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Case Report

Parry Romberg Syndrome with contralateral iridocorneal endothelial syndrome: a unique case

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ABSTRACT

Parry-Romberg syndrome (PRS) is a rare disorder which causes progressive hemifacial atrophy, with ocular manifestations like hypotony, enophthalmos and corneal edema on the ipsilateral atrophic side. This is a report of a unique case of PRS with contalateral manifestations like ectropion uvea, correctopia and endothelial deposits, along with polymegathism and pleomorphism seen on specular microscopy suggestive of Iridocorneal Endothelial (ICE) Syndrome. ICE syndrome and PRS have not been reported together in any literature so far. This case highlights the importance of a thorough glaucoma workup and corneal examination on the atrophic facial side as well as on the apparently normal side in all cases of PRS.

Keywords: Parry Romberg syndrome, ICE syndrome, Hemifacial atrophy, Glaucoma

INTRODUCTION

Parry-Romberg syndrome is a rare disorder characterized by hemifacial atrophy, with ocular involvement being seen in 10-30% of people on the ipsilateral atrophic side. Bilateral involvement is seen rarely, mainly involving the brain.[1] Iridocorneal Endothelial (ICE) Syndrome is an ocular disorder characterized by “hammered silver" or "beaten bronze" appearance of the corneal endothelium leading to corneal edema, iris changes, elevated intra ocular pressure and secondary angle-closure glaucoma.[2] ICE syndrome on the normal or atrophic facial side in a case of Parry Romberg syndrome has never been reported in the literature.[3]

CASE REPORT

The patient, a 25 years old male, was a known case of Parry Romberg Syndrome (Figure 1), having left sided hemifacial atrophy. For this, he underwent skeletal reconstruction with costal cartilage and soft tissue reconstruction with de-epithelized anterolateral thigh flap and fat grafting three years ago in the Department of Plastic Surgery.

He presented to the Department of Ophthalmology with diminution of vision for the past 6 months in the eye on the apparently normal side of the face and poor vision in the eye on the atrophic side since childhood.

On presentation, the best corrected visual acuity (BCVA) on the apparently normal right side of the face was 6/60. The cornea of this eye was clear with a specular count of 2679/mm2 and rest of the anterior segment, dilated fundus examination and intraocular pressure (IOP) was normal. On the atrophic left side, his vision on presentation was perception of light with inaccurate projection of rays. There was also enophthalmos, lagophthalmos with vascularized leukomatous opacity, hazy anterior chamber details and low IOP, suggestive of atrophic bulbi.

The patient underwent uneventful right eye cataract surgery (phacoemulsification) with posterior chamber foldable hydrophobic acrylic intraocular lens (Hoya) implantation. The post-operative unaided vision was 6/6 with a clear cornea and round pupil. The intraocular pressure with Goldmann applanation tonometer was 16 mmHg. The patient was prescribed topical antibiotics and steroids, followed up for 6 weeks and then discharged from the outpatient clinic.

Figure 1: Hemifacial atrophy progressing with age.

Figure 2: Preoperative photo is of the pre senile cataract. Post-operative pictures showing subsequent development of correctopia, ectropion uvea and endothelial deposits.

The patient presented with gradual blurring of vision in the right eye 6 months after the surgery. The BCVA in the right eye had dropped to 6/9. On slit lamp examination, the presence of powdery deposits and pigments on the central corneal endothelium was noted. Ectropion uvea was present nasally distorting the shape of pupil (Figure 2).

The IOP with Goldmann applanation tonometer was 18 mm Hg. The optic disc examination revealed a C:D ratio of 0.3:1 with a healthy neuroretinal rim. The anterior chamber angle was open on gonioscopy. The visual field was normal. The central corneal thickness of was 570 um. UBM showed a mild atrophy of the ciliary body along with mild blunting of the ciliary processes. Specular and confocal examination revealed polymegathism, pleomorphism, and a reduced nerve supply with a total endothelial cell count of 1554/mm2. The coefficient of variation was 41 and the hexagonality 45. On the basis of clinical examination and investigations, a diagnosis of ICE syndrome was made. The patient was advised regular follow ups for IOP measurement and slit lamp examination.

DISCUSSION

PRS is a rare disorder characterised by hemifacial atrophy in which subcutaneous tissues, muscles and osteocartilaginous structures undergo shrinkage and degeneration beneath the skin.[4-7] It is unilateral, non-hereditary and more common in females. The onset is usually in the first decade of life. The etiology of PRS is unknown. Association with herpes and Borrelia burgdoferi infection has been seen.[8-10] Neurological, orthodontal, rheumatological, endocrine, cardiac and ophthalmological associations have been seen. Ocular involvement is seen in 10-30% cases of PRS.

ICE syndrome is an ocular disorder characterised by “hammered silver" or "beaten bronze" appearance of the corneal endothelium leading to corneal edema, iris atrophy, elevated intra ocular pressure and secondary angle-closure glaucoma.[2] Pathologically, the corneal endothelium is replaced by epithelial–like cells that tend to migrate over the trabecular meshwork upto the peripheral iris. Contraction of this layer results in high peripheral anterior synechiae (PAS) and iris changes such as iris atrophy, ectropian uvea, nodule formation which are characteristics of ICE syndrome. High intraocular pressure can be due to advancing corneal endothelium covering trabecular meshwork resulting in open angle glaucoma or it can be because of high PAS leading to angle closure glaucoma. ICE syndrome has never been reported to be associated with PRS.

Patients with PRS have been usually reported to show hypotony of the eye due to atrophy of the ciliary processes.[11] Corneal edema may also be present along with pigments on endothelium. However, ICE syndrome has not been reported in association with PRS. To the best of our knowledge, no case of Parry Romberg Syndrome with presenile cataract and subsequent ICE syndrome on the normal side has been reported yet.

CONCLUSION

ICE syndrome and Parry Romberg Syndrome have not been reported together in any literature, to the best of our

knowledge. This case shows that one must be aware about the possibility of involvement of ICE and glaucoma on the apparently normal side in a case of PRS. A thorough glaucoma workup and corneal examination on the normal, as well as the atrophic facial side in all cases of Parry Romberg syndrome is hence mandatory.

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