syndrome.

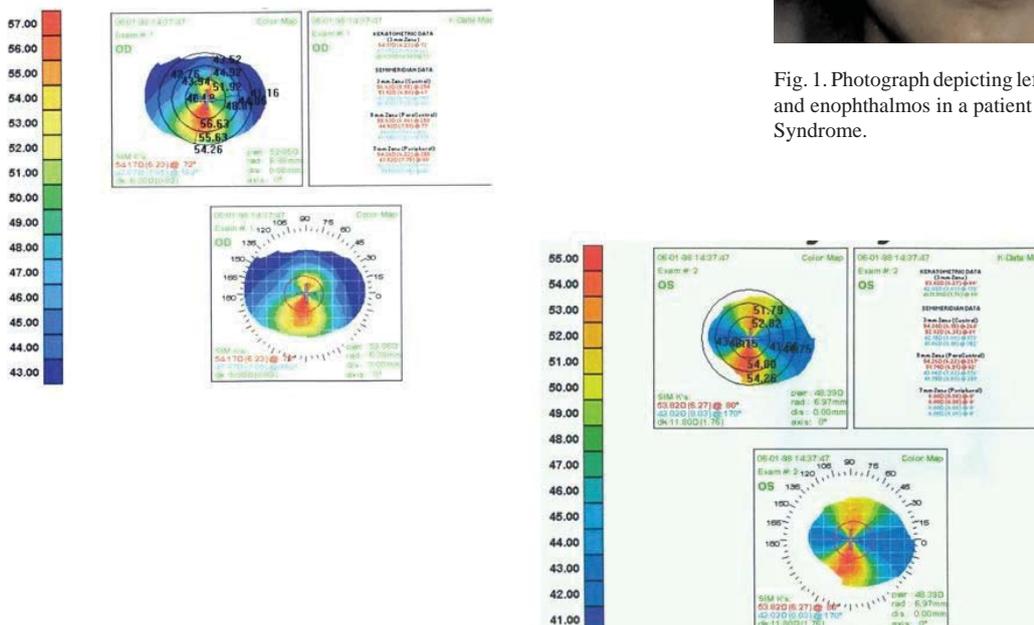


Fig. 2. Topographic map of the patient's right and left eye showing advanced keratoconus in both eyes.

enophthalmos, hypermetropia, high astigmatism and a low intra-ocular pressure, our patient had advanced keratoconus in the LE. Interestingly, contact lens fitting posed a challenge not only because of advanced keratoconus but also because of the associated enophthalmos.

The cause of PHA is unknown. Theories proposed include neurogenic, vascular, exogenous insult and autoimmune mediated processes. A popular hypothesis is the vasomotor trophoneuritis theory, involving the sympathetic nervous system which results in the atrophy of facial tissues.¹ Similarly, an impairment of corneal innervation has been suggested to play a role in the pathogenesis of keratoconus.⁵ We postulate that impaired trophic responses could explain the association of keratoconus with PHA in our patient.

REFERENCES

1. Miller MT, Spencer MA. Progressive hemifacial atrophy. A natural history study. *Trans Am Ophthalmol Soc* 1995;93:203-15.
2. Rudolph G, Haritoglou C, Kalpadakis P, Schworm HD, Ehrh O, Boergen KP. Hemifacial atrophy (Parry-Romberg syndrome, #141300) with

papillitis, retinal alterations, and restriction of motility. *J AAPOS* 2002;6:126-9.

3. Ong K, Billson FA, Pathirana DS, Clifton-Bligh P. A case of progressive hemifacial atrophy with uveitis and retinal vasculitis. *Aust N Z J Ophthalmol* 1991;19:295-8.
4. Cohen JS. Congenital nonprogressive facial hemiatrophy with ipsilateral eye abnormalities and juvenile glaucoma. *Ann Ophthalmol* 1979;11:413-6.
5. Ruddle JB, Mackey DA, Downie NA. Clinical progression of keratoconus following a sixth nerve palsy. *Clin Exp Ophthalmol* 2003;31:363-5.

Arundhati Anshu,¹*MRCSEd (Ophth), MMed (Ophth),*
Li Lim,¹*FRCSEd (Ophth)*

¹ Singapore National Eye Centre, Singapore

Address for Correspondence: Dr Lim Li, Cornea and Refractive Services, Singapore National Eye Centre, 11 Third Hospital Avenue, Singapore 168251.

Email: lim.li@snecc.com.sg