Ocular Findings in Raised Intracranial Pressure

A Case of Terson Syndrome in a 7-Month-Old Infant

Othon J. Mena, MD, Ian Paul, MD, and R. Ross Reichard, MD

Abstract: We present the case of a 7-month-old female infant who was found crying and limp. She was transported to a hospital where a possible subarachnoid hemorrhage was diagnosed radiologically. Before further studies could be pursued, her condition worsened and she died. The autopsy demonstrated diffuse subarachnoid hemorrhage of the brain and along the spinal cord. The brain, spinal cord, and eyes were retained and examined postfixation. An aneurysm of the middle cerebral artery was identified. Examination of the eyes demonstrated bilateral optic nerve sheath hemorrhage and extensive retinal hemorrhages extending to the ora serrata. A rapid increase in intracranial pressure secondary to subarachnoid hemorrhage following rupture of an aneurysm can result in sequelae similar to those found in inflicted traumatic brain injury. In this case, the rise in intracranial pressure resulted in marked hemorrhage within the optic nerve sheaths as well as intra- and preretinal hemorrhages. Patients with subarachnoid hemorrhage, or other causes of rapidly increased intracranial pressure, may develop ocular hemorrhage (Terson syndrome). This case illustrates the importance of ruling out natural disease before attributing the autopsy findings to trauma, as well as the importance of postmortem fixation of pediatric brains and eyes prior to examination.

Key Words: Terson syndrome, intracranial pressure, subarachnoid hemorrhage, aneurysm

(*Am J Forensic Med Pathol* 2010;XX: 000–000)

A 7 month-old female infant with a history of only atopic dermatitis was found crying and limp following a usual nap. Paramedics were summoned, and after resuscitative efforts, she was transported to a local hospital. Radiologic evaluation showed a possible subarachnoid hemorrhage. However, before further studies could be pursued, her condition deteriorated, and she died approximately 18 hours after arrival to the hospital.

The external examination showed a well-nourished and hydrated infant consistent with her age. The dermatitis was most pronounced at the extremities and scalp, and consisted of drying and flakiness of the skin. Full body radiographs showed no evidence of skeletal deformity or acute and subacute injury. Internal examination of the torso showed no congenital defect, evidence of trauma, infectious disease, or other abnormality.

Intracranial examination showed diffuse cerebral edema and subarachnoid hemorrhage, which was most pronounced on the base of the brain. Prior to removal of the brain, a postmortem vertebral artery angiogram was performed. The angiogram did not reveal any abnormality of the vertebral arteries. The optic nerve sheaths were markedly hemorrhagic. Anterior opening of the spinal canal showed diffuse extension of the subarachnoid hemorrhage along the entire length of the spinal cord. No other gross abnormality was identified. Law enforcement contacted the office to determine if any criminal charges should be pursued. Due to no obvious signs of trauma or foul play, it was recommended that no criminal investigation should be pursued pending further examination.

The brain, spinal cord, and eyes were retained for formalin fixation and further examination at a later time. Approximately 2 weeks after the initial examination, the retained organs were again examined. At the level of the left lateral sulcus, and arising from distribution of the middle cerebral artery, a 2.1-cm grossly vascular lesion was identified (Fig. 1). No other lesion was grossly seen. Histologically, the lesion consisted of a tortuous vessel with variable thickening and thinning of the wall, consistent with an aneurysm. There was a marked amount of red blood cells both within and outside the vessel walls. A focus of attenuation and interruption of the vessel wall, consistent with a site of rupture, was identified. The vessel wall had degenerative changes consisting of myxoid changes within it and irregular medial hypertrophy with scattered foci of calcification. An elastin stain showed nonuniformity and irregular absence of the internal elastic lamina.

Examination of the spinal cord confirmed the presence of extensive subarachnoid hemorrhage (Fig. 2). The eye examination demonstrated multiple retinal hemorrhages extending to the ora serrata (Fig. 3). Microscopically, the hemorrhages were both preretinal (subhyaloid) and intraretinal. The retinal hemorrhage involved primarily the optic nerve fibers, and focally extended to the inner nuclear layer, without extension into the outer layers. The optic nerve sheath had extensive intradural and subdural hemorrhage, and mild to moderate subarachnoid hemorrhage (Fig. 4).

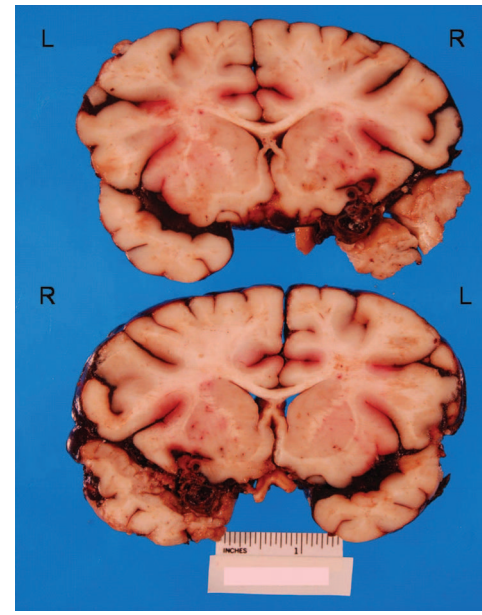

FIGURE 1. Coronal sections of brain with 2.1 cm aneurysm in distribution of right middle cerebral artery.

Manuscript received September 10, 2008; accepted November 13, 2008.
From the New Mexico Office of the Medical Investigator, Albuquerque, NM.
Correspondence: Othon J. Mena, MD, County of San Diego Medical Examiner's Office, 5570 Overland Ave, San Diego, CA 92123. E-mail: othon.mena@sdcounty.ca.gov.

ISSN: 0195-7910/10/0000-0001
DOI: 10.1097/PAF.0b013e3181edee5b

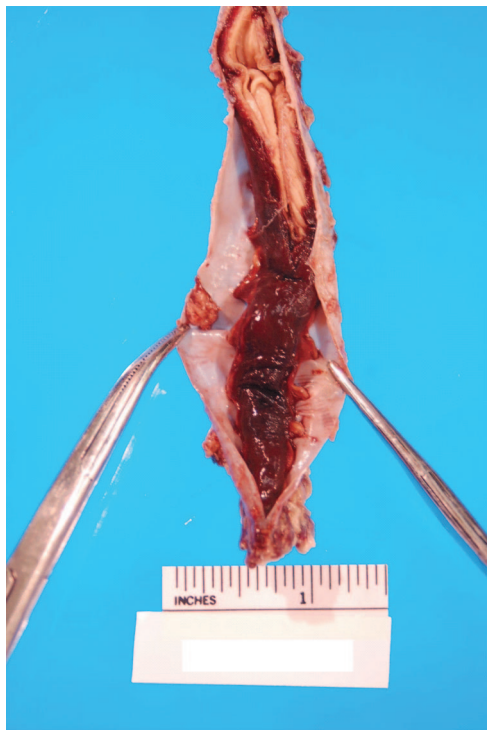

FIGURE 2. Subarachnoid hemorrhage was present along entire length of spinal cord.

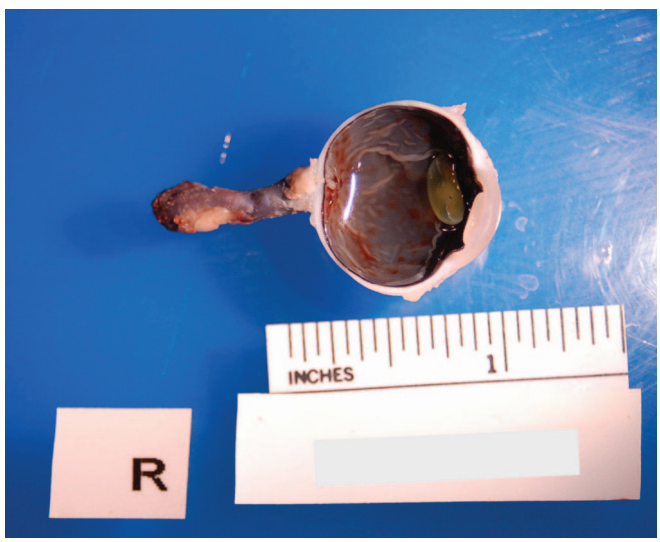

FIGURE 3. Multiple retinal hemorrhages extended to the ora serrata. The entire optic nerve sheath was darkened by marked hemorrhage.

DISCUSSION

It is well known that retinal hemorrhages are associated with inflicted traumatic brain injury. Nevertheless, other causes associated with them include raised intracranial pressure, intracranial subarachnoid hemorrhage, vaginal delivery, blood dyscrasias, and others.[1,2] In the early 20th century, Terson syndrome was originally described as vitreous hemorrhage in association with intracranial hemorrhage, more specifically subarachnoid hemorrhage.[3] Since the original description, Terson syndrome has been expanded to encompass any intraocular hemorrhage associated with intracranial hemorrhage and intracranial pressure elevation.[2,4]

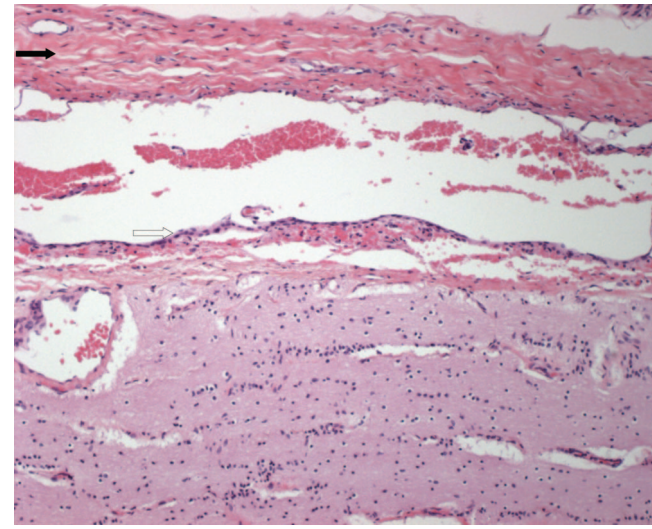

FIGURE 4. The majority of the hemorrhage along the optic nerve sheath was intradural and subdural, with a small amount being subarachnoid (hematoxylin and eosin, 100×) (black arrow indicates dura, white arrow indicates subarachnoid membrane).

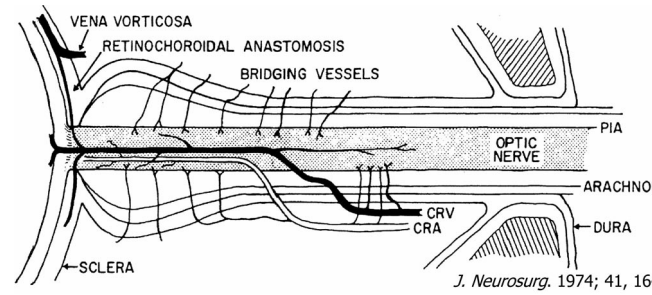

FIGURE 5. Schematic proposed by Muller and Deck (1974), illustrating the intraorbital and intracanalicular portion of the optic nerve and its sheath. Bridging vessel rupture would result in optic nerve sheath hemorrhage. Reprinted with permission from *J Neurosurg*. 1974;41:164.

Different mechanisms have been proposed to explain the occurrence of ocular hemorrhage following a rise in intracranial pressure, whether it is due to an intracranial hemorrhage or any other cause. It has been suggested that a rapid rise in intracranial pressure leads to decreased venous drainage of the eyes, resulting in vitreous hypertension and retinal or vitreous hemorrhage.[5,6] In 1974, it was proposed by Muller and Deck that the rise in intracranial pressure on the subarachnoid compartment dilates the optic sheath, leading to compression of the central retinal vein (Fig. 5). The hemorrhage would then originate from ruptured intradural vessels and bridging vessels, following compression and obstruction of the retinochoroidal anastomoses. Compression of the central retinal vein would also result in blocked or reduced eye venous drainage, resulting in retinal venous hypertension and retinal vein rupture.[5]

There is evidence that there is no connection between the optic nerve sheath subarachnoid space and the vitreous body, and that the optic nerve sheath hemorrhage is not necessarily an extension of

intracranial subarachnoid hemorrhage.[6] Some of the reasons for this are that intraocular hemorrhage can be seen accompanying raised intracranial pressure without intracranial hemorrhage, and the subdural compartment is generally the compartment with the bulk of the hemorrhage.[5,6] Therefore, raised intracranial pressure regardless of the cause (eg, severe brain injury) may result in intraocular hemorrhage.

The findings of the case presented initially mimicked the presentation of inflicted traumatic brain injury. However, a complete and thorough autopsy examination was instrumental in determining that the cause was in fact natural. Thus, this case represents a rare example of a nontraumatic cause of ocular hemorrhage and a case of Terson syndrome in an infant. Whether there is a head injury or an intracranial hemorrhage due to a spontaneously ruptured aneurysm or any other cause, the most significant factor leading to ocular and optic nerve serve sheath hemorrhage appears to be a rapid increase in intracranial pressure.

REFERENCES

1. Kaur B, Taylor D. Current Topic: retinal hemorrhages. *Arch Dis Child*. 1990;65:1369–1372.
2. Jay V, Smith CR. Check Sample, American Society of Clinical Pathologists (ASCP), Forensic Pathology, 1998;40(9).
3. Ou R. Terson Syndrome (e-medicine Web MD Web site). July 3, 2008. Available at: http://www.emedicine.com/oph/topic753.htm#ref1. Accessed August 29, 2008.
4. Giangiacomo J, Frasier LD. Ophthalmic Manifestations. In: Giardino MP, Alexander R, eds. *Child Maltreatment*. 3rd ed. Saint Louis, MO: GW Medical Publishing Inc;2005:83–89.
5. Muller PJ, Deck JH. Intraocular and optic nerve sheath hemorrhage in cases of sudden intracranial hypertension. *J Neurosurg*. 1974;41:160–166.
6. Medele RJ, Stummer W, Mueller AJ, et al. Terson's syndrome in subarachnoid hemorrhage and severe brain injury accompanied by acutely raised intracranial pressure. *J Neurosurg*. 1998;88:851–854.