Cerebral angiography showed severe bilateral stenosis at the distal internal carotid artery and basal moyamoya vessels. It was compatible with moyamoya disease (Fig. 2). Emergent extra-ventricular drainage was performed. His mentality gradually improved to alert. Ophthalmologic examination at our hospital detected a vitreous hemorrhage in his left eye (Fig. 3B), with right eye remaining normal (Fig. 3A). In spite of three months conservative care, his visual acuity was not improved. Vitrectomy with epiretinal membrane removal was performed. After operation his left visual acuity was recovered. Careful ophthalmologic examination is mandatory in patients with hemorrhagic moyamoya disease.

Key Words: Moyamoya disease · Terson syndrome · Intraventricular hemorrhage.
The occurrence of Terson hemorrhage has been described in cases of moyamoya disease, severe closed-head injuries, intracerebral hemorrhage, carotid artery occlusion, cranial subdural hematoma, lumbosacral myelomeningocele, and as a complication associated with intraarterial angiography.

The present case shows that IVH secondary to moyamoya disease can induce an abrupt increase in ICP resulting in Terson hemorrhage. If the only reason of Terson syndrome is venous congestion due to IICP, equal incidence of Terson hemorrhage as seen in SAH should be present in the other IICP conditions. But, the other IICP conditions do not show the same incidence of SAH associated Terson hemorrhage. Therefore, the other factors may be associated with the development of Terson hemorrhage.

Patients with moyamoya disease can exhibit various ocular symptoms which are mainly caused by stenotic and occlusive lesions in the occipital visual cortex or optic pathways or tract, but have relatively normal intraocular findings.

Careful ophthalmological examination including meticulous fundoscopic evaluation remains the method of choice for detecting Terson hemorrhage. The ideal timing for performing an ophthalmological examination remains to be defined. Manischot reported that all Terson hemorrhages were present within one hour from the ictal event. However, delayed Terson hemorrhage might occur up to 47 days after the ictus.

Swallow et al. evaluated the potential of CT scans of the orbits for detecting Terson hemorrhage. They found that in the majority (66.7%) of patients with Terson hemorrhage, characteristic retinal nodularity and retinal crescentic hyperdensities were evident on their CT scans. However, in our case we could not see the evidence of retinal or orbital hemorrhage on initial CT scans.

The visual outcome was excellent in the majority of cases of Terson hemorrhage. In rare instances, however, Terson hemorrhage can be associated with the development of proliferative retinopathy, retinal breaks, retinal detachment, and cataract. Surgical intervention is reserved for those cases in which there is no visual acuity improvement within 6 months from the ictal event. The surgical treatment of choice, when indicated, is pars plana vitrectomy.

Moyamoya disease may accompany various ocular symptoms such as visual field defect, amaurosis fugax, decreased visual
acuity and scintillating scotoma. Majority of these visual symptoms are usually result from ischemic lesions in the visual cortex and optic pathways.

Terson syndrome can be a rare cause of visual disturbance of patients with hemorrhagic moyamoya disease. Careful ophthalmologic examination is mandatory in these patients.

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