G129 Spontaneous Non-Traumatic Subarachnoid Hemorrhage With Retinal and Optic Nerve Sheath Hemorrhages Associated With Segmental Cerebrovascular Fibromuscular Dysplasia

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After attending this presentation, attendees will learn that retinal hemorrhages and optic nerve sheath hemorrhages can occur in young children who have a spontaneous non-traumatic subarachnoid hemorrhage.

This presentation will impact the forensic science community by emphasizing the need for routine postmortem ocular examinations in young children dying suddenly and unexpectedly.

Spontaneous non-traumatic subarachnoid hemorrhage (SAH) in children is usually attributed to arteriovenous malformations, aneurysms, neoplasm, infection, leukemia, hemophilia, or sickle cell disease. Aneurysms may arise with conditions such as polycystic kidney disease, Ehlers-Danlos syndrome, fibromuscular dysplasia (FMD) and other conditions. A case of a child with bilateral retinal hemorrhages (RHs) and optic nerve sheath hemorrhages (ONSHs) associated with a spontaneous SAH arising from a ruptured right vertebral artery aneurysm due to segmental FMD is reported.

A previously healthy 4-year-old boy had a witnessed collapse at his daycare facility. Paramedics found the child in asystole at the scene where he was intubated and received cardiopulmonary resuscitation along with epinephrine and atropine. By the time he reached the medical center he was in sinus tachycardia and had no neurological response. His cranial cranial computed tomography revealed diffuse subarachnoid hemorrhage with marked cerebral edema causing cisternal effacement and downward displacement of the cerebellar tonsils. No parenchymal lesion was visible to account for the hemorrhage. Due to the grim prognosis, organ procurement was contacted and viable organs were recovered for transplantation after declaration of clinical brain death.

No clinical fundal examination was recorded in the medical record; however, postmortem monocular indirect ophthalmoscopy revealed bilateral RHs over the posterior poles. His autopsy revealed diffuse cerebral and spinal cord subarachnoid hemorrhage due to a ruptured right vertebral artery aneurysm arising within segmental fibromuscular dysplasia. Additionally, there was intraventricular hemorrhage, cerebral edema with cerebellar tonsillar herniation and hypoxic ischemic brain injury, pituitary necrosis and patchy acute spinal cord infarcts. He had bilateral optic nerve sheath hemorrhages, most pronounced in the optic nerve-globe junction. The bilateral ONSHs were located not just in the subarachnoid space but were also subdural and intra-dural with extension in the adjacent periorbital soft tissue. Blood extended along the pial septa into the interior of the left optic nerve. The left globe contained multiple RHs involving the nerve fiber layer with focal pre-retinal extension. No FMD was in the globes or orbital soft tissues.

Vitreous bleeding associated with SAH was first described by Moritz Litten in 1881; however, the association of vitreous hemorrhage concurrent with intracranial bleeding has been attributed to Albert Terson and later broadened by some authors to include any intra-ocular hemorrhage. It was initially believed that blood from a SAH could track along the optic nerve sheath and penetrate the lamina cribosa to appear in the vitreous space, but this theory has been refuted because a connection has not been demonstrated on electron microscopy. Another theory suggests that rapid increase in intracranial pressure transmitted through the optic nerve sheath and impairs venous drainage to the cavernous sinus, resulting in venous congestion and rupture of retinal vessels.

A noninflammatory, nonatherosclerotic disorder, FMD is characterized by abnormal cell growth in the walls of medium and large arteries leading to stenosis. The etiology is not clear but believed to relate to hormonal and mechanical factors. It is familial in about 10% of cases and affects the renal arteries about 60-75% of the time. The extra-cranial cerebrovascular arteries are involved in 30-60% of cases.

Extra-renal FMD can cause critical stenosis, aneurysm formation and rupture or cerebral thromboembolism.

Many authors consider that RHs in young children are uncommon in the absence of abusive head trauma. This case highlights the lack of clinical fundal examinations in young children when abuse is not suspected and the importance of routine postmortem ocular examinations in young children who die suddenly and unexpectedly. Although the brain and spinal cord had only subarachnoid hemorrhage, the presence of subdural hemorrhage in the optic nerve sheaths suggests that a sudden rise in intra-cranial pressure preceded the ONSHs and RHs.

Forensic Science, Retinal Hemorrhage, Non-Traumatic Subarachnoid Hemorrhage

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