Abstract:
We present the second reported case of nasal ectopia with pseudostrabismus in retinopathy of prematurity. Simultaneous infrared/spectral domain optical coherence tomography imaging confirmed ectopia and demonstrated preservation of normal retinal layers and foveal contour.

I. Case History
   i. Patient demographics
      64 year old white female
   ii. Chief complaint
      Routine ocular hypertension and macular degeneration dilated exam
   iii. Ocular, medical history
      Ocular History:
      Diabetes Type II without retinopathy OU (6/10)
      Moderate risk dry age-related macular degeneration OU
      Pseudophakia OD (2006)
      Mild cataract OS
      Ocular hypertension OU (brimonidine TID OU)
      Congenital optic nerve anomaly OD
      Mild perifoveal cystic edema OD (ketorolac QID OD)
      Family Ocular History:
      Glaucoma - mother and maternal grandmother
      Medical History:
      Diabetes Type II diagnosed in 2002
      Pulmonary Hypertension
      Congestive Heart Failure
      Hyperlipidemia
      Sleep Apnea
      20 pack year history of smoking
      Gastro-esophageal reflux disorder
      History of carcinoid tumor of colon
      Medical Allergies: NKMA
   iv. Medications
      Albuterol
      Amlodipine besylate
      Atenolol
      Benazepril
      Brimonidine 0.2%
      Furosemide
      Insulin, glargine
      Ketorolac
      Metformin
      Mometasone
      AREDS formula multivitamin with lutein
      Pantoprazole
      Simvastatin
      Tramadol
   v. Other salient information
      Patient reports being prematurely born at approximately 7 months gestational age, weighing 4lbs. 4oz.
II. Pertinent findings

i. Clinical

Best corrected visual acuity OD: 20/30-2, OS: 20/20
Cover Test: Distance: 4 prism diopters (pd) BU OD vertical heterophoria
(with Rx) Near: 4 pd BU OD vertical heterophoria
6pd exophoria
Hirschberg Reflex: 25pd constant right eso deviation
Ocular Motilities: full ductions OD, OS
Amsler Grid: OD: central metamorphopsia
OS: normal

ii. Physical

Pupils: direct and consensual reactions OU, 1+ right afferent pupillary defect
Slit lamp Exam: 2+ diffuse corneal endothelial pigment dusting OU
Eyelids, Conjunctiva, Anterior Chamber, Iris: within normal limits OU

Tonometry:
OD: 22
OS: 18

Dilated Fundus Examination:
OD Findings:
• posterior chamber intraocular lens in good position with large capsular rent
• clear vitreous
• no detectable cupping of nerve, tractional elevation from pre-retinal fibrotic tissue, nerve appears sectorally hypoplastic temporally with double ring sign and no pallor
• dense peripapillary pre-retinal fibrotic membrane extending to equator nasally causing traction and retinal folding
• numerous medium and large sized mixed drusen with no hemorrhage or exudation located in central macular region, pre-retinal fibrotic tissue overlying macula
• senile reticular and pavingstone degeneration

OS Findings:
• 2+ Nuclear Sclerosis
• Posterior vitreal detachment
• 0.35/0.35 CD ratio
• Posterior pole and peripheral retina normal

iii. Heidelberg Spectralis Spectral Domain Optical Coherence Tomographer (OCT)

OD: Dense peripapillary pre-retinal membrane with extension nasally. Retinal folds in posterior pole and nasal peripapillary region; tractional elevation of the optic nerve; focal area of vitreoretinal traction nasal to disc, numerous medium and large sized drusen centrally, ectopic macula displaced nasally and inferiorly. Foveal contour relatively normal; few small perifoveal cystic spaces.

OS: Numerous medium and large sized drusen
III. Differential diagnosis

Pseudoesotropia secondary to nasal macular ectopia
Acquired esotropia with/without anomalous correspondence
Congenital esotropia with/without anomalous correspondence

IV. Diagnosis and discussion

Patient presented with appearance of constant esotropia while cover test revealed an exophoric movement. Additionally the patient exhibited no suppression, no diplopia, and no restriction of ductions. Strabismus with anomalous retinal correspondence was suspected, however the 20/40 acuity conflicted with the amount of strabismus present. A retinal scan of the OCT was performed showing retinal dragging with foveal displacement inferonasally. Simultaneous infrared and spectral domain OCT imaging confirmed ectopic foveal fixation by demonstrating steady fixation of the internal target and observation of normal retinal layers with foveal contour. Foveal location nasally is consistent with an esotropic eye turn in order for the patient to fixate. This causes an appearance of strabismus despite bifoveal fixation, resulting in pseudostrabismus.

Heterotopia of the macula is an uncommon consequence of retinopathy of prematurity (ROP). Typically in macular heterotopia due to ROP, the macula is displaced temporally due to cicatrical forces from vascular proliferation toward the temporal retina. Developing retinal vasculature in the fetal eye spreads from the optic nerve in a radial fashion until it reaches the ora serrata. Due to the nasal displacement of the optic nerve, the primitive vasculature reaches the temporal retina last; being the last to mature it is the most susceptible to hypoxic injury. Our patient had the unusual case of a nasal heterotopia which has only been reported in 11 patients with regressed ROP.

Pseudostrabismus results from foveal ectopia; the patient appears to have a strabismic eye when they are actually foveating with both eyes. It is a rare condition, necessitating intact vision with an ectopic fovea from an early age allowing for development of fusion and/or adaptation. As most foveal ectopia occur temporally, the eye correspondingly develops an exotropic appearance. There are several instances of exotropia and esotropia associated with ROP in presence of foveal ectopia, however it is rare to develop pseudostrabismus, having only been reported in 10 patients. We report only the second case of pseudostrabismus associated with a nasal macular ectopia in ROP.

And finally, the extramacular drusen in the right eye of this patient with otherwise classic age-related macular degeneration (AMD) call into question some of the existing theories of AMD pathogenesis; this will be the subject of further investigation.

i. Bibliography, literature review encouraged

V. Conclusion

i. Clinical pearls

1. OCT imaging can be used to determine heterotopia by locating foveal structure and by observing the patient’s fixation of the internal target

2. It is important to evaluate the foveal location in patients with a history of premature birth before referring for strabismus surgery or vision therapy. In cases of pseudostrabismus, the dysfunction is primarily aesthetic and there is no form of intervention.