Title: Amblyopia or Not: A Review of Amblyogenic Factors and Importance of Clinical Picture

Abstract: This case explores unilaterally decreased vision in a 6-year-old previously diagnosed with amblyopia but discovered to be macular pathology on second inspection. The importance of amblyogenic factors when evaluating decreased visual acuity is reviewed.

I. Case History

- Demographics: 6-year-old Asian male
- Chief complaint: Reduced vision in the left eye. Secondary complaint of left outward eye turn
- Ocular history: Intermittent Left Exotropia, Strabismic Amblyopia with Eccentric Fixation
- Medical history: none
- Medications: none
- Other salient information
  - Patient was seen for a comprehensive exam 2 months previous to this exam. Visual Acuity: OD 20/20, VA OS 20/32 (E-ETDRS protocol).
  - Equal, low hyperopic refractive error found with 1% cyclopentolate (+0.75DS OD/OS). Unsteady temporal eccentric fixation noted OS. CLXT at distance noted with 80% intermittent LXT noted at near. Ocular health within normal limits for anterior and posterior segment.
  - Diagnosed with mild strabismic amblyopia OS secondary to constant LXT at distance and high frequency intermittent LXT at near. Associated condition of unsteady 2pd temporal EF OS. Absence of disease that would cause reduced BCVA OS.
  - Started patching right eye for 2 hours a day to improve vision in left eye. Seen for a one-month follow-up where unsteady EF was noted again. No improvement in vision. Added fine eye-hand coordination to patching and recommended 1 month f/u with consideration to start active OVT for amblyopia treatment.

II. Pertinent findings

- Follow-up visit (2 months after comprehensive exam)
- Visual Acuity: 20/20, right eye. 20/32, left eye measured with E-ETDRS protocol
- Ocular Alignment: Constant left exotropia at distance (previous exam showed 200” RDS in the distance), intermittent left exotropia at near with 20% frequency (lower frequency than was noted at comprehensive exam)
- Monocular fixation
  - Visuscopy: central and stable OD. Unable to view foveal reflex OS
  - Haidinger’s Brushes: Reported central OD and OS
- Physical Exam (with 1% Tropicamide)
  - Anterior Segment: Healthy
  - Posterior Segment: Healthy OD. OS, mild macular elevation with no foveal reflex. Patch of presumed vitreo-macular traction superior/temporal macula.
  - Optos: OD normal and fully attached. OS defect noted in macula (faint outline of traction area with wrinkled appearance of macula in circular fashion).
  - OCT: OD normal tissue structure. OS: Vitreo-macular traction noted with contraction of internal limiting membrane.

III. Differential diagnosis

- Previous diagnosis: Strabismic Amblyopia
  - Ruled out with the presence of macular pathology and absence of constant, unilateral strabismus
- Primary Leading: Idiopathic Vitreo-macular Traction
- Secondary: Epiretinal Membrane
- X-Linked Retinoschisis
- Retinopathy of Prematurity
- Toxocariasis
- Familial Exudative Vitreoretinopathy

IV. Diagnosis and discussion

- Vitreoretinal adhesion is stronger in children than adults\textsuperscript{1} with both adults and children demonstrating a strong adhesion throughout the posterior pole, including the macula. This stronger adhesion accounts for a variety of vitreomacularopathies. Of particular concern in this case is idiopathic vitreo-macular traction and epiretinal membrane.
- Epiretinal membranes (ERM) are less common in pediatric patients than in adults and are much more likely to be fibrotic as opposed to “cellophane” in appearance.\textsuperscript{2,3} A premacular fibrosis type of ERM presents with a thickened membrane that has contracted on itself, becoming opaque. Superficial retinal folds and traction lines are also present.\textsuperscript{2}
- Other causes of vitreo-retinal complications in children include: X-Linked Retinoschisis, Retinopathy of Prematurity, Toxocariasis, and Familial Exudative Vitreoretinopathy. These diagnoses were excluded based on patient history and clinical presentation. The pathology in this case was limited to the macula of the left eye with healthy peripheral retina OS and completely healthy retina OD.
- The emphasis of this case is to highlight the importance of diagnosing amblyopia; it is not simply a diagnosis of exclusion. There must be an amblyogenic factor present to make a correct diagnosis of amblyopia.
Amblyogenic factors are listed and reviewed below. For amblyopia to be present there must also be an absence of ocular pathology or structural abnormality.

- **Unilateral Amblyopia**
  - Strabismic Amblyopia: Constant, unilateral strabismus that results in abnormal binocular input. Deviated eye’s image is suppressed by brain and vision does not develop normally.
  - Anisometropic Amblyopia: Unequal refractive error that causes abnormal binocular input. The brain receives a blurry image from the more ametropic eye while receiving a clear image from the fellow eye. The blurry image from the more ametropic eye is suppressed, preventing normal visual development.
  - Unilateral Form Deprivation: Secondary to media opacity or other mechanism obstructing the visual axis, such as a ptosis. The resulting inequality in images between the two eyes causes abnormal binocular input. The blurry and/or obscured image does not allow for normal visual development in the eye with the pathology. The pathology will also have a direct impact on visual acuity, but this secondary obstruction is what leads to amblyopia.

- **Bilateral Amblyopia**
  - Isoametropia: Bilateral high refractive error that doesn’t allow for clear retinal image quality in either eye. Bilateral reduction in best-corrected visual acuity that tends to be shallower than anisometropic amblyopia.
  - Bilateral Form Deprivation: Bilateral obstruction of visual axes that doesn’t allow for a clear retinal image to either eye. Bilateral reduction in best corrected visual acuity that tends to be shallower than unilateral deprivation.

- **Other causes of decreased vision where significant refractive error and ocular pathology are also absent.**
  - Malingering: Patient intentionally giving incorrect answers. Usually bilateral and in children often stems from a desire for glasses
  - Psychogenic: History of emotional trauma that results in a decrease of best-corrected visual acuity that the patient does not find bothersome. Also a bilateral phenomenon that is characterized by constricted, tubular fields.

**V. Treatment, management**

- **Vitreo-macular Traction**
  - Referral to retinal specialist for assessment and monitoring for stability
  - Observation. Preferred option for patients with VA 20/50 or better.  
  - Pediatric Surgical Options
    - ERM peel in conjunction with vitrectomy. Gives improvement in VA with success rates similar to adult outcomes though with higher recurrence rate than in adults.
- Intermittent Exotropia
  - Return to center to begin vision therapy
  - Good prognosis for decreasing/eliminating manifest deviation as patient shows good control of exotropia at near with good sensory fusion at distance and near. Therapy program will focus on anti-suppression when deviated and convergence training.

VI. Conclusion

In this case, it was very tempting to pinpoint the strabismus as the cause for decreased visual acuity, especially when ocular health initially appeared normal and the presence of EF was mimicked by subtle macular changes. Re-evaluation is indicated when clinical data does not add up. Less common pediatric pathologies, such as macular changes, should be kept in mind as a cause of decreased visual acuity.

VI. Bibliography


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