Retinoschisis in Coats Disease: clinical picture, therapeutic considerations and management outcomes

Audina M. Berrocal MD
Professor of Clinical Ophthalmology
Medical Director of Pediatric Retina and ROP,
Vitreoretinal Fellowship Director
Bascom Palmer Eye Institute
University of Miami, Miller School of Medicine
No relevant disclosures for this presentation

ALCON, DORC, VISUNEX, ZEISS, PHOENIX, ALLERGAN
Aerie, ProQR, AGTC
Coats Disease: Background

- Congenital, Non-hereditary
- Unilateral
- Males > Females
- Presenting Symptom:
  - Decreased VA (43%)
  - Strabismus (37%)
  - Leukocoria (20%)

Coats Disease: Diagnosis/Monitoring

• Clinical Diagnosis: angiography and ultrasound

• Long-Term Multimodality Imaging
  • Only 41% of patient present with correct diagnosis
  • 27% referred for Retinoblastoma
    • Shields et al. AJO 2001
Coats Disease: Globe salvage

Globe Salvation rate 94%

Visual Acuity 20/20-20/50 in 50% of patients
Coats Disease: Current advance treatment

August 2002 – January 2014 (26 eyes)

Primary Outcome: Anatomic Success, Globe Salvation, Final Visual Acuity

<table>
<thead>
<tr>
<th>Median Laser</th>
<th>5 sessions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean IVB</td>
<td>3 injections</td>
</tr>
<tr>
<td>Globe Salvage</td>
<td>100%</td>
</tr>
</tbody>
</table>
Coats Disease: Macrocyst/schisis

Clinical Variations and Complications of Coats Disease in 150 Cases: The 2000 Sanford Gifford Memorial Lecture

JERRY A. SHIELDS, MD, CAROL L. SHIELDS, MD, SONTASH G. HONAVAR, MD AND HAKAN DEBCIRCI,

<table>
<thead>
<tr>
<th>Posterior Segment Finding</th>
<th>Number (%)*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Telangiectasia</td>
<td>158 (100%)</td>
</tr>
<tr>
<td>Intraretinal exudation</td>
<td>157 (99%)</td>
</tr>
<tr>
<td>Exudative retinal detachment</td>
<td>128 (81%)</td>
</tr>
<tr>
<td>None</td>
<td>30 (19%)</td>
</tr>
<tr>
<td>Partial</td>
<td>54 (34%)</td>
</tr>
<tr>
<td>Total</td>
<td>74 (47%)</td>
</tr>
<tr>
<td>Retinal hemorrhage</td>
<td>20 (10%)</td>
</tr>
<tr>
<td>Retinal macrocyst</td>
<td>18 (11%)</td>
</tr>
<tr>
<td>Vasoproliferative tumor</td>
<td>9 (6%)</td>
</tr>
<tr>
<td>Optic disk neovascularization</td>
<td>3 (2%)</td>
</tr>
<tr>
<td>Retinal neovascularization</td>
<td>1 (1%)</td>
</tr>
<tr>
<td>Vitreous hemorrhage</td>
<td>0 (0%)</td>
</tr>
</tbody>
</table>

Coats: Pathology

- Splitting of layers in the neurosensory retina
- Tractional
- Exudative
Acquired Schisis: Coats Disease

- **Tractional:** Tractional forces combined with vascular compromise leading to complex schisis
- **Exudative:** Leakage of microaneurysm into the retinal layers leading to schisis
- **Ischemic**
Coats Disease: Retinoschisis

Cavity collapsed with treatment 14 months later:
7 IVFA guided laser treatments
7 IVB
1 STK

In globe salvage early PPV/EL/IVB/STK controls disease
Methods

- Retrospective review with IRB approval
  - January 2014 to May 2018

- Identified:
  - 138 subjects with Coats
  - 133 had color images

- 18 subjects with retinoschisis (14%)
Methods

• Included:
  • Patients with more than 5 month follow up
  • Color images

• Two groups:
  • Schisis group
  • Control group
Results:

• Gender:
  • 75% male and 25% women

• Median age in the control group:
  • 17.8 years of age +/-15

• Median age in the schisis group:
  • 6.5 +/- 5
Results: Complete schisis control

<table>
<thead>
<tr>
<th></th>
<th>Schisis</th>
<th>Control</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diode Laser Photocoagulation Sessions</td>
<td>4.8± 2.9</td>
<td>n/a</td>
</tr>
<tr>
<td>Intravitreal Anti-VEGF</td>
<td>4.5 ± 2.9</td>
<td>n/a</td>
</tr>
<tr>
<td>Sub-Tenon’s Injections Triamcinolone</td>
<td>1.70 ± 1.20</td>
<td>n/a</td>
</tr>
<tr>
<td>Intraocular Surgery (PPV or SB)</td>
<td>39% (P=0.14)</td>
<td>22 %</td>
</tr>
</tbody>
</table>
Discussion:

- Retinoschisis is a rare entity in Coats Disease

- The pathophysiology of retinoschisis in Coats Disease is not completely understood could be exudative or tractional (combined)

- Angiography guided laser and multimodal imaging are necessary to monitor progression of disease

- Early PPV may be indicated in the setting of rapidly progressive schisis
Thank you!

- Linda Cernichiaro-Espinosa
- Nathan Scott
- Timothy Murray
- William Harbour
- Sander Dubovy
- Catherine Negron