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CASUISTIC PAPER

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Traumatic "TERSON SYNDROME PLUS": Pneumocephalocele with optic atrophy

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ABSTRACT

Introduction. Terson Syndrome is subarachnoid hemorrhage (SAH) with sub retinal hemorrhage flowing through channel. Reduced vision in such fresh case is due to hemorrhage itself, blocking macula/other photo receptors in the long run macular cellophane retinopathy which causes profound visual loss. SAH causes neurological problems which can become a risk factor for evacuating blood from vitreous. Hypertension is commonest cause to cause Terson Syndrome, but trauma is also devastating cause as it can lead to irreversible visual consequences like total loss of perception of light or blindness.

Aim. Here we describe a case of Terson Syndrome plus disease features SAH in frontal lobe.

Description of the case. When there is traumatic pneumocephalocele, it gives space to blood to imbibe towards bony optic canal and form hematoma around nerve sheath which causes compression around the same and leads to optic atrophy. Optic nerve can be injured by direct traumatic dissection during road traffic accidents (RTA), but even without that blood may accumulate around optic nerve and in turn leads to formation of hematoma and subsequently pressure induced optic atrophy. Moreover, blood can slowly travel to sub hyaloid space/sub retinal space (beneath internal limiting membrane or sub ILM) with probable gliosis covering typical boat shaped blood as seen in this case. This sub ILM hemorrhage or gliosis may have resolved through three injections of Triamcinolone in the orbital floor (OFTA) near apex, but optic atrophy snatches vision. This protocol was followed to treat traumatic compressive (peri optic hematoma) optic neuropathy and traumatic retinopathy associated with sub hyaloid hemorrhage.

Conclusion. Diagnosis of Terson syndrome plus disease was established by addressing all features on computed tomography (CT) scan and magnetic resonance imaging (MRI). Plus, features include pneumocephalus, optic nerve sheath hematoma, optic atrophy and gliosis over sub-hyaloid hemorrhage, typical boat shaped. The part of hemorrhage still endured as seen on optical coherence topography, but vision was lost by virtue of optic atrophy. OCT shows clot in sub hyaloid space.

Keywords. optic atrophy, sub retinal, subarachnoid hemorrhage, sub hyaloid, Terson syndrome plus

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Introduction

First described by Litten in 1881 and then in 1900 by French ophthalmologist Albert Terson.[1,2] Terson syndrome is now recognized as intraocular haemorrhage associated with subarachnoid haemorrhage (SAH), intra-cerebral haemorrhage, or traumatic brain injury.[1] Haemorrhage may be present in the vitreous, sub hyaloid, or intraretinal/sub-internal limiting membrane.

Several possible pathophysiologic mechanisms for Terson syndrome are known. Blood present in subarachnoid may be directly transmitted forward through the optic nerve sheath.[1,3] More commonly, a sudden increase in intracranial pressure leads to rapid effusion of CSF into the optic nerve sheath which causes dilatation of the retro bulbar optic nerve mechanically compressing central retinal vein and ensuing venous hypertension results in rupture of thin retinal vessels. This mechanism is consistent with the fact that Terson syndrome can be seen in patients without intracranial haemorrhage.[4]

Aim

Here we describe a case of Terson Syndrome plus disease features SAH in frontal lobe.

Description of the case

A 38-year-old male came to our hospital with history of road traffic accident (RTA) 4 months back, he sustained head injury and was treated outside for brachial plexopathy. After recovering from head injury components, the patient presented with total loss of vision in left eye. On examination his best corrected visual acuity (BCVA) was 6/6 in right eye and no perception of light (PL negative) in left eye. Anterior segment was within normal limits. Relative afferent papillary defect (RAPD) was present in left eye. Fundus examination showed white boat shaped dirty white sub hyaloid lesion involving macula with disc pallor (Figure 1).

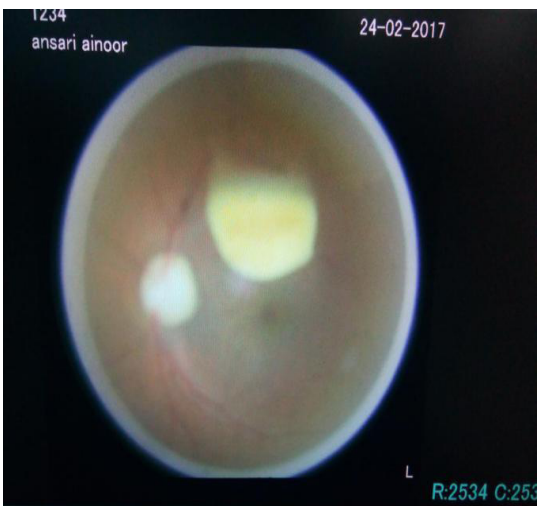

Fig. 1. Initial presentation showing optic atrophy with peri macular gliosis

Extra ocular movements were free and full in both eyes. Initial computed tomography (CT) of head showed contusion involving left frontal region with perilesional edema with subarachnoid haemorrhage in basal cisterna (Figure 2). MRI brain showed normal study except thickening of optic nerve in left side and haemorrhage around optic nerve (Figure 3).

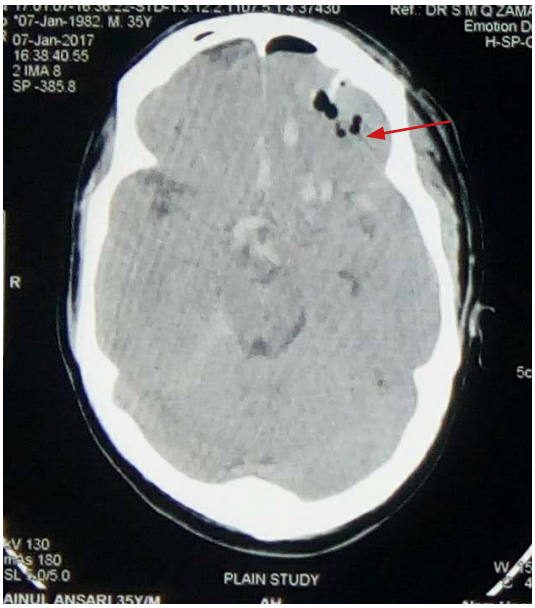

Fig. 2. CT scan of brain showing subarachnoid haemorrhage and pneumocephalus in frontal lobe – black arrow shows subarachnoid haemorrhage, blue arrow shows pneumocephalus in frontal lobe

Presence of RAPD with MRI-proven perineural haematoma confirmed traumatic optic neuropathy. Moreover, sub hyaloid gliosis over macular area was the indication for OFTA. Three injections on monthly interval have resolved gliosis exposing a faded sub hyloid haemorrhage (Figure 4). OCT findings ruled out sub ILM haemorrhage (Figure 5). On subsequent follow up fundus photography, the haemorrhage resolved but the patient's visual acuity did not improve

Key points of the case:

1. Frontal lobe haemorrhage leading to haematoma near optic nerve
2. Retinal Haemorrhage with gliosis
3. Gliosis resolved with periocular steroid
4. No mortality even with SAH and Terson syndrome
5. Optic nerve atrophy due to pressure of optic nerve haematoma leading to high ocular morbidity

Discussion

When vitreous hemorrhage occurs in association with subarachnoid hemorrhage is known as Terson syndrome. Intraocular hemorrhages of any type, call it retinal, sub hyaloid, or vitreous all of these have been documented in 10–40% of individuals with subarach-

noid hemorrhage. It can also occur in association with intracranial hemorrhage and elevated intracranial pressure. Diagnosis of Terson syndrome is clinically important, as it is associated with significantly higher mortality. Up to 77% of these cases are overlooked on daily reports. Timely ophthalmologic intervention to prevent long term visual loss is of utmost importance.

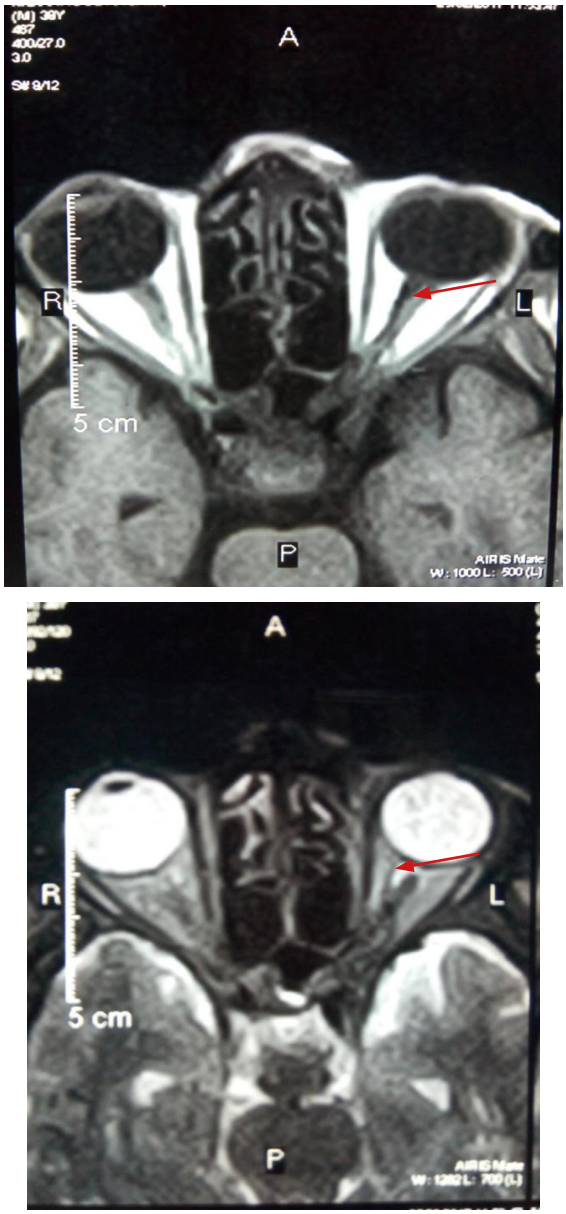

Fig. 3. T1 and T2 weighted images showing hematoma near optic nerve in left eye (blue and black arrows)

Fluorescein angiography has demonstrated a leakage site at the disc margin in a patient with Terson syndrome with vitreous haemorrhage. This theorizes potential damage to the peripapillary retina induced by increased intracranial pressure transmitted through the optic nerve sheath.[4]

Terson syndrome can present with dome-shaped haemorrhages in the macula.[5] A macular "double ring" sign may be seen with the inner ring caused sub-ILM haemorrhage and the outer ring caused by sub-hyaloid haemorrhage.[6]

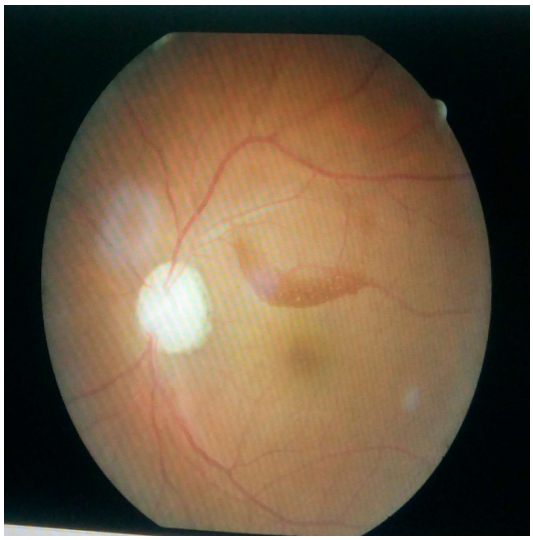

Fig. 4. Treatment with peri ocular steroid resulting in disappearance of gliosis exposing retinal haemorrhage

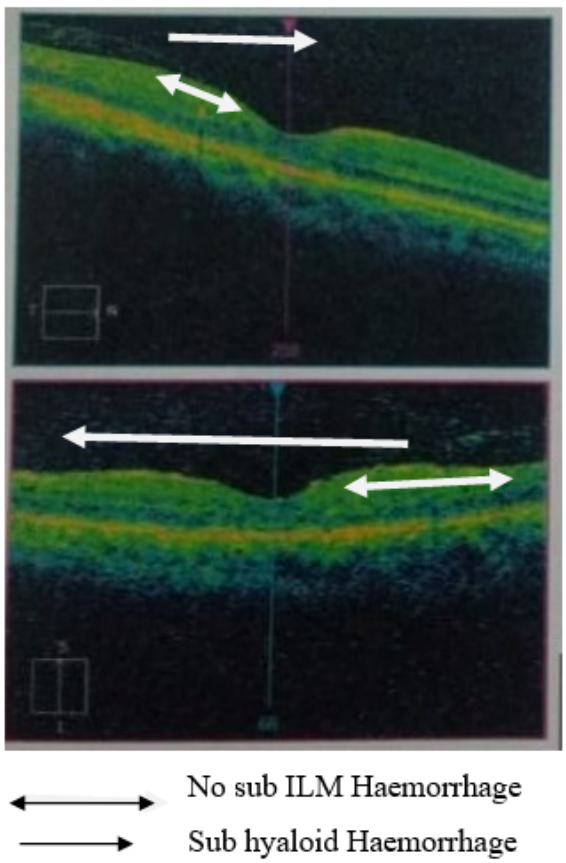

Fig. 5. OCT image showing sub hyaloid haemorrhage

Even if intraocular haemorrhages most frequently develop in the first hour after SAH, Terson syndrome can have a delayed onset, with reports of intraocular haemorrhage occurring up to 47 days after SAH.[1,7,8]

Low Glasgow coma scale, high Hunt and Hesse grade, and high Fisher grade are associated with a higher incidence of Terson syndrome.[1]

Neurological outcomes and mortality rate are worse in patients with SAH and Terson syndrome than patients with SAH alone.[1] In a study by Pfausler, mortality was 90% in patients with SAH and Terson syndrome and 10% in those with SAH without Terson syndrome.[9-11]

Swallow investigated the use of orbital CT to identify intraocular haemorrhage in patients with Terson syndrome. There was presence of retinal crescentic hyper densities and retinal nodularity in CT in two-thirds of patients with Terson syndrome.[12-14] Thus CT may be useful to identify possible Terson syndrome prior to an eye exam.

Multiple complications have been reported after Terson syndrome. Epiretinal membrane is the most common sequel of Terson syndrome, with an incidence of 15-78%.[15-18] Vitreous blood may cause ERMs by inducing glial proliferation and disruption of the ILM.[19]

In 20% of patients with Terson syndrome retinal folds/perimacular folds occur. Also, occurrence of retinal detachment in 9% and ghost cell glaucoma in around 4% cases has been seen reported.[19] Proliferative vitreoretinopathy and preretinal fibrosis have also been reported after Terson syndrome.

Studies have shown no significant difference in final visual acuity between patients who were conservatively managed and those who underwent PPV. However, visual recovery was more rapid in the vitrectomy group despite these patients having denser vitreous haemorrhage.[17]

Role of systemic and periocular steroid is the mainstay of to prevent visual loss in neuro ophthalmology cases but use of OFTA proved beneficial in this case.[20] So this approach may be tried as it is very safe, non-oculo-hypertensive and without any systemic side effects.[21] However this needs more studies in future.

Conclusion

Terson Syndrome is a known entity particularly in cases of brain haemorrhage due to hypertensive crisis and haemorrhagic stroke. Traumatic subarachnoid haemorrhage with sub hyaloid/sub retinal haemorrhage is rare. Several possible path physiologic mechanisms for Terson syndrome are well known, but it is always good to assess any disease from different perspective, as minute finding can be vital for new outcome. So the case of "Terson Syndrome plus" a new clinical entity discussed here is having signs of initial sub arachnoid hemorrhage in frontal lobe along with pnemoencephalocele with hematoma in optic canal and sub hyaloid haemorrhage. Essentially Terson syndrome plus is the combination of multiple features, namely Subarachnoid haemorrhage in frontal lobe with pneumoencephalocele, optic canal haematoma sub hyloid/sub retinal haemorrhage and subsequent blindness due to pressure atrophy of optic nerve.

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