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Case Report

Managing Spontaneous Scleral Melt and Intractable Intra Ocular Pressure in Parry-Romberg Syndrome - A 20 Years Case Journey

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Abstract

Purpose: To report the 20-years case journey of review and management of intractable intraocular pressure (IOP) following surgical interventions in an unusual case of spontaneous scleral melting and macular hypotony in a patient with diagnosed Parry-Romberg Syndrome (PRS).

Materials and Methods: This is an observational case report over 20 years. A 59-year-old Caucasian male patient with known PRS was referred with unilateral "swelling of sclera" above cornea. Ocular examination revealed a unilateral overhanging conjunctival bleb due to spontaneous auto-scleral melt.

Over next 8 years, he initially developed hypotonus maculopathy necessitating bleb repair with scleral patch graft (SPG), resulting in intractably high IOP requiring urgent trans-scleral cyclophotoablation.

Results: SPG stabilised the best corrected vision (BCVA) and the IOP. At the 20-years review, the BCVA remains stable (6/12) and the IOP remained well-controlled. Early cataractous changes and an early bleb recurrence and exposure remained the other notable findings. The other eye remained stable throughout.

Conclusion: Parry-Romberg syndrome is an unusual disorder of unknown aetiology. Although mainly involving the neuro-cutaneous system, PRS co-exists with multi-systemic problems. The ocular involvement, though uncommon can be extensive and varied. It can result in both ocular hypotony and ocular hypertension.

We share our experience of long-term management of various ocular complexities associated with such a case. Literature review leads us to believe our case is the second-ever case, reported for the management of spontaneous scleral melt and the intractable IOP associated with PRS. This case report is also the longest reported follow-up experience for PRS related ophthalmic manifestations.

Keywords: *Parry-Romberg Syndrome; Spontaneous Scleral Melt; Intractable Intraocular Pressure; Conjunctival Bleb; Ocular Hypotony*

Abbreviations

PRS: Parry-Romberg Syndrome; SPG: Scleral Patch Graft; IOP: Intraocular Pressure; BCVA: Best Corrected Visual Acuity

Introduction

Parry-Romberg syndrome is a rare disorder of unknown aetiology [1,2]. Multiple hypotheses on aetiology include infective, degenerative, inherited and traumatic causes [3]. Autonomic dysfunction has also been suggested as a possible pathogenesis. Progressive hemi facial atrophy (Parry-Romberg syndrome, Romberg syndrome, PRS) is characterized by slowly progressive atrophy, frequently involving only one side of the face, primarily affecting the subcutaneous tissue and fat. Progressive atrophy of face and skin is seen in 75% of cases [3]. The atrophy of the face can be associated with atrophy of subcutaneous fat, the muscles and sometimes of the bone (maxillary and palatine) [4]. It frequently involves ipsilateral muscles of the tongue and soft palate although atrophy can extend to the neck and beyond in upper trunk [5]. This condition ultimately stabilises after periods of slow progression and remission. Progressive hemi facial atrophy has also been associated with transient numbness of the extremities and seizures with white matter densities in the cerebral hemispheres [6].

10% - 35% of all cases show ocular involvement in PRS or progressive hemi facial atrophy [3,7]. The most common association is enophthalmos due to the atrophy of orbital fat. Other reported ocular features include Horner's syndrome, mydriasis, heterochromic cyclitis, ptosis, ocular motility disorder, nystagmus, blepharophimosis, band keratopathy, retinal vascular abnormalities, scleral melt and primary corneal endothelial failure requiring penetrating keratoplasty [8-10].

We consider that this is second case report of spontaneous scleral melt after Hoang-Xuan., *et al*. considered scleral melt as a late manifestation of Romberg disease [3,8]. We share our experience in managing complex issues related to such a case over a period of twenty years.

Case Report

In the year 2000, a 59-year-old Caucasian gentleman was referred to the eye clinic with a possible "swelling of the sclera" by his general practitioner with slight unilateral ocular irritation being the only presenting symptom. He was previously already diagnosed as a case of PRS at the age of 38 years. He had undergone right facial reconstructive plastic surgery in 1986 and still had persisting atrophic changes on right side of the face and of the tongue musculature (Figure 1). Patient also complained of transient numbness of the right leg which seemed to be associated with PRS.

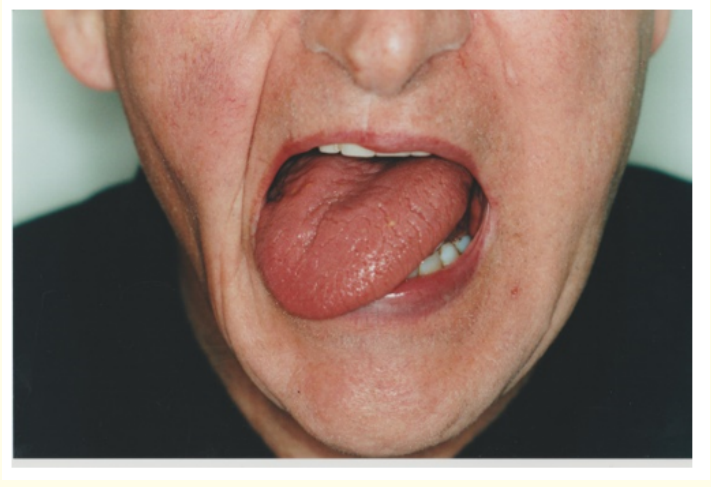

Figure 1: *Atrophy of the right sided facial and tongue musculature.*

Unilateral ocular irritation was the only presenting symptom. There was no history of trauma, previous ocular surgery or any previous episodes of viral infections. On slit lamp examination, there was an area of scleral defect with conjunctival bleb formation over three clock hours supero-temporally (Figure 2). The BCVA in the affected eye was 6/9 (Snellen's) and the IOP was 9 mm Hg (Goldmann Applanation tonometer). An extensive laboratory evaluation was performed including full blood count, ESR, liver function test, thyroid function test, rheumatoid factor and antinuclear antibody essay. All the results were within the normal range.

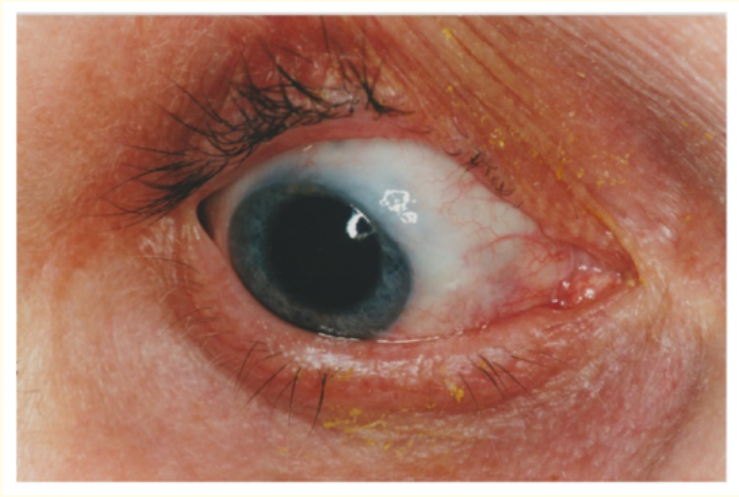

Figure 2: *Area of scleral defect with conjunctival bleb formation.*

The patient was prescribed topical lubricant drops and his vision and IOP remained stable over the serial ocular reviews. He was informed of significant risk of endophthalmitis due to thin bleb in 2008 and was started on chloramphenicol 0.5% preservative free drops once daily in right eye, prophylactically. Over next 12 months his dysaesthesia became significant. On examination, the bleb became bigger and IOP dropped to 3 mm Hg with reduction in visual acuity by 2 lines. Ocular examination showed a large area of scleral melt (3 mm by 2 mm) with 3 adjacent small satellite areas of scleral perforation. Further, Choroidal folds suggestive of early Hypotony maculopathy were also noted. He was keen for surgical repair with donor scleral graft. Following a detailed counselling regarding secondary rise of IOP due to irreversible angle damage, a scleral repair with donor corneo-scleral graft was performed in July 2010.

Postoperatively, he developed significant pain with IOP of 50 mm Hg. He was admitted for IOP control with intravenous Acetazolamide, topical Glaucoma drops. The intractable IOP further required intravenous Mannitol and Trans-scleral Cyclophotoablation.

Over next week, his IOP dropped and came down to 9 mm Hg. The BCVA improved to 6/6 and a reversal of hypotony maculopathy was noted at 3 months.

Over the next 10 year of follow-up, the bleb has recurred with IOP being stable and no signs of hypotony. He did develop corneal Dellen under the bleb on 2 occasions needing intensive ocular lubricants.

At 20 years of total follow-up, the BCVA was 6/9 in the affected eye with early cataractous changes. Further, orbital fat atrophy (Figure 3) was noted which left the bleb slightly exposed. A lid procedure was considered to cover the bleb (surgically induced ptosis) but he chose not to opt for this surgery as he is subjectively comfortable with topical Ocular lubricants. No new systemic features associated with PRS were noted. He remains under our ongoing review.

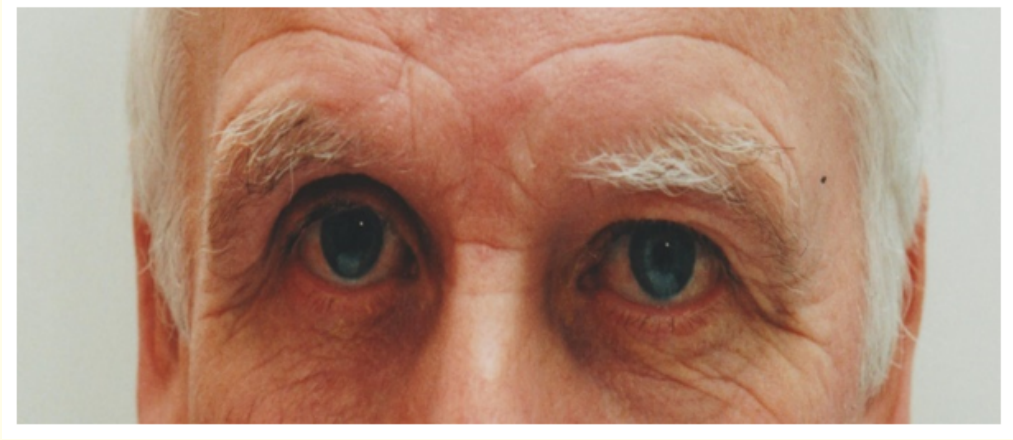

Figure 3: *Consequent Orbital fat atrophy.*

Discussion and Conclusion

PRS is a rare, self-limiting, largely neuro-cutaneous disorder of unknown aetiology. It usually manifests within the first two decades of life and is more common in females. Though, neurological involvement is commonest, this syndrome can also have associated rheumatological, endocrine, cardiac, maxillo-facial, orthodontal and ophthalmological disorders. Neurological manifestations can range from migraine, intracranial vascular anomalies, atrophic changes in brain and hemiplegia.

Ocular involvement can be associated with up to 35% of all cases and can be manifested pre, per or post facial atrophy. Though corneal and retinal changes are more prevalent, palpebral pigmentation, uveitis, heterochromic cyclitis, ptosis and phthisis have been reported. The commonest periocular manifestations include enophthalmos, eyelid retraction, lagophthalmos, asymmetrical eyebrow contour, alopecia and a late-onset pseudo coloboma of lid. The other neuro-ophthalmological associations include Horner's syndrome, tonic pupil, optic neuropathy, neuro-retinitis and third nerve palsy to name a few.

Our experience, spans over two decades and reveals the glaucoma surgeon's journey encompassing both the extremes of IOP changes (hypotony and intractably high IOP) encountered secondary to the condition. The scleral patch graft in such a case can be extremely tricky due to the atrophic nature of the tissues involved, the fluctuating IOPs and the risks of graft failure owing to periocular changes. The ocular hypotony can be further worsened secondary to phthisis and choroidal folding.

Scleral melt should be considered as an established feature of PRS. In early stages, hypotony may not be severe but it causes irreversible changes in the drainage angle. Scleral repair with donor graft causes intractable intraocular pressure which can potentially cause irreversible visual loss. A detailed discussion about these risks is required with documentation of understanding of risks in such rare cases. Hypotony maculopathy can be reversed if treated early. A tertiary centre is perhaps the best place to carry this corrective surgery as intensive postoperative treatment is required. All these efforts may stabilise the vision in the long term but as this is a progressive disease some recurrence of symptoms may occur.

Literature evidence suggest only one previous case of scleral melt documented with PRS [8]. Our case is possibly the second ever reported spontaneous scleral melting in PRS. To our knowledge, this is also the longest reported Ophthalmology case journey and ocular follow-up experience of PRS case. Therein, lies the novelty of our case journey. We hope that detailed description of our experience of dealing with this complex condition will help all others in future.

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All the authors attest that they meet the current ICMJE criteria for authorship.

Patient Consent

Informed written consent for publication of personal identifying clinical record details and facial photographs was obtained from the patient.

Declaration of Conflicting Interests

The authors have no potential conflicts of interest with respect to the authorship, research and/or publication of this case report.

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