Terson syndrome secondary to aneurysmal subarachnoid hemorrhage in a child: illustrative case

Jacob M. Mazza, MD,1,2 Parth Tank, BS,3 Melissa A. LoPresti, MD, MPH,1 Jonathan P. Scoville, MD, MSPH,4 Brenda L. Bohnsack, MD, PhD,5 and Sandi Lam, MD, MBA1

1Division of Pediatric Neurosurgery, Department of Neurosurgery, Lurie Children’s Hospital, Northwestern University Feinberg School of Medicine, Chicago, Illinois; 2Department of Neurological Surgery, Rush University Medical Center, Chicago, Illinois; 3Chicago Medical School of Rosalind Franklin University, Chicago, Illinois; 4Department of Neurological Surgery, University of Utah School of Medicine, Salt Lake City, Utah; and 5Division of Pediatric Ophthalmology, Department of Ophthalmology, Lurie Children’s Hospital, Northwestern University Feinberg School of Medicine, Chicago, Illinois

BACKGROUND Terson syndrome is the phenomenon of intraocular hemorrhage in the setting of subarachnoid hemorrhage (SAH). Vision loss can lead to morbidity for the affected individual. Aneurysmal SAH related to intracranial aneurysms is rare in children. Studies have shown the incidence of Terson syndrome in adults with aneurysmal SAH to be over 40%; however, few cases of Terson syndrome in pediatric aneurysmal SAH have been reported.

OBSERVATIONS A 9-year-old male presented with altered mental status and seizures. Computed tomographic angiography showed aneurysmal SAH from a ruptured, left-sided posterior inferior cerebellar artery aneurysm. The patient underwent endovascular treatment with coiling and external ventricular drainage for SAH. Ophthalmological consultation for blurry vision revealed the diagnosis of Terson syndrome with decreased vision in the left eye, which was managed conservatively.

LESSONS Terson syndrome after SAH can occur in children. Prompt ophthalmological evaluation in pediatric patients with aneurysmal SAH is vital for recognition and management to decrease overall morbidity.

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KEYWORDS Terson syndrome; pediatric; aneurysm; subarachnoid hemorrhage; intraocular hemorrhage

Subarachnoid hemorrhage (SAH) is the extravasation of blood within the subarachnoid space that most commonly results from ruptured intracranial aneurysms (aneurysmal SAH), second only to trauma.1 Intracranial aneurysms (IA)s occur in approximately 3% of the general population, with uneven distribution across the age spectrum: only 5% of all aneurysm cases are found in children age < 18 years, even when the pediatric population constitutes over one-fifth of the population.2 Fifty-three percent of pediatric patients with IA present with aneurysmal SAH, while only 37% of adults present with aneurysmal SAH at some point throughout their life.3

Terson syndrome (TS) is the phenomenon of intraocular hemorrhage that occurs in the setting of SAH, including aneurysmal SAH. This entity can cause reversible and, in some cases, permanent vision loss resulting in significant morbidity.4 In recent retrospective and prospective studies of adults with aneurysmal SAH, upwards of 40% have been shown to develop TS.5,6 In pediatric aneurysmal SAH, it is believed that only 2% of patients are affected by TS.2,7 Only 3 cases in the literature describe findings of TS in pediatric patients with aneurysmal SAH, one of which was only discovered after autopsy.7,8 This highlights a scarcity of information regarding the workup and management of this patient population. Here, we describe a pediatric patient with aneurysmal SAH found to have TS. We highlight this case to review the key evaluation, diagnostic, and management considerations in TS in children with aneurysmal SAH.

Illustrative Case

A 9-year-old male without significant past medical history presented to a hospital with nausea and severe headache. The patient
subsequently had a generalized seizure. Head computed tomography (CT) demonstrated SAH primarily located in the pre-pontine and interpeduncular cisterns, as well as cisterna magna (Fig. 1). Intraventricular hemorrhage (IVH) was noted mostly in the fourth ventricle with diffuse ventriculomegaly consistent with hydrocephalus.

The patient was transferred to our institution for neurosurgical evaluation and management of the Hunt and Hess grade III, modified Fisher grade 4 SAH. CT angiogram revealed a proximal left posterior inferior cerebellar artery (PICA) aneurysm (Fig. 2). The patient underwent right frontal external ventricular drain (EVD) placement. Digital subtraction angiography confirmed a postero-medially projecting, multilobed, large left-sided PICA aneurysm measuring 3.0 $\times$ 5.5 mm, with a wide neck (Fig. 2).

The patient underwent coil embolization of the left PICA aneurysm with radiographic occlusion (Fig. 3) using 4 coils (Axiom Prime 3-D coils, $3 \times 6$ mm, $2 \times 4$ mm, $1 \times 3$ mm, and $1 \times 2$ mm, Medtronic, USA). The patient was treated in the pediatric intensive care unit for EVD management and monitoring for vasospasm. The patient’s course was complicated by fungal meningitis treated with antimicrobial therapy for 21 days and subsequent endoscopic third ventriculostomy for hydrocephalus treatment. On post-bleed day 20, when the patient’s condition had improved to the point that he was able to verbally communicate, he reported decreased vision in his left eye. The visual acuity was 20/30 in the right eye, and he only had the ability to detect hand motion in the left eye. There were scattered-flame intraretinal hemorrhages, not involving the fovea, throughout the right eye. In the left eye, the view to the retina and optic nerve was obstructed by a dense hemorrhage consisting of partially de-hemoglobinized blood within the vitreous. A B-scan ultrasound of the left eye showed possible preretinal hemorrhage trapped within the subhyaloid space.

FIG. 1. Noncontrast enhanced CT scan of the brain with the (A) axial plane showing casting of the fourth ventricle with acute hemorrhage, (B) coronal plane showing dilation of the lateral and third ventricular system with prominent temporal homs and intraventricular hemorrhage within the third ventricle, and (C) sagittal plane showing diffuse preponitine, interpeduncular, and cisterna magna subarachnoid hemorrhage.

FIG. 2. (A–C) Three-axis CT angiography images showing a left PICA aneurysm. Oblique (D) and lateral (E) digital subtraction angiographic projections showing a multilobed left PICA aneurysm.

FIG. 3. (A–C) Digital subtraction images showing a right frontal external ventricular drain in the lateral ventricle. (D) and (E) Digital subtraction angiography images showing the left PICA aneurysm with a wide neck.
The patient continued to make progress in recovery and was subsequently discharged on post-bleed day 39 to inpatient acute rehabilitation. Repeat eye examination at 2 months showed uncorrected visual acuity of 20/20 in the right eye and 20/500 in the left eye. The intraretinal hemorrhages in the right eye had resolved. In the left eye, there was still a moderate amount of red blood cells within the vitreous, preventing adequate visualization of the optic nerve and macula (Fig. 4).

Discussion

Observations

We present a case of a 9-year-old male who experienced a Hunt and Hess grade III, modified Fisher grade 4 aneurysmal SAH after left PICA aneurysm rupture, with decreased vision in the left eye secondary to TS. We highlight this case to review the prevalence and considerations in the evaluation and management of TS in children with SAH.

TS Is Rare but Important to Recognize

Named for ophthalmologist Dr. Albert Terson, TS is the association of intraocular hemorrhages with SAH. Often reported in the literature in relation to blunt head trauma with associated intracranial and subarachnoid hemorrhage, this entity can arise from different pathologies, leading to SAH. The pathophysiology remains contested, but the predominant theory is that elevated intracranial pressure in the setting of SAH leads to ingress of cerebrospinal fluid within the optic sheath, subsequently dilating the nerve posterior to the bulb. As a result of the dilation, there is compression of the central retinal vein causing venous congestion and rupture of small vessels. A second theory involves lymphatic reflux that parallels the general disruption of normal fluid dynamics within and around the globe.

While this report focuses on TS in the setting of aneurysmal SAH, there are other well-documented etiologies of TS in the pediatric population. Similar to adults, traumatic brain injury, both with and without intracranial hemorrhage, can lead to TS. The pathologic mechanism in these instances mimics that of aneurysmal SAH; increased intracranial pressure leads to nerve edema, subsequent venous congestion, and resultant small vessel rupture, leading to intracranial hemorrhage. The incidence of TS in children related to traumatic brain injury is also rare but considered to have a similar incidence to TS related to aneurysmal SAH. In children, a long-standing pillar of screening pediatric patients for nonaccidental trauma is the presence of intracranial hemorrhage, highlighting this phenomenon in the setting of trauma of varying severity. This continues to be a component of the non-accidental trauma workup in this population.

An important aspect of TS within the aneurysmal SAH patient population is that there can be a large portion of this patient population who are severely disabled from the inciting neurological injury. As such, these patients may not be able to communicate visual complaints, especially if compounded with visual field defects secondary to neurological injury. Thus, TS may be underdiagnosed due to a lack of ophthalmic screening within a few weeks of injury. Prospective studies identify significantly higher percentages of patient cohorts that have TS, as compared with retrospective studies—a finding that shows the underreported nature of TS in aneurysmal SAH. This highlights the importance of standardized approaches to screening for proper identification of TS in this vulnerable population.

Terson syndrome in the setting of aneurysmal SAH has been seen in adults with more severe Hunt and Hess and modified Fisher grades, and is believed to lead to increased morbidity and mortality within this patient population. A systematic review of both prospective and retrospective studies in adults with TS showed that patients with aneurysmal SAH with associated intracranial hemorrhage have worse outcomes, underscoring the severity of disease in these patients and perhaps suggesting underlying links in pathophysiology. In a study of 102 patients with SAH, 20 of whom developed TS, 55% of patients with TS had Hunt and Hess IV and V, as compared to 13% in those without TS. In a similar study of 60 patients with SAH, all patients with TS had a Hunt and Hess grade III, IV, or V (82% of which were Hunt and Hess 4 or 5). Only 20% of those without TS had Hunt and Hess grade IV or V. In a review of 45 patients with spontaneous SAH, the median Hunt and Hess grade between those with TS and those without was 4 and 2, respectively. These studies highlight the importance of ophthalmological screening in the SAH population presenting with more severe injury. Additionally, all 3 studies described correlated greater mortality and worse functional outcomes for patients with TS as compared with those without.

TS in Children: A Review of the Literature

This is the fourth reported case of pediatric TS secondary to aneurysmal SAH. This is perhaps due to the rarity of the disease, although also may be due to pediatric patients being unable to verbalize visual disturbances due to their young age or neurological injury. Reported cases are summarized in Table 1. Two patients aged 7 months, 1 aged 1 month, are compared with our patient with
regard to patient demographics, presentation, evaluation, and management. One patient expired,\(^8\) one underwent aneurysm clipping in addition to our case,\(^7\) and one was treated conservatively.\(^8\) At this time, none of the reported cases in the literature, nor our patient, required ophthalmological intervention for TS.

Our reported patient was 9 years old who presented with both intraretinal and vitreous hemorrhages. Although this patient’s ophthalmic course continues to evolve, based on his last eye examination he is likely to require vitrectomy for nonclearing vitreous and subhyaloid hemorrhage. In children, the vitreous is well formed with higher viscosity and the hyaloid face is still intact compared with that in adults. As a result, in the pediatric population, vitreous hemorrhages are less likely to spontaneously clear. On the other hand, complications common in adults, such as retinal detachments and folds and epiretinal membranes, are less likely to occur in children.

Notably, the 3 previously reported pediatric cases of TS were all younger than 12 months of age, such that this diagnosis had only been reported in infants and adults. The true incidence of TS is not known. With this report, we complement the literature with a novel case,\(^7\) and one was treated conservatively.\(^8\) At this time, none of the reported cases in the literature, nor our patient, required ophthalmological intervention for TS.

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age</th>
<th>Sex</th>
<th>Presentation</th>
<th>Imaging</th>
<th>Diagnosis</th>
<th>Surgical Intervention</th>
<th>Ophthalmological symptoms and findings</th>
<th>Treatment/ outcome</th>
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<tbody>
<tr>
<td>Bhardwaj et al., 2010(^7)</td>
<td>7 mos</td>
<td>Female</td>
<td>Unresponsiveness, apnea</td>
<td>CT: right temporo parasietal SAH</td>
<td>Ruptured aneurysmal SAH</td>
<td>Clipping of aneurysm</td>
<td>Preterinal &amp; intraretinal hemorrhage of rt eye</td>
<td>Clinical monitoring; spontaneous resolution of hemorrhage at 3 mos</td>
</tr>
<tr>
<td>Mena et al., 2011(^8)</td>
<td>7 mos</td>
<td>Female</td>
<td>Irritability, hypotonia</td>
<td>CTA: fusiform aneurysm at anterior division of rt middle cerebral artery bifurcation</td>
<td>Ruptured aneurysmal SAH</td>
<td>No treatment</td>
<td>Bilateral optic sheath hemorrhage, extensive retinal hemorrhage extending to the ora serrata</td>
<td>Mortality prior to intervention/treatment</td>
</tr>
<tr>
<td>Scheller et al., 2015(^9)</td>
<td>1 mo</td>
<td>Female</td>
<td>Seizure</td>
<td>Gross specimen: rt middle cerebral artery aneurysm</td>
<td>Ruptured aneurysmal SAH with tt subarachnoid hemorrhage</td>
<td>No treatment</td>
<td>Bilateral retinal hemorrhage</td>
<td>No documented treatment/ follow-up</td>
</tr>
<tr>
<td>Present Case</td>
<td>9 yrs</td>
<td>Male</td>
<td>Headache, nausea</td>
<td>CT: rt-sided SAH, subdural hemorrhage &amp; round lesion in suprasellar area</td>
<td>Ruptured aneurysmal SAH with tt subarachnoid hemorrhage</td>
<td>Coiling of aneurysm</td>
<td>Bilateral retinal hemorrhage with tt subarachnoid hemorrhage</td>
<td>Clinical monitoring; improvement of rt eye acuity to 20/20, lt eye acuity to 20/500; moderate hemorrhage in lt eye</td>
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**TABLE 1. Comparison of TS cases in pediatric SAH**

CTA = computed tomography angiogram; IVH = intraventricular hemorrhage.

### Considerations in Management

The ophthalmological management of TS is based on the extent and location of hemorrhages and subsequent complications. Further, in infants and young children, clearing of the visual axis may be required to prevent vision loss from amblyopia.\(^10\) Vitrectomy surgery is indicated for nonclearing vitreous or subhyaloid hemorrhages, retinal detachments and folds, and visually significant epiretinal membranes. There is not 1 specific recommended time window for vitrectomy because multiple variables are taken into clinical consideration from ophthalmology specialists. From the literature, in adults, faster time-to-surgery following diagnosis and younger age have been predictive of better visual outcomes.\(^23\) One study indicated that time-to-referral after symptom onset was anywhere from 4 to 5 months, highlighting a significant delay in diagnosis and management.\(^13\) In the same study, 22 of 25 adults who underwent vitrectomy had improvement in acuity to 20/30 or better.\(^24\) Another cohort of 30 adults, 16 of whom were observed and 14 of whom underwent vitrectomy, had improvement in acuity to at least 20/50 in 75% and 86%, respectively.\(^25\) A study of TS in 28 eyes among 19 children, some of which experienced traumatic brain injury, vitrectomy was associated with improved visual acuity but not without mention of complications such as retinal detachment and epiretinal membrane formation.\(^19\) Overall, the data on children remain sparse. In adults, management decisions must be guided by clinical severity with the selection of conservative (observation) and operative management options determined by the extent and location of hemorrhages and...
subsequent complications. For children, the variable of age must be taken into consideration, among the concurrent clinical comorbidities involved. The younger the child, the more quickly surgical intervention may be considered for the affected eye because of amblyopia and vision development during formative years. For instance, if a patient were under 5 years of age, the intervention would likely happen within a few months of clinical follow-up from the initial insult; this is especially the case in infants under 12 months of age. In the present case, the patient is 9 years of age, which explains the longer time frame allowed for conservative management and follow-up. Vitreectomy may be recommended 6–12 months after the initial bleed in his case (a time frame that may sometimes be longer in adults).

While the numbers reported in the pediatric literature are too few to fully examine the impact of diagnosis and management on outcomes, the identification of TS as a disease in children, though rarer than in adults, allows for prompt diagnosis, early consideration of management, and coordination of care in these often-sick patients. Given that aneurysmal SAH can lead to significant neurological deficit, it is often not feasible to clinically assess these patients for signs and symptoms of TS without an ophthalmological examination. This is in conjunction with challenges in eliciting visual complaints in young children on physical examination. Furthermore, complex medical sequelae that accompany aneurysmal SAH, such as hydrocephalus, stroke, seizure, and other encephalopathy-inducing processes, may often further obscure clinical signs of TS, leading them to go unnoticed. If TS is diagnosed in a timely manner and clinical decision making is driven by a multidisciplinary approach, including ophthalmology, neurosurgery, neurology, and intensive care physicians, among others, any additional visual morbidity may be avoided in these patients. Thus, we suggest a role for ophthalmological evaluation for screening in the setting of pediatric SAH as part of acute multidisciplinary assessment and management within the first few weeks of injury.

**Lessons**

Terson syndrome is a phenomenon that must be recognized to ensure appropriate evaluation, management, and follow-up in pediatric patients who experience aneurysmal SAH. This syndrome has been well documented in adult aneurysmal SAH: while pediatric aneurysmal SAH is less common, this patient population is also at risk of developing TS and experience devastating ophthalmological sequelae. Routine screening and evaluation, in conjunction with subspecialty consultation, can help limit long-term neurological deficits and aim to decrease morbidity. This is especially important in patients presenting with Hunt and Hess grade III or worse SAH and those too young to reliably verbalize ophthalmological complaints. While our study is limited due to the low sample size given the rarity of the disease, we believe this case nicely highlights a rare pathology in children, which, with prompt recognition and multidisciplinary care, may help improve outcomes.

**References**


Disclosures
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions
Conception and design: Lam, LoPresti. Acquisition of data: Mazza, Tank. Analysis and interpretation of data: Mazza, Scoville. Drafting of the article: Mazza, Tank, LoPresti, Scoville. Critically revising the article: Lam, Mazza, LoPresti, Scoville, Bohnsack. Reviewed submitted version of the manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Lam. Administrative/technical/material support: Lam, Mazza. Study supervision: Lam, LoPresti.

Correspondence
Sandi Lam: Lurie Children’s Hospital, Chicago, IL. slam@luriechildrens.org.