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Case Report

Terson Syndrome: Current View Points

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ABSTRACT

Terson syndrome is characterized by intraocular hemorrhage combined with any form of intracranial hemorrhage and sharp increase of intracranial pressure. However, most patients are first diagnosed in neurology department. Terson syndrome is often neglected due to lack of relevant professional knowledge. This study discusses the current cognition of pathophysiological mechanisms, clinical presentation, management and prognosis of Terson syndrome.

Keywords: Terson syndrome; Intracranial pressure; Intraocular hemorrhage; Surgery

INTRODUCTION

In 1900, a French ophthalmologist Albert Terson first reported Subarachnoid Hemorrhage (SAH) related Vitreous Hemorrhage (VH). Since then, the disease has been named Terson. With further study, Terson syndrome is not limited to VH secondary to SAH. At present, the definition of Terson syndrome usually reflects an intraocular hemorrhage secondary to a rapidly elevated intracranial pressure, including any types of SAH, traumatic brain injury or intracerebral hemorrhage combined with vitreous, sub-hyaloid, sub-retinal, intraretinal and/or preretinal hemorrhage [1,2]. The risk of developing Terson syndrome is associated with an increased clinical and radiological severity of aneurysmal SAH, a low Glasgow Coma Scale, high Hunt and Hess grade, and high Fisher grade [2]. In particular, Terson syndrome is not uncommon; the prevalence rate is reported 8%-44% [3]. Aneurysmal SAH is the most common cause of Terson syndrome; moreover, the latter can lead to temporary vision loss or even permanent blindness if severe. Some studies have shown that Terson syndrome occurs more frequently in anterior circulation aneurysms, especially in anterior cerebral artery complex aneurysms, which are 10 times as many as other aneurysms [4]. Aneurysm size has also been related to the fundus abnormality and the occurence of Terson syndrome [5]. Larger aneurysms are associated with a greater risk for developing Terson Syndrome. In a study of 34 patients with Terson Syndrome, 30 had a dome diameter no less than 5 mm, while only 4 had a dome diameter less than 5 mm [3]. In addition, although Terson Syndrome can occur in children, it usually happens in adults.

Pathophysiological mechanisms

Although it has been more than 120 years since the first description, the exact pathophysiology of intraocular hemorrhage in Terson syndrome remains controversial. Different theories have been proposed to explain its pathogenesis, including trans lamina cribrosa theory, peripapillary leak theory, perivascular leak theory, primary vitreous origin, retinal venous congestion theory and so on [6].

At present, there are mainly two accepted possible pathophysiological mechanisms. The optic nerve is covered by the optic nerve sheath, which comes from the three-layer meninges. The cerebrospinal fluid flows freely between the intracranial and orbital subarachnoid space. Therefore, the intracranial and orbital subarachnoid space will be affected by the same pressure changes [7]. One hypothesis is proposed based on these features that the scleral cribriform plate connects peripapillary subdural space with intracranial subarachnoid space, maintaining pressure gradient on both sides. Under physiological conditions, SAH will not enter eye directly. Since silicone oil was used as intraocular tamponade after vitrectomy, it was occasionally reported that silicone oil was found in subarachnoid space or even ventricle, indicating that intracranial hemorrhage might enter eye under pathological conditions [8].

An alternative hypothesis is that SAH causes sudden increase of intracranial pressure, spreading to interorbital space of optic nerve, resulting in central retinal vein oppression, venous circumfluence obstruction, elevated intraocular vein pressure, vascular rupture and bleeding [9]. In the process of Terson

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syndrome, these two mechanisms may be reasonable and complement each other. Czorlich et al. reported that the incidence of Terson syndrome was associated with elevated intracranial pressure and an intracranial pressure of P25 mmHg was the only independent predictor for Terson syndrome in the multivariate analysis [10].

Recently, Kumaria et al. [11] put forward another novel hypothesis: The glymphatic reflux theory. Under physiological conditions, ocular glymphatic flow terminates in meningeal lymphatics. When intracranial pressure exceeds intraocular pressure, subarachnoid blood in skull base cisterns enters the lamina cribrosa through paravascular glymphatic channels to cause Terson Syndrome.

CASE PRESENTATION

Patients present with different clinical manifestations, depending on the degree of intraocular hemorrhage and previous neurological status [12]. The ocular manifestations may be accompanied by symptoms of increased intracranial pressure, such as headache with or without nausea and vomiting, loss of consciousness or coma. The history of loss of consciousness associated with headache episodes of ruptured cerebral aneurysms sensitively identifies 100% of VH related to Terson syndrome [13].

Visual impairment can occur two weeks after the onset of SAH at the earliest and it has also been documented to appear before SAH [12]. Patients with Terson syndrome present with unilateral or bilateral ocular hemorrhage of asymmetrical nature [5]. Depending on the amount of intraocular hemorrhage, there may be different degrees of visual impairment. If there is only a small amount of retinal hemorrhage, the visual acuity will not decline significantly. If the hemorrhage is located in the macular area or a large amount of hemorrhage enters the vitreous cavity, the visual acuity will decline sharply. The degree of intraocular hemorrhage is related to the speed of intracranial hemorrhage and whether there is cerebral edema. VH can occur at the same time as SAH or later, some patients develop VH two weeks after the occurrence of SAH. Some hemorrhage can accumulate under the internal limiting membrane without entering the vitreous body. Some VH occurs in the rebleeding of the brain. VH is the diffuse red turbidity firstly at the posterior pole, and the peripheral retina can be seen clearly in some patients. In patients with chronic VH of Terson syndrome, VH provides an opportunity for retinal glial cells to proliferate and promote the formation of an organized membrane. The hemorrhage also contains cell growth factors, which also promote the migration and proliferation of cells, and ultimately organized traction will be formed, causing retinal detachment.

Necessary laboratory examinations should be carried out according to different causes of intracranial hemorrhage to determine the initiating factors of intravitreal hematocele. The location and scope of intracranial hemorrhage can be determined by cranial Computed Tomography (CT) scan (Figure 1) and Magnetic Resonance Imaging (MRI), and the amount of hemorrhage can be estimated to determine the cause.

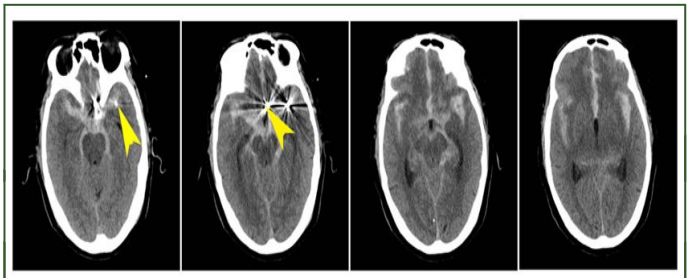

Figure 1: CT scan of a 63-year-old man on the day of second onset of subarachnoid hemorrhage. The yellow arrow heads indicates the coil used in the operation for the first subarachnoid hemorrhage.

Considering the prognostic impact of Terson syndrome on morbidity and mortality, it is recommended that all coma SAH patients undergo ophthalmic examination upon admission [9]. About 4% of Terson syndrome patients may have VH within one hour to several days after SAH [12]. Funduscopy is the gold standard for diagnosing Terson syndrome. However, when the fundus view is obscured, ophthalmic B-ultrasound examination (Figure 2) can be used to determine VH, posterior detachment and whether there is retinal detachment.

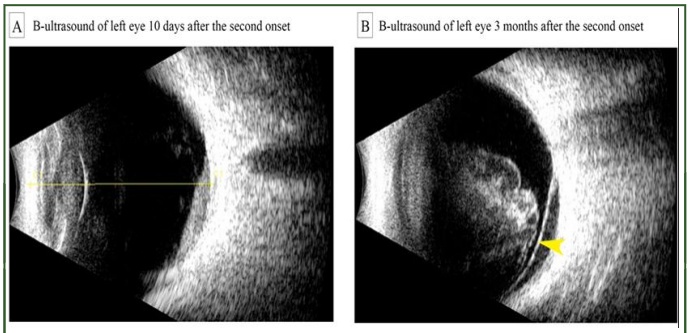

Figure 2: A 63-year-old man suffered from a second subarachnoid haemorrhage 10 days later, (A) B-ultrasound showed vitreous and preretinal haemorrhage of left eye; (B) 3 months later, B-ultrasound showed massive vitreous haemorrhage and limited retinal detachment (yellow arrow head) of left eye.

Management

The treatments of Terson syndrome include conservative ophthalmic treatment by observation, surgery and other treatment methods. For patients with slight unilateral hemorrhages, unsuitable for surgery and in the absence of other complication threatening vision, conservative treatment is recommended through observation. The resorption rate of intraocular hemorrhage depends on the severity, and which is slow for large, dense hemorrhages. Though a majority of patients with VH resolves spontaneously 6-12 weeks after SAH [14], some usually takes several months, even up to about a year. The absorption of accumulated blood is generally from the periphery to the center. The vision of patients can often return to normal after that. The patients with less hemorrhage can wait for self-absorption, and the prognosis is better. These methods include mobilizing patients to stand upright and avoiding the use of anticoagulants (aspirin and NSAIDs), because which may increase bleeding at the pathological site [6]. The existing

literature shows that although surgery speeds up the process of vision recovery, the results of conservative treatment are similar to those of surgery [15].

However, in some cases, the hemorrhage removal may take several years or too massive to absorb by itself. If not treated in time, VH can cause deposition of blood components and inflammatory cells, fibrous proliferation, epiretinal membrane formation, and eventually traction retinal detachment development. In addition, the disintegration products of blood components may have long-term toxic effects on retina with vision loss or serious vision damage. Therefore, vitrectomy should be performed in patients without signs of spontaneous hemorrhage absorption. The surgical treatment of pars plana vitrectomy is mainly applicable to bilateral dense intraocular hemorrhage that cannot be resolved for a long time (4-6 months), prevent amblyopia in children, as well as in instances where there is concurrent hemorrhage with intraocular complications, such as retinal detachment [16]. Many studies have confirmed that patients with a course of less than 3 months have better postoperative visual acuity than that of more than 3 months [17]. Although some studies demonstrated no significant difference for visual acuity in patients with VH underwent surgery within 3 months compared with those who underwent surgery after 3 months [18,19], minimizing the latency before surgery and timely vitrectomy can significantly reduce intra and postoperative complications [19,20]. Therefore, most scholars advocate early surgery, at least for intraocular hemorrhage less than 6 months [9,10].

For patients with failed conservative treatment and high surgical risk, other treatment methods have been tried, such as minimally invasive laser hyaloidotomy (Nd: YAG (neodymium-doped yttrium–aluminium–garnet) laser posterior hyaloidotomy) and intravitreal injection therapies (sulfur hexafluoride gas and tissue plasminogen activator) [6].

RESULTS

There is a report collected 320 patients with SAH and VH. The results revealed that the fatality rate was 53.6%, while that of patients without VH was only 19.7%. If there is bilateral VH, the mortality is higher. If the patients with intracranial hemorrhage can survive, the retinal and pre-retinal hemorrhage can generally be absorbed without leaving obvious sequelae, but a few of them will cause permanent visual damage. Early detection and timely vitrectomy when necessary can improve the prognosis.

DISCUSSION AND CONCLUSION

Terson syndrome, an uncommon eye-brain syndrome, may cause severe vision loss if not diagnosed and treated early. Terson syndrome has many different pathogenesis theories, but the most recognized one is acutely raised intracranial pressure. With the deepening of research, new hypotheses are occasionally put forward. Considering that the neuropsychiatric manifestations of most patients with Terson syndrome are too serious to express visual symptoms when they are admitted to hospital, it is easy to underdiagnosis. Thus, it is recommended that all SAH patients should be conducted routine ophthalmic examination when they are on hospital admission, as well as those with acute intracranial hypertension or visual impairment of unknown cause. Furthermore, it is crucial to make a clear diagnosis by combining clinical manifestations, fundoscopy and imaging clues. The spontaneous absorption of intraocular hemorrhage should be observed for at least 6-12 weeks. If the intraocular hemorrhage persists for more than 3 months and cannot be absorbed, prompt vitrectomy should be performed. Therefore, early diagnosis and treatment of Terson syndrome should be concerned by both neurologists and ophthalmologists.

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