H173 Retinal (RH) and Optic Nerve Sheath Hemorrhage (ONSH), Papilledema, and Spinal Cord Nerve Root/Ganglia Hemorrhage Associated With a Cerebral Cavernous Malformation

Dongfang Yu, MD, PhD, Department of Pathology, Winston Salem, NC 27157; Ryan T. Mott, MD, Medical Center Boulevard, Winston Salem, NC 27157-1072; William J. Beuerlein, DO, Wake Forest Baptist Health, Winston Salem, NC 27157; Patrick E. Lantz, MD, Wake Forest University School of Medicine, Winston-Salem, NC 27157-1072*

Learning Overview: After attending this presentation, attendees will learn that extensive RHs, ONSH, and papilledema can occur in young children who have increased intracranial pressure, and spinal cord nerve root/ganglia hemorrhages are not specific for hyperflexion-hyperextension injury in young children.

Impact on the Forensic Science Community: This presentation will impact the forensic science community by reinforcing the necessity for routine postmortem ocular and spinal cord examinations of children with fatal intracranial hemorrhage, cerebral edema, and elevated intracranial pressure.

RH and ONSH in young children have been considered specific for inflicted traumatic brain injury. Current studies suggest that non-traumatic, markedly elevated Intracranial Pressure (ICP) rarely causes RHs, and when it does, the RHs are few in number and peripapillary. Dorsal root ganglial hemorrhage has been attributed to hyperflexion-hyperextension neck injury (shaken baby syndrome) in infants and young children. A case of a young child with papilledema, bilateral RHs, ONSHs, and spinal nerve root/ganglia hemorrhages associated with non-traumatic intracranial hemorrhage arising from a ruptured left temporo-parietal cavernous malformation is presented. The clinical, autopsy, and investigative findings of this case refute the purported specificity of RHs, ONSHs, and spinal dorsal root ganglial hemorrhages in young children.

Case Report: A 5-year-old boy with no significant medical history was found unresponsive at home. At the local hospital, cranial Computed Tomography (CT) revealed a large intraparenchymal hemorrhage. The previous day, he had been diagnosed with Streptococcal pharyngitis and sent home with antibiotics following complaints of fever and headache. On arrival at the tertiary medical center's Pediatric Intensive Care Unit, he had dilated pupils and a Glasgow coma scale of 6. A repeat cranial CT revealed extensive intraparenchymal hemorrhage centered in the left temporal lobe measuring to 6.1cm, extensive surrounding edema and mass effect resulting in 11mm rightward midline shift and crowding of suprasellar cistern concerning for uncal herniation, multifocal subarachnoid hemorrhage along the left convexity, and intraventricular extension of hemorrhage into lateral ventricles with developing hydrocephalus. His family offered no reports of recent trauma. His initial ICP was 67mmHg and remained elevated (40–50mmHg) despite external ventricular drain placement and mannitol treatment. Due to the extent of his neurological injury and high ICP that was refractory to treatment, his parents decided to withdraw care; he was pronounced dead 26 hours after admission. No clinical fundal examination was recorded in the medical record.

Postmortem indirect ophthalmoscopy revealed bilateral papilledema (left > right), numerous splinter, flame-shaped, and dot retinal hemorrhages over the posterior poles and extending past the equators. His autopsy was significant for a 7 x 5 x 4cm intraparenchymal hemorrhage of the left temporo-parietal cortex with associated cerebral edema, left uncal herniation, and cerebellar tonsillar herniation. The intraparenchymal hemorrhage was from a ruptured, thrombosed, necrotic cavernous malformation. Diffuse spinal cord nerve root and ganglial hemorrhages involved the cervical, thoracic, and lumbar regions. The RHs were multilayered and the bilateral ONSH was intradural, subdural, and subarachnoid.

This case describes severe hemorrhagic retinopathy, papilledema, ONSH, and diffuse spinal cord nerve root/ganglia hemorrhages associated with a fatal spontaneous, non-traumatic, intracranial hemorrhage arising from a ruptured cavernous malformation of the left temporo-parietal lobe. The reported ocular and spinal cord findings have been considered diagnostically specific for abusive head trauma (shaken baby syndrome); however, this case illustrates the need for caution in attributing these findings as diagnostically specific for abusive head trauma. This case emphasizes the importance of consistent and thorough postmortem ocular and spinal cord examination in young children with intracranial hemorrhage, cerebral edema, and elevated intracranial pressure.

Retinal Hemorrhages, Papilledema, Increased Intracranial Pressure

*Presenting Author