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Shaken Baby Syndrome (SBS) Ocular Findings with Legal Implications

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Important Prologue

- Injury (disruption of form or function) is influenced by a large number of variables
- No two episodes of injury are exactly alike
- Non-Accidental injuries: Injuries are due to trauma intentionally inflicted by another person
- The diagnosis of non-accidental injury to a child requires input from both clinicians and pathologists
Background Information (SBS)

• Definition
  • Caffey (1946) Whiplash-shaken infant syndrome
  • Lancon (1998) Child abuse
  • Chase (1999) Abusive head injuries
  • Case (2001) Shaken Baby Syndrome
  • Keenan (2002) Inflicted childhood neuro trauma
The purpose of this talk

- Assist pathologists in identifying and reporting ocular findings in cases of suspected non-accidental injury
  - Hemorrhage
  - Retina
  - Sclera
  - Optic nerve
  - Tissue displacement
  - Vitreous
  - Retina
  - Optic nerve
Background Information (SBS) (Continued)

• **Pathophysiology**
  - Not clearly understood
    - Meta studies in progress
    - New studies planned
  - Transfer of kinetic energy
    - Shaking
    - Blunt force trauma
  - The eye-wall (cornea + sclera) is intact
  - Ocular
    - Hemorrhage
    - Tissue displacement
The Result of Pathophysiologic Mechanisms

• Hemorrhage
  • Retina, location (subhyaloid, superficial, full thickness, subretinal)
  • Retina, extent (focal posterior pole, focal peripapillary, confluent, extension to the ora serrata)
• Intrascleral (Circle of Zinn-Haller)
• Optic nerve (subdural, subarachnoid)
The Result of Pathophysiologic Mechanisms (continued)

- Tissue displacement
  - Schisis of the retina (splitting)
  - Retinal detachment (separation from the retinal pigment epithelium)
  - Traction of the dural insertion in the sclera
“Challenges” to the Surgical Pathologist

• Interpreting ophthalmologist’s notations
• Critical anatomy of the eye
• Autolysis and artifacts of the infant eye
• Acquiring the specimen
• Gross Dissection
• Macroscopic Characteristics
• Microscopic characteristics
• The diagnosis
• The Comment
Data from Clinicians
Indirect Ophthalmoscopy

20D Lens
Virtual, inverted, reversed image
• When in doubt; call the ophthalmologist
Optic Disc

"Macula"

"The Posterior Pole"

Superior Temporal Arcade

Ora Serrata

Fovea Centralis

Inferior Temporal Arcade
Interpreting the ophthalmologist’s notations

- **VA** = visual acuity
  - **FF** = fix and follow an object
  - **CF** = count fingers
  - **Pictures** = images of common objects
- **EXT** = external examination
- **SLT** = slit lamp examination
- **PLE** = pen light examination
- **Indirect ophthalmoscopy** = basic tool
Indirect Ophthalmoscopy (IO)

- Most common bedside evaluation
- Viewing an aerial virtual image
- Requires dilation of pupil (mydriacyl, neosynephrine)
- SD: Scleral depression: external pressure to bring the peripheral retina into view
- Clear media = retina clearly visible
- VH = vitreous hemorrhage
- Retinal hemorrhage
Data From Pathologists
Retinal Hemorrhage

• NFL = nerve fiber layer

• Posterior pole: posterior hemisphere of the eye

• Peripapillary: around the optic nerve

• Macular: between the superior and inferior temporal arcade of vessels

• Peripheral: outside of the macula

• To ora: to the most peripheral retina
Essential Anatomy of the Eye
P.O. = Pupil-
Optic Nerve Section
Calottes = caps

Peripheral artifact  Lange’s fold

Intraretinal Hemorrhage
Morphology unique to children

- Immature anterior chamber angle
- Flat contour of the lens
- Posterior umbilication of the lens (fixation)
- Delicate, elongated ciliary processes
- Folding of the peripheral retina (Lange’s fold) (fixation)
Epithelial irregularity = post mortem
drying artifact

Immature anterior chamber angle

“Flattened” profile of the crystalline lens
The Immature Eye

Primary Vitreous

Posterior umbilication of the lens;
Fixation artifact

Tunica Vasculosa Lentis

Persistent Hyaloid Vessels
• Under age 3 years
• Identifies the junction of the retina and the ciliary body = Ora Serrata
Retina

- Transparent, cellular
- No extracellular space (necessary for transparency)
- Poorly supported architecture (Mueller cells)
- Regions
- Retinal blood supply
Basic “Systems” of the Retina

Transmission

Computation

Reception

No extracellular space --- Hemorrhage = destruction
Vascular System of the Retina

- Defined vascular channels inner half
- Parallels route of nerve fiber layer
- Dominant superior, inferior temporal arcades
- Avascular periphery
- Capillary-free zone
Blood Supply of the Retina

• Central retinal artery; end artery, no collateral vessels

• Located in the superficial retina, anterior 50% (no vessels in the external retina)

• The central macula is avascular (capillary-free zone)

• Separated from vitreous by the internal limiting membrane

• Density of circulation is proportional to the thickness of the retina
  • Ora = thin
  • Peripapillary = thick
**Periphery of the Retina**

- **Avascular**
- **Few functional photoreceptors**
- **Nutrition from the choriocapillaris**
- **Microcystoid spaces**
Regions of the Retina

- **Ora serrata**: termination of the peripheral retina
- **Macula (foveola centralis)**; temporal to the optic disc, most sensitive portion of the retina (best visual acuity)
- **Peripapillary**: region around the optic disc, thickest portion of the retina
Vitreous

- Transparent, acellular
- 80% volume of the eye
  - 4 cc
  - 4 grams
- Extracellular materials
  - Type II collagen
  - Hyaluronic acid (proteoglycan)
  - Water
Vitreous

- A scaffolding structure
- Function complete at age 3 years
- Shrinks throughout live
- Origin of “floaters”
Regions of the Vitreous

- Base
- Weiggert’s ligament
- Potential space of Berger
- Adhesions
  - Optic cup
  - Parafovea
  - Blood vessels
  - Lattice degeneration
Vitreous (continued)

- Degenerates after age 3 (floaters)
- Strong attachment: vitreous base at ora
- Relative attachments
  - Peripapillary retina (optic disc)
  - Perimacular retina (near fovea)
  - Along the course of retinal blood vessels
Optic Nerve

- **Meninges**
  - **Dura**: inserts into sclera
  - **Arachnoid**: contiguous with the CNS
  - **Pia**: distributes blood to the optic nerve axons

- **Optic disc blood supply**: Circle of Zinn-Haller

- **Optic nerve blood supply**: arachnoid

- **Retina blood supply**: central retinal artery
Central Retinal Artery and Vein
Short posterior ciliary arteries

- Branches of the ophthalmic artery
- Supply tissues of the optic disc
- End arteries
Cross Section of the Optic Nerve
12 mm posterior to the Sclera
Dura

Arachnoid

Pial septa defining Axonal columns

Route of the Blood Supply
Orbit

- Orbital bones: usually intact
- Orbital soft tissues: usually no hemorrhage or disruption
- Preseptal soft tissues (eyelids) usually no hemorrhage or disruption
Autolysis and Artifacts

- Mechanical disruption or displacement of the lens during sectioning
- Detachment of the retina
- Degeneration of the photoreceptor outer segments
- Mechanical disruption of the neurosensory retina
- Incomplete or off-axial sections
  - Pupil
  - Optic disc
Autolytic Separation of Vireo-retinal Junction

Autolytic Degeneration of Photoreceptor Outer Segments
Retinal Artifacts

- Detachment
  - No subretinal material
  - Amputation of RPE microvilli
  - Sharp contours of retinal separation
Acquiring the Specimen

  - Anterior – more direct
  - Trans frontal – more control (adequate optic nerve)

- **Do not open the eye or penetrate the sclera before immersion fixation**
  - All important diagnostic information is within 2 mm of the external surface of the eye
  - The sclera is not a barrier to the diffusion of formalin
  - Opening the eye before fixation may result in loss of diagnostic information
Gross Dissection

• Protocols
  • Traditional anterior-posterior
    • Pupil
    • Optic nerve
    • Horizontal includes macula
  • Equatorial section
    • Simulates clinical findings
    • Requires secondary sectioning

• Obtain a cross section of the optic nerve

• Photograph positive and negative findings
Macroscopic Characteristics

- Few if any external signs of trauma
- Note “color” of sclera
  - Important finding in osteogenesis imperfecta
  - Indicates relative thickness of sclera
  - Sclera may be thin in premature infants
“Blue”

“White”

Osteogenesis Imperfecta
Macroscopic (continued)

- Retina
  - Retinal hemorrhage
    - Location
    - Extent
      - Peripapillary
      - Confluent
      - Extension to the ora
  - Tissue disturbance
    - Elevation of the macula
    - Retinal detachment
Peripapillary hemorrhage

Ora Serrata

Macular Traction Detachment

Photograph for Files
Macroscopic (continued)

- **Optic nerve**
  - Normal cross sectional diameter of myelinated nerve = 3 mm
  - Preservation of neural axis
  - Subdural hemorrhage
  - Subarachnoid hemorrhage

- **Optic nerve atrophy**
  - Seen with prolonged survival
  - Diminished axonal column
  - Expansion of subdural space
Microscopic Characteristics

- Few if any signs of disruption of the anterior eye
- Cataract, very infrequent
- Retinal hemorrhage
  - Within the retina (intraretinal)
  - Under the retina (subretinal)
  - Internal to the retina (subhyaloid)
- Displacement of the internal limiting membrane
- True retinal detachment
Retina Hemorrhage
Area of macular traction, disruption and hemorrhage
Thick ganglion cell layer
Subretinal hemorrhage = true retinal detachment
Circle of Zinn-Haller

Optic nerve
Axona

Hemorrhage

Sclera
Subarachnoid & Subdural Hemorrhage
The Role of Iron Stains

- The accuracy of intraretinal hemosiderin as an indication of previous injury
- Hemosiderin detected within 2 days in intraretinal hemorrhage caused by central retinal vein occlusion
- Hemosiderin may be associated with organization of extravasated blood
- Absence of hemosiderin does not exclude prior retinal hemorrhage
The Diagnosis

- Eye, right, autopsy specimen: extensive intraretinal and subretinal hemorrhage extending to the ora, focal detachment of the macula, hemorrhage present in the region of Zinn-Haller, extensive subdural and subarachnoid optic nerve hemorrhage (please see comment)

- Eye, left, autopsy specimen: focal intraretinal hemorrhage posterior pole (please see comment)
The Comment

• The findings of the right and left eyes are consistent with non-accidental injury, however, this report should be considered in the overall context of all available historical information, clinical findings, and the remainder of the autopsy findings.

• Other comments
Summary Points (SBS)

1. Input from both clinicians and pathologists is necessary
2. Findings: Hemorrhage, tissue displacement of ocular tissues
3. Report location and extent
4. Obtain a long section of optic nerve
5. Eyes should be fixed before sectioning
6. Hemosiderin staining correlates poorly with time intervals
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