Terson’s syndrome: a reversible cause of blindness following subarachnoid hemorrhage

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✓ Terson’s syndrome refers to the occurrence of vitreous hemorrhage with subarachnoid hemorrhage (SAH), usually due to a ruptured cerebral aneurysm. Although it is a well-described entity in the ophthalmological literature, it has been only rarely commented upon in the neurosurgical discussion of SAH.

Fundus findings are reported in a prospective study of 22 consecutive patients with a computerized tomography- or lumbar puncture-proven diagnosis of SAH. Six of these patients had intraocular hemorrhage on initial examination. In four patients vitreous hemorrhage was evident on presentation (six of eight eyes). In the subsequent 12 days, vitreous hemorrhage developed in all the additional two patients (three of four eyes) due to breakthrough bleeding from the original subhyaloid hemorrhages.

The initial amount of intraocular hemorrhage did not correlate with the severity of SAH. Two of the six patients with intraocular hemorrhage died, whereas five of the 16 remaining SAH patients without intraocular hemorrhage died. Of the four survivors with intraocular hemorrhage, three showed gradual but significant improvement in their visual acuity by 6 months. The fourth underwent vitrectomy at 8 months after presentation and had a good visual result. With modern and aggressive medical and microsurgical management, Terson’s syndrome should be recognized as an important reversible cause of blindness in patients surviving SAH.

KEY WORDS • intraocular hemorrhage • subarachnoid hemorrhage • subhyaloid hemorrhage • Terson’s syndrome • vitreous hemorrhage

IN 1900, the French ophthalmologist, Albert Terson, first described vitreous hemorrhage occurring as the direct result of subarachnoid hemorrhage (SAH). In the 50 years that followed, only 16 cases of this seemingly rare entity were reported in the world medical literature. Since the early 1950’s, several retrospective studies have looked at the incidence of vitreous hemorrhage in patients presenting with SAH. These studies, with between 200 and 500 patients each, showed Terson’s syndrome to be more common than was originally thought. Vitreous hemorrhage was demonstrated in 3% to 5% of patients presenting with SAH. In addition, intraocular hemorrhage that did not initially involve the vitreous was observed in 20% to 40% of patients.

Shaw and Landers found the mortality rate to be twice as high for SAH in the presence of intraocular hemorrhage than for patients with SAH alone. Others have shown similar results. For patients presenting with suspected SAH, therefore, the presence of intraocular hemorrhage has both diagnostic and prognostic significance.

Clinical Material and Methods

Twenty-two consecutive patients presenting with SAH to the Sir Mortimer B. Davis Jewish General Hospital were studied prospectively. Each patient was examined by computerized tomography (CT) and if no blood was observed in the subarachnoid space, a lumbar puncture was performed to establish the diagnosis of SAH. Patients were graded according to the Hunt and Hess scale. Those patients who were clinically stable underwent early angiography. A screening ophthalmological examination was performed routinely upon presentation, and patients with intraocular hemorrhage received serial examinations that included fundus photography and ultrasound studies with B-mode scans.

One of our cases of Terson’s syndrome was studied at autopsy. The globes and the intraorbital portion of the optic nerves were removed intact and were fixed in formalin. Prior to embedding in paraffin, the specimens were stained with hematoxylin and eosin (H & E). Microtome-sectioned specimens were then studied and photographed.
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TABLE 1
Characteristics of six patients with Terson’s syndrome

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs.)</th>
<th>Sex</th>
<th>Source of Hemorrhage*</th>
<th>Initial Clinical Grade†</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>43, M</td>
<td></td>
<td>ACoA aneurysm</td>
<td>I</td>
</tr>
<tr>
<td>2</td>
<td>47, F</td>
<td></td>
<td>ACoA aneurysm</td>
<td>III</td>
</tr>
<tr>
<td>3</td>
<td>36, F</td>
<td></td>
<td>MCA aneurysm</td>
<td>IV</td>
</tr>
<tr>
<td>4</td>
<td>65, F</td>
<td></td>
<td>AChA aneurysm</td>
<td>V</td>
</tr>
<tr>
<td>5</td>
<td>51, F</td>
<td></td>
<td>trauma</td>
<td>IV</td>
</tr>
<tr>
<td>6</td>
<td>72, F</td>
<td></td>
<td>ACoA aneurysm</td>
<td>V</td>
</tr>
</tbody>
</table>

* Abbreviations: ACoA = anterior communicating artery; MCA = middle cerebral artery; AChA = anterior choroidal artery.
† According to the scale of Hunt and Hess.11

Results

Of the 22 patients with SAH, six (27%) were identified as having intraocular hemorrhage. The six patients ranged in age from 36 to 72 years (mean 52 years), and five were women. The 16 patients with SAH alone ranged in age from 34 to 77 years (mean 56 years); 11 of them were women. The mean initial clinical grade of 3.7 was slightly higher for SAH patients with intraocular hemorrhage than the 3.2 for those with SAH alone; however, the difference between the two groups did not reach statistical significance.

Aneurysmal rupture of the anterior communicating artery (ACoA) was responsible for SAH in three of the six patients with intraocular hemorrhage. Anterior choroidal artery rupture was evident in another. One patient developed SAH secondary to a head injury and no aneurysm was identified on angiography. The final patient had a clinically suspected middle cerebral artery aneurysmal hemorrhage; however, multiple aneurysms were demonstrated on angiography. The characteristics of our patients with Terson’s syndrome are shown in Table 1 along with their initial clinical grade.

TABLE 2
Ocular findings in six patients with Terson’s syndrome*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Type of Intraocular Hemorrhage</th>
<th>Visual Acuity</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Initial 6 Months</td>
</tr>
<tr>
<td>1</td>
<td>rt: retinal only</td>
<td>20/20 20/20</td>
</tr>
<tr>
<td></td>
<td>lt: retinal, subhyaloid, &amp; vitreous</td>
<td>20/200 20/40</td>
</tr>
<tr>
<td>2</td>
<td>rt: subhyaloid &amp; vitreous</td>
<td>HM 20/50</td>
</tr>
<tr>
<td></td>
<td>lt: subhyaloid only</td>
<td>CF 20/40</td>
</tr>
<tr>
<td>3</td>
<td>rt: subhyaloid &amp; vitreous</td>
<td>LP HM</td>
</tr>
<tr>
<td></td>
<td>lt: subhyaloid &amp; vitreous</td>
<td>LP CF</td>
</tr>
<tr>
<td>4</td>
<td>rt: subhyaloid &amp; vitreous</td>
<td>LP (died)</td>
</tr>
<tr>
<td></td>
<td>lt: subhyaloid &amp; vitreous</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>rt: subhyaloid only</td>
<td>20/20 20/20</td>
</tr>
<tr>
<td></td>
<td>lt: retinal &amp; subhyaloid</td>
<td>CF 20/200</td>
</tr>
<tr>
<td>6</td>
<td>rt: retinal &amp; subhyaloid</td>
<td>20/40 (died)</td>
</tr>
<tr>
<td></td>
<td>lt: retinal only</td>
<td>20/20</td>
</tr>
</tbody>
</table>

* Abbreviations: rt = right; lt = left; HM = perceives hand motion; CF = can count fingers; LP = perceives light.

Death from SAH occurred in two of the six patients with intraocular hemorrhage and five of the 16 without. The difference between the two groups was not statistically significant. The first obtainable visual acuity measurements are shown in Table 2 and compared with the results of a follow-up acuity test approximately 6 months after initial presentation. The specific type of intraocular hemorrhage is described for each eye.

Vitreous hemorrhage was evident on presentation in four of six patients, with six of eight eyes involved. In the subsequent 12 days, the remaining two patients with intraocular hemorrhage each developed vitreous hemorrhages (in three of four eyes). A fundus photograph of the left eye of the 43-year-old man in Case 1 is shown in Fig. 1 left. Two subhyaloid hemorrhages are seen below the optic disc and obscure the underlying retinal vasculature. The other hemorrhages are retinal.

Fig. 1. Ocular fundus photographs of the left eye. Left: Case 1. Two subhyaloid hemorrhages appear inferior to the optic disc and obscure the underlying retinal vasculature. Retinal hemorrhages surround the fovea in a stellate pattern. Right: Case 6. Retinal hemorrhages alone are visible.
and surround the fovea for the most part. The vitreous hemorrhage, which was present inferiorly, is not evident in this photograph.

Figure 1 right is a fundus photograph of the left eye of the 72-year-old woman in Case 6. Retinal hemorrhages are seen, yet no subhyaloid or vitreous hemorrhage is present. These fundus photographs illustrate the spectrum of intraocular hemorrhage seen in patients with SAH. The first patient did well clinically and the second patient died 9 days after initial presentation.

With the exception of Case 3 (a 36-year-old woman), all surviving patients demonstrated a significant improvement in visual acuity by 6 months. Examination of Case 3 showed no evidence of retinal membrane formation or detachment. Eight months after presentation, the patient underwent a vitrectomy in her right eye and the visual acuity improved from perception of hand motions only to 20/50. She has recently undergone a similar procedure for the left eye and visual acuity has improved from counting fingers to 20/70.

Pathological study in our autopsy case with Terson’s syndrome showed intraretinal, subhyaloid, and vitreous hemorrhages in both eyes. There was a moderate expansion of the retrobulbar portion of the optic nerve sheath immediately adjacent to the sclera. Optic nerve sheath hemorrhage was clearly evident, with the subdural space showing more extensive involvement than the subarachnoid. Axial and transverse H & E-stained sections from this case are presented in Fig. 2.

Discussion

Definition

Classically, Terson’s syndrome has referred to vitreous hemorrhage seen in patients with SAH secondary to aneurysmal rupture. It is most commonly reported with aneurysms of the anterior circulation, particularly those of ACoA and internal carotid artery (ICA) origin. Terson’s syndrome has also rarely been described with subdural hematomas, trauma causing SAH, and aneurysms of the posterior circulation.25-32,33

Mechanisms for Intraocular Hemorrhage

Several mechanisms have been proposed to explain the occurrence of intraocular hemorrhage with SAH. One early theory suggests that blood simply tracks through the subarachnoid space into its continuation within the optic nerve sheath. The blood then penetrates the sclera in the porous region which transmits the optic nerve (lamina cribrosa), and finally appears in the vitreous space within the globe.7 Pathological studies using electron microscopy have not been able to demonstrate a connection between the optic nerve sheath subarachnoid space and the vitreous.2,20,27,35 In addition, intraocular hemorrhage has been shown to occur with a sudden increase in intracranial pressure (ICP) not due to SAH. In spite of this evidence, many textbooks including some recent neurological texts still attribute Terson’s syndrome to this mechanism.1,6

A second mechanism is proposed whereby intraocular hemorrhage is due to retinal venous stasis, which occurs secondary to the rapid increase in ICP with SAH.3,13,16,27,35 This sudden rise in ICP is thought to decrease venous return to the cavernous sinus from the veins that drain the globe. Increased retinal venous pressure results in stasis followed by vessel rupture and the observed intraocular hemorrhage. The orbital vessels may drain through outflow into the extracranial facial and pterygoid venous systems should increased ICP reduce venous return through the cavernous sinus.15 Thus, this mechanism for intraocular hemorrhage is unlikely, since increased ICP would presumably not affect the extracranial path of venous return from the eye. Riddoch and Goulden24 and Symonds25 have pro-
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posed that retinal venous return could be reduced by compression of the central retinal vein and that this would result in venous stasis and retinal hemorrhage. The compression would be caused by extravasated blood, which is seen both within and surrounding the optic nerve sheath near the exit of the central retinal vein. Hayreh\(^5\) occluded the central retinal vein at its exit from the optic nerve sheath, and reported that this did not produce retinal hemorrhages in rhesus monkeys. Venous drainage was thought to occur through another site, the retinochoroidal anastomoses. These drain through the posterior ciliary vessels, as is shown in Fig. 3.

A more recently proposed mechanism suggests that, with increased ICP due to SAH, there is a rapid effusion of cerebrospinal fluid through the communication of the subarachnoid space within the optic nerve sheath.\(^{18,19,21,32}\) When dilated, the retrolubar portion of the optic nerve sheath could compress and therefore obstruct both the retinochoroidal anastomoses and the central retinal vein. This would then produce an acute decrease in venous drainage from the retina that would result in stasis and hemorrhage.

The retinochoroidal anastomoses are situated near the junction of sclera and the optic nerve, and are well anterior to the subarachnoid space, as shown in Fig. 3. These would only be susceptible to compression with significant retrolubar optic nerve sheath dilatation and hence only certain patients with SAH could develop intraocular hemorrhage. Finally, stretching of the optic nerve sheath would shear bridging vessels which would bleed into the subdural and subarachnoid spaces of the nerve sheath, thereby further compromising venous drainage.

**Clinical and Pathological Review**

Muller and Deck\(^21\) reported a pathological review of the eyes of 23 patients who had died from sudden intracranial hypertension. Expansion of the retrolubar portion of the optic nerve sheath was evident in virtually all of the patients. Optic nerve sheath hemorrhage was seen in 87% of the cases studied. Hemorrhage into the subdural space was more common than into the subarachnoid space, indicating that the blood present did not simply track through the subarachnoid space. With dilatation of the optic nerve sheath, rupture of intradural and bridging pial-dural vessels did occur and probably contributed to the acute decrease in retinal venous drainage. Only 37% of the eyes studied had intraocular hemorrhage, demonstrating that optic nerve sheath hemorrhage and dilatation were not always sufficient to result in the complete obstruction of venous return. The light microscopic study of our autopsy case showed changes similar to those found in their investigation.

Walsh and Hedges\(^45\) reviewed the clinical and pathological findings in 90 cases of spontaneous SAH. They observed that the amount of hemorrhage surrounding the optic nerve sheath was directly related to the rate of onset of the clinical symptoms prior to death. Those patients with a significant amount of optic nerve sheath hemorrhage were more likely to have had papilledema, cerebral edema, and retinal and subhyaloid hemorrhages.

In a retrospective study of 195 patients with ruptured intracranial aneurysms, Fahmy\(^4\) found that patients with intraocular hemorrhage had a greater amount of bleeding into the subarachnoid space. He noted an increased association of intraocular hemorrhage with ACoA aneurysms and suggested that this was due to the tendency for these aneurysms to bleed more profusely. He even suggested that the presence of intraocular hemorrhage be used as an independent risk factor in grading patients with SAH.\(^9\) This was not supported by our study.

**Visual Implications**

The vitreous hemorrhage seen in Terson's syndrome is usually bilateral; however, it may be very asymmetrical in nature. It is suggestive but not pathognomonic of a specific intracranial process. Blood may mechanically break through the internal limiting membrane of the retina and the posterior hyaloid membrane of the vitreous from initial subhyaloid or preretal hemorrhages. For this reason, vitreous hemorrhage may appear several days after SAH. In our study, two patients developed new vitreous hemorrhage in the 2 weeks following their SAH. This was attributed to extension of the intraocular hemorrhage seen on initial examination, because there was no clinical evidence of aneurysmal rebleeding. Thus, it is important to document the initial fundus examination.

One of our patients, a 47-year-old woman with an ACoA aneurysm (Case 2), presented with an acute decrease in vision in her right eye several days before...
she developed SAH. There was no history of hypertension, vascular disease, or diabetes mellitus, and headache was not a prominent symptom. On examination, there was a dense vitreous hemorrhage in the right eye and a subhyaloid hemorrhage in the left with normal-appearing optic discs. While undergoing outpatient workup, she developed a severe right-sided headache and focal neurological signs indicative of SAH. The appearance of intraocular hemorrhage before the clinical signs of SAH in a patient with Terson’s syndrome has not been previously reported in the literature.

Patients with preretinal, subhyaloid, or vitreous hemorrhages may complain of “floaters,” blurred vision, or an overall decrease in vision depending on the extent of the intraocular hemorrhage. Further decrease in a patient’s vision or increase in the number of floaters seen may indicate an extension of a previous intraocular hemorrhage. It is important to recognize that papilledema, unless very severe and prolonged, does not usually result in blurred vision or a significant change in visual field. Compromised visual acuity resulting from vitreous hemorrhage or its potential complications may become a serious problem for patients who survive SAH.

The prognosis for recovery of vision in patients with Terson’s syndrome has been reported to be good, and this was evidenced by the patients in this study. Patients should be followed closely for complications of intraocular hemorrhage, including elevated intraocular pressure and retinal membrane formation with resulting retinal detachment. Conservative therapy is advocated, since these hemorrhages usually clear spontaneously by 6 to 12 months. After this time, vitrectomy should be considered if progressive improvement in visual acuity is not demonstrated.

Clarkson, et al., treated with vitrectomy five patients with persistent vitreous hemorrhage secondary to Terson’s syndrome. They reported excellent postoperative visual acuity with no significant surgical complications. Vitrectomy resulted in a substantial improvement in the visual acuity of our one patient with persistent vitreous hemorrhage. Others have had a similar experience. Conclusions

The occurrence of intraocular hemorrhage as a direct result of SAH is not infrequent and represents a potentially serious complication in patients surviving SAH. Since the 1950’s, there have been several large retrospective studies showing that intraocular hemorrhage occurs in as many as 20% to 40% of patients with SAH, which is consistent with the frequency of 27% reported in our prospective series.

There is a need to look specifically for intraocular hemorrhage in patients with suspected SAH since its presence could aid in early diagnosis and guide later management. It is also important to recognize that intraocular hemorrhage has been linked to a poorer prognosis for the SAH patient, although this was not demonstrated in our study.

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A. M. Garfinkle, et al.
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