

Review

The best of the best: a review of select pediatric ophthalmology and strabismus case reports published in 2015

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Abstract

This review highlights 4 case reports in the field of pediatric ophthalmology and strabismus published in the year 2015. The first article describes ocular presentations of acute childhood leukemia relapse. The second demonstrates the association of sector iris hemangioma with diffuse choroidal hemangioma. The third article provides a secondary procedure for persistent hypertropia in thyroid-related orbitopathy, and the fourth article describes a treatment for cyclic esotropia with temporary use of prismatic correction.

Introduction

Pediatric ophthalmology and strabismus is a wide and varied field, encompassing children and adults as well as intraocular, extraocular, and systemic pathology. The published literature reflects this diversity, and case reports are essential in sharing rare and novel developments. This review highlights several published cases in pediatric ophthalmology and strabismus in the year 2015, emphasizing reports that have the most potential to impact patient care.

A thorough, manual search of the ophthalmology literature for case reports in pediatric ophthalmology and strabismus as well as an automated search of MEDLINE and World of Science focused on pediatric case reports and series related to eyes and vision, yielded 102 articles, which were reviewed with respect to (1) potential for improving patient care, (2) uniqueness of clinical case or novelty of management technique, and (3) comprehensiveness in description of the clinical or surgical scenario.¹ Of the 102 publications, 4 were selected as outstanding according to all three criteria, as determined by the author. This review summarizes those articles, each focusing on a specific aspect of pediatric ophthalmology pathology: systemic disease, intraocular disease, and surgical and nonsurgical strabismus treatment.

Ophthalmic manifestations of relapsing acute childhood leukemia

In this case series, Chocron et al² present 3 patients with a history of acute lymphoblastic leukemia (ALL) with ocular recurrence of disease. The patients ranged in age from 9 to 15 years; 2 had T-cell ALL, and 1 had pre-B-cell ALL; recurrence occurred 2–13 months after completion of maintenance chemotherapy and 17 months after treatment for a prior remission. Two patients experienced eye redness and photophobia with either pain or epiphora and were found to have anterior uveitis, which did not completely respond to topical corticosteroids for 4 and 5 months. One patient was diagnosed via repeat anterior chamber paracentesis (as the first was without blast cells) and the other via lumbar puncture after bilateral optic disc edema was seen. The third patient presented with a red conjunctival lesion that extended into the orbit, and was found to be recurrent tumor on biopsy. All 3 patients eventually succumbed to their disease or complications thereof.

Chocron et al emphasize the importance of a high index of suspicion when evaluating ocular symptoms in patients with a history of ALL. The systemic work-up of these patients, performed in close collaboration with the oncologic and primary care teams, included repeat blood work, computed tomography or magnetic resonance imaging (MRI) of the brain and orbits, lumbar puncture

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for cerebrospinal fluid analysis, and bone marrow biopsy. Anterior chamber paracentesis was not performed in the first case because the patient's oncologist did not believe uveitis was an indication of recurrence. In the other uveitis case, the first paracentesis did not show blast cells, and when the patient developed a hypopyon 8 months later, repeat paracentesis showed leukemic blast cells. This accentuates the possibility of a false negative anterior chamber paracentesis, and the need for continued investigation if suspicion is high.

ALL is the most common childhood malignancy, with 90% 5-year survival rate.⁽³⁾ Orbital or ocular lesions are associated with increased frequency of central nervous system and bone marrow relapse, resulting in increased mortality.^{4,5} Ocular signs in patients with a history of ALL should be promptly and thoroughly evaluated for the possibility of recurrence to prevent delay in the diagnosis of relapse.

Sector iris hemangioma in association with diffuse choroidal hemangioma

In this report, Shields et al⁶ describe 2 patients with sector iris hemangiomas associated with choroidal hemangioma, each notably without any evidence of Sturge-Weber disease. The first patient was noted to have an iris lesion at 6 months of age, when he was diagnosed with amblyopia. It was diagnosed as an iris hemangioma at age 4, at which point, vision was counting fingers, thought to be due to noncompliance with patching. Further examination showed a diffuse choroidal hemangioma with overlying serous retinal detachment, and vision improved to 20/400 after plaque radiotherapy resolved subretinal fluid and reduced tumor thickness. The second patient was a 5-year-old boy with a stable, congenital iris hemangioma, and diffuse choroidal hemangioma not causing visual compromise. In addition to clinical examination, iris hemangiomas were characterized with ultrasound biomicroscopy and fluorescein angiography, and choroidal hemangiomas were further evaluated with optical coherence tomography, fluorescein angiography, and ultrasonography. Both patients had normal brain MRI and no cutaneous abnormalities.

This report highlights the association of iris hemangiomas with choroidal hemangiomas, an association evidently occurring in patients *without* Sturge-Weber syndrome. Iris vascular tumors are rare, comprising 2% of all iris tumors.⁽⁷⁾ Of 57 reported iris vascular tumors, none were associated with Sturge-Weber syndrome or choroidal hemangiomas.⁷ Conversely, there are no known reports of Sturge-Weber patients with iris hemangioma, although iris neovascularization can occur.⁸ The

authors propose that as in Sturge-Weber syndrome, in which a somatic mosaic mutation is thought to cause maldeveloped vascular tissue in a geographic distribution including the brain, eye and skin, the association of iris hemangioma and choroidal hemangioma results from an analogous mutation at a different point in development.⁶

Inferior oblique recession in thyroid related orbitopathy

In this case series, Salchow⁹ details 3 patients with thyroid-related orbitopathy and vertical strabismus, who, despite previous inferior rectus recession, continued to have manifest hypertropia. All 3 patients underwent 8, 10, or 12 mm recession of the inferior oblique in the hypertropic eye, based on grading system by Apt and Call.¹⁰ At 3-months' follow-up, the hypertropia in primary gaze was decreased from 9 ± 3 prism diopters (PD) to 1.3 ± 1.5 PD, and the largest hypertropia in side gaze from 18.3 ± 2.1 PD to 3.3 ± 1.5 PD. All patients were diplopia free in primary, side, and down gazes postoperatively. Excyclotorsion was reduced in most positions of gaze, although 1 patient had symptomatic incyclotorsion of 4° , measured by Harms tangent screen. Horizontal alignment had ≤ 5 PD of change in each patient. One patient was noted to have diplopia due to contralateral hypertropia, with onset 2 years after inferior oblique recession, presumably due to reactivation of thyroid-related orbitopathy, and was successfully treated with inferior rectus recession.

Thyroid-related orbitopathy and strabismus is a challenging disorder, in part due to its restrictive nature, which dictates the use of muscle recession, rather than resection. When more than one surgery is required, as is often the case,¹¹ the choices may become rapidly limited. Inferior rectus recession remains the first-line intervention for hypertropia in thyroid-related strabismus.¹¹ However, reoperation on a thyroid muscle may be increasingly technically difficult. This report shows that the inferior oblique, an elevator of the eye, may be weakened via recession with good success as a secondary procedure.

Nonsurgical treatment of cyclic esotropia

This case report presents a healthy 6-year-old girl with a history of anisometropic amblyopia in the right eye and small-angle esophoria, who returned for follow-up with complaints of horizontal diplopia.¹² She was found to have an esotropia of 40 PD at distance and near, with full ocular motility and otherwise normal ophthalmologic examination, neurologic examination, and MRI of

the brain. A careful history revealed that diplopia was present only every other day, and when examined on a nondiplopic day, the patient had 6 PD of esophoria at distance and 10 PD of esophoria at near, with 60 arcsec of stereopsis, similar to the initial examination. This 48-hour pattern persisted for 1 month. The authors prescribed a 6 PD base out Fresnel prism over the right eye, corresponding with the distance esophoria, which broke the cycle of esotropia. After 1 month of prism therapy, the prism was removed, and the patient had no recurrence of strabismus, continuing to 24 months of follow-up.

The traditional treatment of cyclic strabismus is strabismus surgery, dosed for the angle of misalignment when manifest, which is generally successful and without cyclic overcorrection in children,^{13,14} although cyclic esotropia has been reported to convert to cyclic exotropia in adults.¹⁵ Botulinum toxin injection has shown varied success.^{16,17} The authors propose that their method of prismatic correction of the latent deviation breaks the esotropic cycle by relieving the compensatory effort to maintain an esophoria, indicating that cyclic strabismus is due to a progressive loss of compensation.¹² The patient detailed above was already diagnosed with an esophoria, which may not be known in patients newly presenting with cyclic strabismus. Nevertheless, this nonsurgical treatment may be an excellent first-line method prior to pursuing more invasive management.

Summary

- Acute lymphoblastic leukemia relapse may present with anterior uveitis or conjunctival lesion.
- Sectoral iris hemangiomas can be associated with diffuse choroidal hemangiomas in children without Sturge-Weber syndrome.
- Inferior oblique recession may be an effective second-line procedure for persistent hypertropia in patients with thyroid-related orbitopathy.
- Cyclic esotropia may be definitively treated with prismatic correction of latent misalignment on nonstrabismic days.

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