Outcome of Strabismus Surgery for Exotropia in a 4-year-old with Silver-Russell Syndrome (SRS): A Case Report

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ABSTRACT

Silver-Russell Syndrome (SRS) is a rare disorder associated with prenatal and postnatal growth retardation with associated characteristic facial and ocular features including strabismus. We report the outcome of strabismus surgery performed for exotropia in a 4-year-old patient with SRS.

The patient presented with decreased visual acuity and constant exotropia of the right eye noted since 3 months of age. Systemic SRS characteristics consisted of relative macrocephaly, short stature, forehead prominence and stunted growth pattern. An X-pattern exotropia is consistent with bilateral tight lateral recti muscles with over-elevation in adduction of the left eye was present. Patient underwent unilateral right lateral rectus recession and right medial rectus resection for a 50-prism diopter constant exotropia. Patient had a favorable outcome of within 8 prism diopters from orthotropia at 1st, 3rd-, 6th- and 12th-month post-operatively.

Keywords: Silver-Russell Syndrome, exotropia, X-pattern exotropia, strabismus surgery, macrocephaly

INTRODUCTION

Silver-Russell Syndrome (SRS) is a distinct, rare disorder associated with prenatal and postnatal growth retardation with characteristic features of relative macrocephaly or enlargement of the head, forehead prominence, asymmetry in growth patterns and feeding difficulties.[1] SRS occurs in 1 in 30,000-100,000 people in the United States although the exact incidence remain unknown worldwide.[2] Notable ophthalmologic features associated with this syndrome are decreased visual acuity, refractive errors commonly hyperopia or farsightedness, difference in refraction between two eyes of more than 1 diopter, shorter axial lengths, small optic discs and large retinal vessels.[3] While concomitant refractive errors and large anisometropia in SRS may predispose the patients to strabismus and amblyopia, it is uncommon for SRS patients to present with strabismus. Outcomes of strabismus surgery in SRS patients remain unknown. The significant characteristic craniofacial features of SRS patients pose a challenge to the ophthalmologist in treating patients both medically and surgically.

We present the outcome of strabismus surgery in a pediatric Filipino patient with SRS who had a large-angle exotropia.

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CASE PRESENTATION

A 4-year-old female was brought for consultation for constant exotropia of the right eye. The exotropia was noted since 3 months of age. This began as a non-alternating intermittent exotropia without fixation preference. There was no abnormal head posturing, abnormal head tilt and nystagmus. At 9 months of age, exotropia became constant. No consultation was done previously.

The patient was born preterm at 35 weeks age of gestation via spontaneous vaginal delivery for premature labor contractions and oligohydramnios and was small for gestational age at 1.580 kg. The mother denied intake of medications and prohibited drugs during pregnancy.

The patient was diagnosed with SRS based on systemic clinical features of macrocephaly, short stature, forehead prominence, stunted growth pattern, narrow chin and micrognathia. Ocular examination showed visual acuity of finger play at 20 feet on both eyes, with a left eye fixation preference; pupils were briskly and equally reactive to light with no afferent pupillary defect. External adnexae were normal. Lid opening and closing were unremarkable. No lagophthalmos was noted. Extraocular muscle examination showed a large angle exotropia with bilateral adduction deficits. Alternate prism cover test (APCT) revealed 50 prism diopters exotropia with and without spectacle correction (Figure 1) with no episodes of orthotropia during the strabismus measurement. Full cycloplegic refraction showed a refraction of -1.50 sph with -4.00 cylinder at axis 180 degrees on the right eye, and plano with -1.50 cylinder at axis 180 degrees on the left eye. Fundoscopy showed normal fundi on both eyes without signs of retinopathy of prematurity. Optic nerves were unremarkable. Patching for amblyopia was initiated on the left eye because of the left eye preference. On subsequent visits, refraction was stable while the deviation was persistent at 50 prism diopters using APCT.

Cranial magnetic resonance imaging (MRI) with orbital cuts done showed unremarkable orbits and extraocular muscles. Imaging was done to pre-operatively identify the status of the extraocular muscles.

Intraoperatively, a forced duction test showed bilaterally tight lateral recti muscles. A unilateral right lateral rectus recession of 8 mm and a medial rectus resection of 7mm for 45-50 prism diopters exotropia was performed based from Parks surgical table of strabismus surgery for exotropia. Intraoperatively, right lateral and medial recti insertions were at 5 millimeters and 4 millimeters from the limbus, respectively indicating the more anterior location than the regular LR and MR insertions. In addition, the lateral rectus was superotemporally displaced that could possibly be explained by the chronic and longstanding exotropia, while the thin medial rectus was anteriorly inserted.

The patient's exotropia was corrected (Figure 2B) to within 6-8 prism diopters from orthotropia up to her first month follow-up. This further improved to 2-4 PD after four months and patient's eye became orthotopic after one year. There was no diplopia. Myopic spectacles were continued and patching of left eye was tapered to 2-3 hours/day.

DISCUSSION

Silver-Russell Syndrome is a sporadic, rarely genetic, syndrome which affects premature and small for gestational age children.[2] The pathophysiology of SRS is still unknown. The diagnosis of SRS was made after the clinical features of patient fulfilled Netchine-Harbison clinical scoring system.[1] The patient fulfilled 5 out of 6 characteristics: small for gestational age (SGA), postnatal growth failure, relative macrocephaly at birth, protruding forehead and low body mass index. The patient did not have feeding difficulties. Common associated ocular findings in patients with SRS are subnormal visual acuity, refractive errors with more hyperopes than myopes,[3] anisometropia of >1 diopter, shorter axial lengths, small optic discs and large retinal vessels.[4]

For the ophthalmologist, the characteristic ocular features in patients with SRS, such as alterations in the position and thickness of the extraocular muscles, pose a challenge in management Our case presented with a large-angle exotropia, large anisometropic astigmatism and myopia. Orbital imaging was requested in order to help identify and prepare the surgeon for the anticipated difficulties, such as locating an abnormally displaced, thin extraocular muscle.

A unilateral right lateral rectus recession of 8 millimeters (mm) and right medial rectus resection of 7 mm corrected the large 50 prism diopter deviation. Although several studies still debate on the better surgical procedure for exotropias, a unilateral procedure on the poorer eye instead of a bilateral lateral rectus recession was preferred by the surgical team. Review of literature reported no pathologic findings as yet on the extraocular muscles.[3,4] Neither was there any reported literature on any ocular surgeries performed for patients with SRS. Despite the more anterior locations of the extraocular muscles, a regular strabismus surgery worked well in this case. After a successful and careful monocular recession and resection, the patient remained within monofixation range, or within a range of 8 prism diopters.

Limited information is available on the ophthalmologic presentation, treatment and management among patients with SRS. The diagnosis and management of SRS greatly rely on the clinical abilities of both the pediatrician and the ophthalmologist. Management of refractive errors and strabismus remain a challenge for the pediatric ophthalmologist in terms of surgical approach. Standard strabismus protocols for preoperative work-up and surgery do not differ from the otherwise normal child.

CONCLUSION

SRS is a rare disorder and the approach and outcome of surgical management for strabismus deviation remain

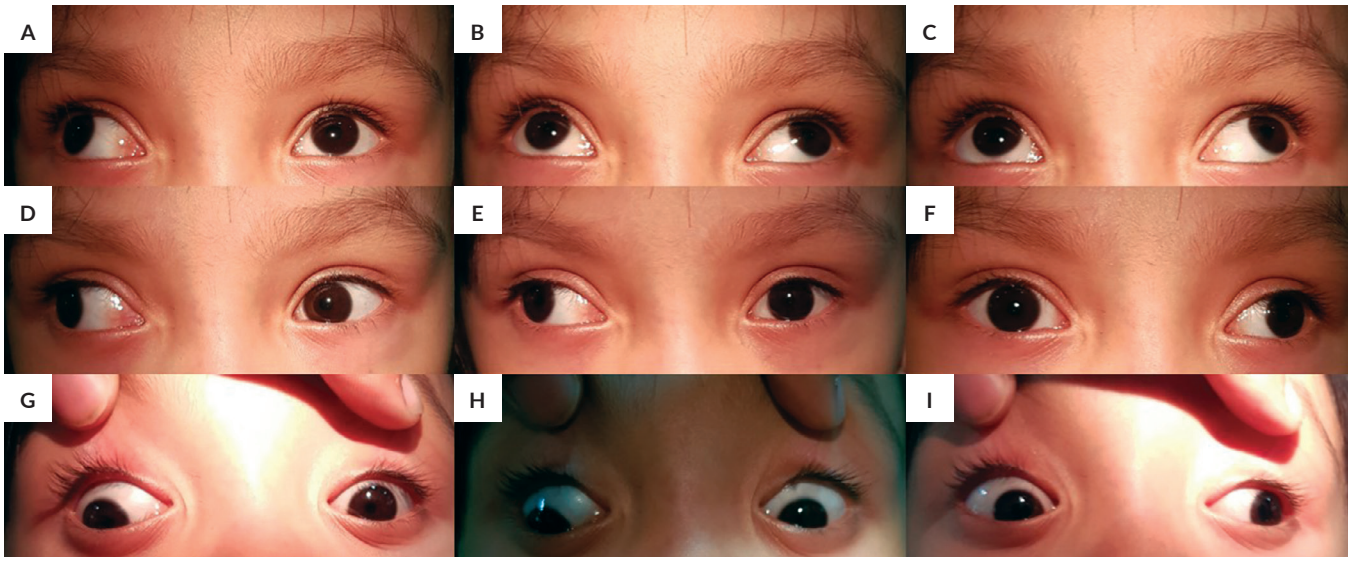

Figure 1. Preoperative nine gaze composite pictures showing a large constant right exotropia (XT) on primary gaze with the left eye fixating **(E)**. There was severe adduction limitation of the right eye **(F)** while moderate adduction limitation in the left eye **(D)**. The right inferior oblique **(A)** and the right superior oblique **(G)** showed pseudo-overaction. An X-pattern **(B, E and H)** was observed consistent with a tight lateral rectus on the right eye.

Figure 2. Postoperative composite pictures after 1 week showing an alignment of within 8 prism diopters of orthotropia. Adduction deficits were corrected post-operatively as seen in photos **(A)** and **(C)**. Myopia was not expected to progress in this case.

unknown. The surgical correction of strabismus in SRS can be challenging in several ways because of the differences in the orbit and locations of the muscles in different SRS patients, thus, the ophthalmologist and the surgeon should manage parent and patient expectations. This report presented a case of SRS who underwent strabismus surgery with excellent outcome with at least a year of post-operative follow-up. On the basis of our findings, it is recommended to perform a thorough pre-operative evaluation which includes comprehensive ophthalmologic evaluation and orbital imaging if necessary, such as in cases where a pre-operative imaging would help identify the status of the extraocular muscles.

Ethical Consideration

All authors secured consent from the parents of the patient for publication of the report.

Statement of Authorship

All authors contributed in the conceptualization of work, acquisition and analysis of data, drafting and revising and approved the final version submitted.

Author Disclosure

All authors declared no conflicts of interest relevant to the conduct of the proposed study. Consent from the patient and guardians were given.

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