Terson syndrome with ipsilateral severe hemorrhagic retinopathy in a 7-month-old child

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In infants with intracranial hemorrhage, the most common cause of intraocular hemorrhages is abusive head trauma. Terson syndrome is rare in infants, and the retinal findings, although not well reported in the literature, are generally limited to the posterior pole. We report a case of a 7-month-old boy who developed ipsilateral, extensive preretinal and intraretinal hemorrhage after subarachnoid hemorrhage from a ruptured intracranial aneurysm.

Case Report

A previously healthy 7-month-old boy suddenly became floppy, unresponsive, and apneic at home. Cardiopulmonary resuscitation was initiated, and en route to the hospital, he had 2 generalized seizures. At the local hospital, his Glasgow coma score (GCS) fluctuated between 5 and 7. His anterior fontanelle was tense, and he was hypertensive and bradycardic, consistent with Cushing reflex. He was sedated and paralyzed for intubation; he received seizure prophylaxis with phenytoin and intravenous cefotaxime to prevent central nervous system infection.

Computed tomography (CT) of the brain showed right parieto-temporal subarachnoid hemorrhage with significant cerebral edema and 1–2 cm of midline shift. His initial recorded intracranial pressure was 50 mm Hg, which fluctuated up to 110 mm Hg. During the transfer to our facility, his intracranial pressure fluctuated between 20 and 30 mm Hg. His right pupil was transiently dilated to 4 mm during an intracranial pressure spike of 82 mm Hg, but this normalized after treatment with mannitol, hypertonic saline, fentanyl, and hyperventilation. All hematological parameters, including complete blood count and coagulation profiles, were within normal limits.

A CT angiogram (Figure 1) showed a complex fusiform aneurysm of the anterior division of the right middle cerebral artery with a large amount of acute subarachnoid blood extending anteriorly to the right temporal pole. There was also an intraparenchymal hematoma measuring 3 × 4 cm superior to the aneurysm and evidence of infarction of the posterior right frontal lobe.

During surgery, the aneurysm was clipped (Figure 2) and the Sylvian fissure hematoma evacuated. Intraoperatively, evidence of right subdural hematoma was discovered. The postoperative course was unstable, with persistently increased intracranial pressure; a ventriculoperitoneal shunt was inserted on day 19 as the result of hydrocephalus.

The patient's fluctuating neurological status did not allow for pharmacological mydriasis, which prevented an ophthalmological examination until 9 days after his initial presentation. On binocular indirect ophthalmoscopy at this stage, extensive preretinal and intraretinal hemorrhages were noted in the right eye (Figure 3); the left eye appeared normal.

Follow-up examination at 1 and 3 weeks showed improvement of the retinal hemorrhages. On stabilization of his neurological status, patching of the left eye was commenced for 1 hour daily. On follow-up 3 months later, the intraocular hemorrhages had all but resolved, apart from some mild pigmentation in the macula. There were no obvious signs of optic atrophy. A left homonymous hemianopia, consistent with cortical visual impairment, and a left hemiparesis were present. The patient had a right exotropia, and ocular rotations were full. Vision was worse in the right eye than the left eye, although he was able to fix and follow with both eyes.

Discussion

Terson syndrome is the clinical entity of intraocular hemorrhage in the setting of intracranial hemorrhage.[1] The authors of prospective studies[2,3] have reported that 17% to 46% of adults with subarachnoid hemorrhage develop Terson syndrome. Because of the low incidence of spontaneous intracranial hemorrhage in infants, Terson syndrome has not been well studied in this age group. In the only published case series of nonabuse instances of intracranial hemorrhage in children,[4] 1 of 57 children (2%) had intraocular hemorrhages; a 7-year-old with 5 intraretinal hemorrhages along the vascular arcades in one eye after a severe motor vehicle accident. The authors estimated the incidence of Terson syndrome in children with intracranial hemorrhage to be less than 8%.

There have been 2 case reports of intraocular hemorrhages in infants with intracranial hemorrhage from demonstrated vascular anomalies. McLellan and colleagues[5]

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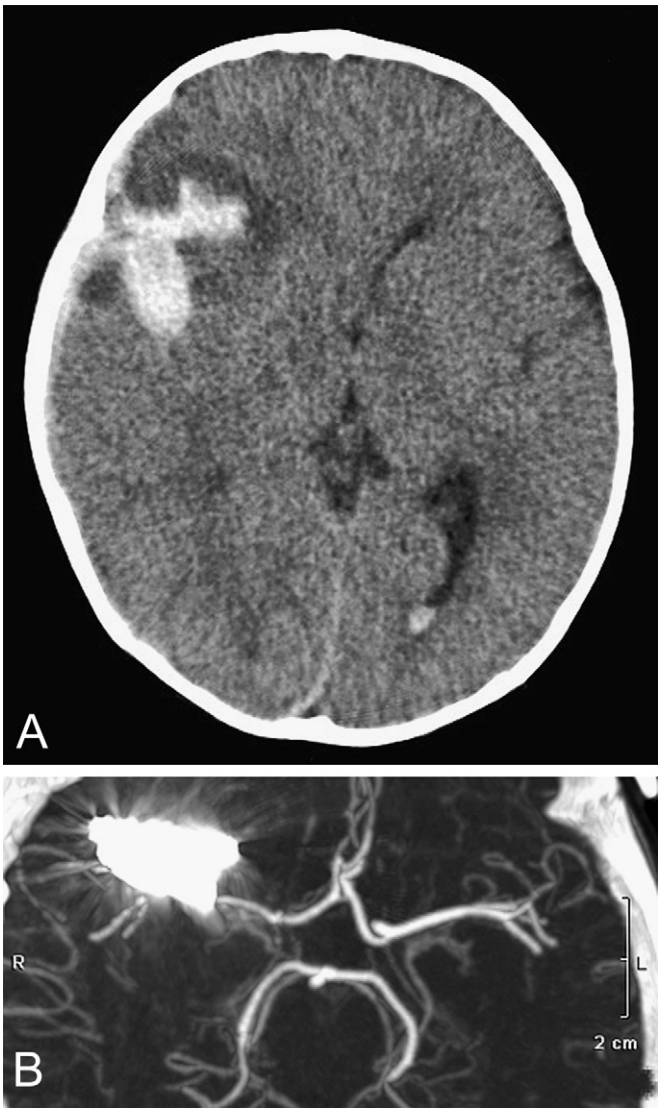

FIG 1. Preoperative noncontrast axial CT brain and CT angiogram of the circle of Willis. A, Acute subarachnoid hemorrhage centered on the right Sylvian fissure with probable areas of infarction in the posterior right frontal lobe. There is mass effect and midline shift to the left by 4 mm and effacement of the right lateral ventricle; a small amount of acute blood is visible in the posterior horn of the left lateral ventricle. B, Reconstruction: a large complex aneurysm arising from the anterior branch of the right middle cerebral artery distal to the bifurcation of the M1 segment.

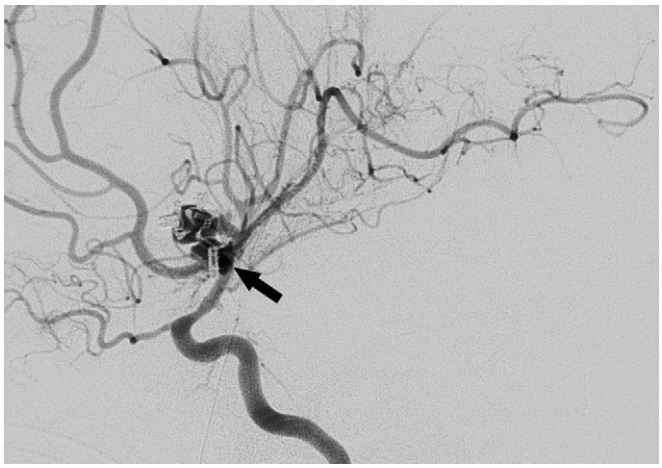

FIG 2. Digital subtraction angiogram of the right internal carotid artery obtained 7 days after neurosurgery. A 5- to 6-mm irregular aneurysm is visible at the middle cerebral artery bifurcation with aneurysm clips in place (arrow).

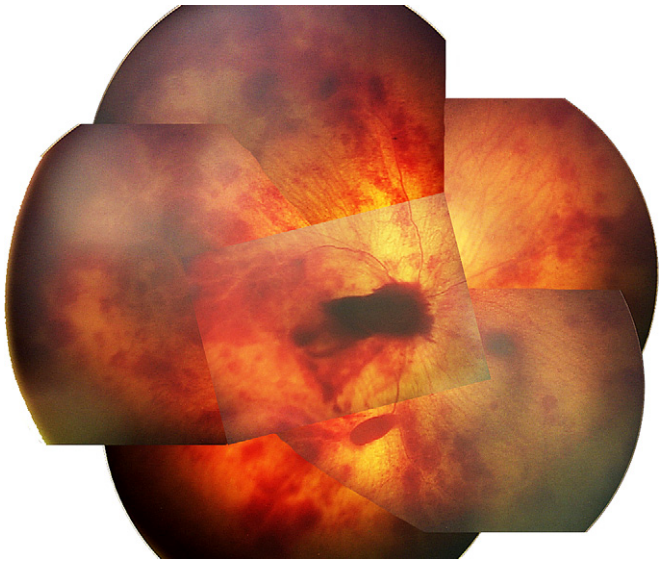

FIG 3. Montage fundus photographs of the right eye showing extensive preretinal and intraretinal hemorrhages radiating toward the periphery.

reported a 6-week-old girl with an intracerebral hematoma attributable to a ruptured aneurysm in the anterior branch of the right middle cerebral artery. The intraocular hemorrhages were described as "extensive bilateral retinal hemorrhages" with a "large right subhyaloid hemorrhage," although photographic documentation was not provided. Reddy and colleagues[6] reported a 5-week-old girl with mixed subarachnoid and subdural blood overlying the right cerebellar hemisphere due to a ruptured angiodysplasia. This girl had ipsilateral intraretinal hemorrhages in the posterior pole of the right eye not extending to the periphery.

The pathogenesis of Terson syndrome remains controversial.[2,7] It is unclear whether blood in the retrobulbar optic nerve sheath directly enters the subhyaloid space[7] or whether intraocular hemorrhage is a result of obstructed retinal venous and retinochoroidal outflow from raised intracranial pressure and dilatation of the optic nerve sheath.[8]

Spontaneous Terson syndrome is one of the differential diagnoses for intraocular hemorrhages in infants with suspected abusive head trauma. Distinguishing characteristics,

which have been suggested by Levin,[9] are the mild severity and confinement of retinal findings to the posterior pole in spontaneous Terson syndrome, which are in contrast to the usually severe, multilayered intraocular hemorrhages extending to the retinal periphery in abusive head trauma. A recent review article also stated that Terson syndrome "in the sense of major retinal hemorrhages of the type seen in inflicted traumatic brain injury has not yet been reported in the pediatric literature."[1]

In our patient, the obvious appearance of a ruptured aneurysm on CT scan confirmed the diagnosis. Our case photographically demonstrates the possibility of a severe hemorrhagic retinopathy from a nontraumatic cause and confirms the existence of Terson syndrome in infants.

Literature Search

PubMed was searched (1970 to present) for the following terms: *Terson's syndrome* OR *subarachnoid hemorrhage* and *retinal hemorrhage*. The same terms were also searched in MEDLINE (1950-present), EMBASE (1947-present), and Evidence-Based Medicine Reviews simultaneously via OVID. Articles cited in the reference lists of other articles were also searched.

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