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SNI: Neurovascular

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Case Report

Atypical Terson syndrome after subarachnoid hemorrhage from middle cerebral artery aneurysm rupture during coitus

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Received: 12 April 2024
Accepted: 15 July 2024
Published: 23 August 2024

DOI
10.25259/SNI_287_2024

Quick Response Code:

ABSTRACT

Background: Terson syndrome (TS) is a neuro-ophthalmologic disease arising due to subarachnoid hemorrhage (SAH), resulting in the formation of subhyaloid hemorrhagic spots. These spots can affect the ability to see due to the alteration of the optic cameras. Although it often affects both eyes, the symptoms and the eye involvement can be asymmetrical in rare cases.

Case Description: We described the case of a 52-year-old female patient who developed Terson disease following the rupture of a right middle cerebral artery aneurysm occurring during coitus with SAH (Fisher grade III). The aneurysm was treated by endovascular coiling. Interestingly, despite the major involvement of the right eye, the patient primarily manifested symptoms of visual changes in the left eye.

Conclusion: TS is a frequent ocular complication of SAH, with symptoms typically affecting both eyes. Characterized by hemorrhagic spots in both subhyaloid layers, the syndrome's symptomatology is generally bilateral. However, in the case described, the manifestation is deemed atypical, primarily appearing contralateral to the hemisphere exhibiting a greater pattern of SAH.

Keywords: Terson syndrome, Subarachnoid hemorrhage, Middle cerebral artery aneurysms, Intracranial hypertension, Subhyaloid hemorrhage

INTRODUCTION

Terson syndrome (TS) is a disease of both ophthalmic and neurologic interest. It is characterized by intraocular hemorrhage (IOH), and it appears in 15–50% of patients with subarachnoid hemorrhage (SAH).[10] The presence of blood collection in the vitreous body was first described in 1881 by German Physician Moritz Litten. The French Ophthalmologist Albert Terson was the first one who documented a strict association between vitreous hemorrhage and SAH.[10] In subsequent studies, it was highlighted that bleeding can also involve peri-retinal and even subhyaloidal regions.

Although it is most evident in aneurysmal SAHs, TS may also be an occasional finding in post-traumatic SAH, Moyamoya disease, or thrombosis of dural venous sinuses.[18] Treatment of TS can be conservative or surgical. If not treated promptly, in cases where no spontaneous clot re-absorption occurs, it can result in photoreceptor degeneration. Although the exact etiology is still being investigated, the main pathophysiological mechanisms are probably linked to impaired retinal venous drainage or glymphatic theories.[6] In general, TS manifests within hours from SAH, but it can also be delayed for weeks. Moreover, there are TS atypical forms, described as cases where visual symptoms precede neurological symptoms.[25]

CASE DESCRIPTION

A 52-year-old female patient presented with a prior history of autoimmune dermatitis and hypertension. During sex, she suffered a headache and blurred vision, followed by loss of consciousness. After regaining consciousness, she vomited and continued to complain about visual alterations with a stronger headache.

After her admission to our emergency room, she underwent a brain computed tomography scan, documenting diffuse Fisher type 3 SAH [Figure 1]. An angiographic study documented the presence of a ruptured saccular aneurysm located at the right middle cerebral artery (MCA) bifurcation, with a diameter of 6 × 4 mm, whose dome pointed antero-caudally, with an irregular bilobed morphology. Considering morphological ratios and the size of a neck <3 mm, aneurysmal sac was angiographically embolized through three Guglielmini coils (Target 360° model) [Figure 2].

After the procedure, the patient was admitted to our hospital. Due to the persistence of visual alterations, the patient underwent a first ophthalmologic examination. Tropicamide 1% was instilled, and through binocular indirect ophthalmoscopy, subhyaloid hemorrhage was detected in both the eyes in the macular region (bigger in the right eye than the left one) and vitreous hemorrhage in the left eye [Figure 3]. Moreover, the right eye showed optic disk swelling.

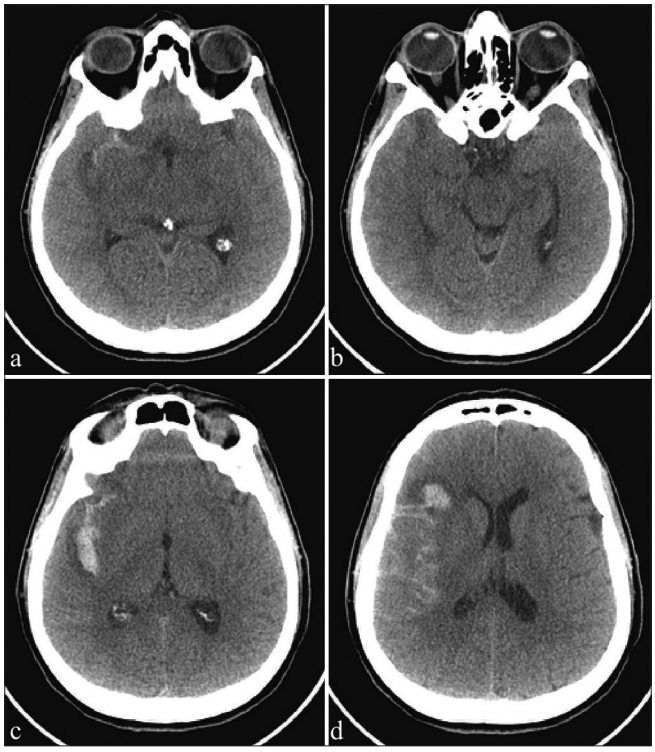

Figure 1: (a,c,d) Brain CT scan showing subarachnoid hemorrhage (SAH) due to middle cerebral artery (MCA) aneurysm rupture. (b) Note that basal cysterns are almost blood-free and SAH is highly localized in right hemisphere while Terson syndrome is in the left eye.

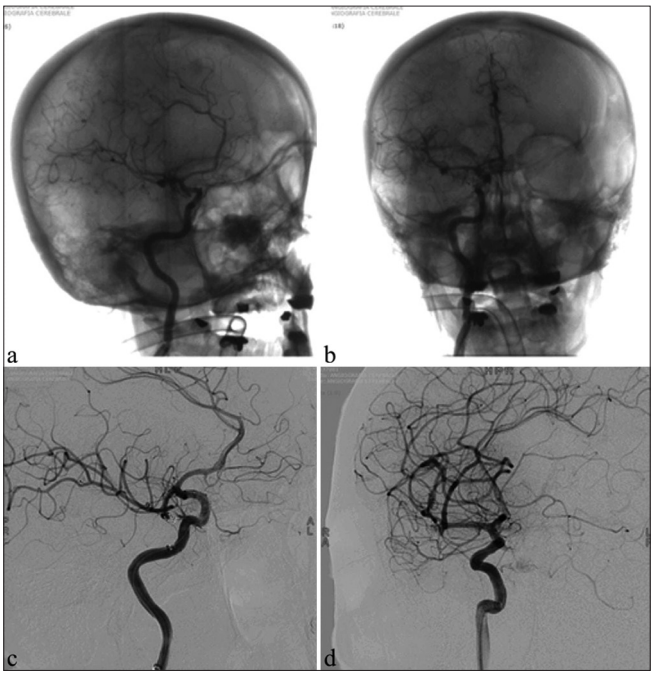

Figure 2: (a,b) Brain Angiogram showing middle cerebral artery (MCA) aneurysm (c,d) Brain Angiogram showing the correct endovascular treatment

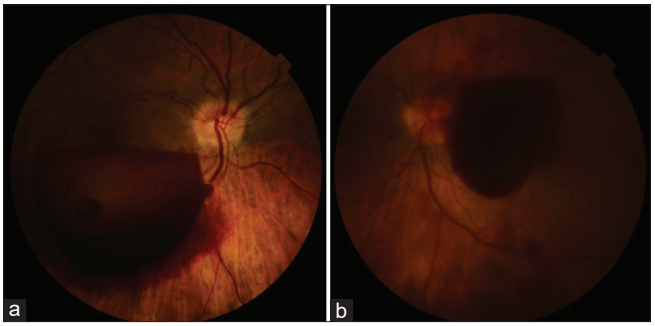

Figure 3: (a) Right eye: Retrohyaloid hemorrhage covering part of the macula and mild papilledema (optic disk swelling). The patient complained of decreased vision. (b) Left eye: Retrohyaloid hemorrhage covering the fovea and vitreous hemorrhage. The patient complained of blurred and decreased vision.

Medical therapy with tranexamic acid was prescribed. 1 week later, the patient referred an improved of her vision, that was less blurred. A second examination was performed. Her best-corrected visual acuity was 2/10 on the right eye and 10/10 on the left one. The last ocular examination showed the presence of hyper-refractive material beneath the hyaloid, which masked part of the posterior pole in the right eye and just a little part of the macula on the left one. Besides, on the right eye, ocular tests detected the presence of hyper-refractive fog over the fovea. This could explain the reason why the patient complained about right blurred vision, even if TS involved both eyes and vitreous hemorrhage was found only in the left eye.

DISCUSSION

TS is a rare ophthalmologic disorder characterized by vitreoretinal hemorrhage secondary to blood expansion into the subarachnoid space and rapid intracranial pressure (ICP).[10] It is also seen in cases of intraparenchymal hemorrhage or severe head trauma, even in some subdural or extradural hematomas.[4,13] Older hypotheses postulated a blood overflow directly into the eyeball through the optic nerve sheaths due to increased arachnoid pressure, but eye microanatomy studies established the absence of direct communication between the subarachnoid space and the vitreous cavity.[6] One of the most vetted hypotheses is that abrupt change in pressure within the subarachnoid space could result in abnormal vascular outflow in the retinal region with the development of ischemic and hemorrhagic damages extending to the vitreous chamber.[6,17,18] This pathology was thought to be more common in SAH from the rupture of anterior communicating artery aneurysms, but according to Pfausler *et al*., no direct correlation with aneurysm location was confirmed.[19] Several cases associated with rupture of MCA aneurysm have been described. Takeuchi described an exceptional case of TS following the rupture of a superior cerebellar artery aneurysm [2,30], while Inoue described a case of initial ophthalmic presentation in SAH by vertebral artery aneurysm rupture.[7]

During intercourse, physiological phenomena are known to result in a transient change of balance between sympathetic and parasympathetic systems. It leads to increased blood pressure that is typically transient in healthy subjects, but it can develop persistently in patients with long-lasting history of hypertension.[22,35] Low Glasgow coma scale score, high Hunt and Hess score, or high Fisher scores are negative prognostic factors.[12,23] The atypical manifestation of the reported case was the onset of visual alterations preceding neurological symptoms as well as the transient loss of consciousness without subsequent coma state. In fact, TS usually correlates with devastating intracerebral hemorrhage forms with high SAH scores, in which patients manifest important neurological deficits and lose consciousness for a long time up to coma. Subbiah *et al*. described an unusual TS case that preceded intracranial hypertension (IH) symptoms with gradual visual loss in a setting of flu-like pathology and loss of consciousness 3 weeks after severe intraventricular hemorrhage.[28] Murthy *et al*. described the case of TS caused by ruptured carotid-ophthalmic artery aneurysm SAH with late onset of headache and cervicalgia.[17] Raevis and Elmalem described a case of Terson-like syndrome in pseudotumor-cerebri regressed following treatment with acetazolamide.[21] Some sporadic and incidental forms of TS in epileptic patients have been noted among the different forms of Valsalva retinopathies since they are not associated with IH signs but with intracranial-intra-abdominal over-pressure during severe generalized seizures.[1,13,32] For instance, Mulholland and Page described a case of TS in a patient with an epilepsy history. The syndrome developed 3 days before the onset of meningeal and neurological signs, and SAH due to MCA aneurysm rupture was lately detected.[16] Compression of the hypothalamic-pituitary axis occurring during severe IH is typical in cases of cerebral herniation or midline shift.[9,27,31] It has been related to TS neurologic etiopathogenesis that possibly involves the activation of autonomic mechanisms for retrobulbar retinal vasoconstriction.[5,15]

Although in literature there is no strict relation between the SAH side and the eye affected by TS, it seems to occur bilaterally or unilaterally on the same aneurysm side.[19,21,25] In our case, the patient had a right MCA aneurysm, but she manifested left visual changes. Bilateral IOHs were evident on fundus oculi examination, but TS was symptomatic only in the left eye.

A possible explanation of our case is based on the new venous hypertension theory.[15,19] Although in SAH from MCA aneurysm, a predominant hyperdense pattern is more evident in the aneurysm side, the blood may extend into both hemispheres.[8] On considering the subarachnoid level, a not divided sector with a continuous flow of cerebrospinal fluid (CSF), the increased pressure is widespread to all compartments.[14,20,24] According to the recent theories of venous hypertension, TS could be caused by altered venous drainage in ocular veins due to raised ICP.[15] IH could force an abnormal CSF flow into the second cranial nerve or optic nerve sheath, causing its imbibition, and the venous hypertension would arise from mechanical compression by the dilated central retinal vein or the other choroidal veins.[13,33] In addition, the propagation of the pressure stress within the orbital cavity through the intervaginal space of the optic nerve sheath would be the cause of papillary edema, papillary damage, and visual impairment.[29] Papillary damage, and in some cases even macular atrophy, can be determined by the activity of reactive oxygen species that accumulate and activate during venous stasis, autoimmune

immune reactions in response to an infectious agent.[3,26,32,34] Lee *et al*. reported the first case of TS in the presence of atypical acute retinal necrosis in immunocompromised patient (presence of multiple antibodies for Herpes Simplex Virus, Varicella-Zoster virus, Toxoplasma, and Epstein-Barr), characterized by severe vaso-occlusive vasculitis determining panuveitis and vitritis, treated with intravitreal injections of ganciclovir and intravenous corticosteroids.[11] The patient in our report was treated with calcium channel blockers, vasodilators (nimodipine), and corticosteroids to ensure careful normovolemia and normotension. Moreover, as reported in the literature as the first choice treatment of TS, intravenous tranexamic acid was administered for 14 days.

CONCLUSION

TS is a rare condition characterized by hemovitreous extending even into the peri-retinal region and is associated with neurosurgical hemorrhagic disorders such as SAH from ruptured cerebral aneurysm. The exact etiopathogenesis is doubtful, but processes related to IH affecting venous and glymphatic drainage would be the main reasons. Unlike the typical forms, atypical forms may not be preceded by clinical and radiological signs of IH. We described an unusual case of fundoscopically evident bilateral TS in a patient with intercourse rupture of the right MCA aneurysm, but symptomatic only in the contralateral eye despite the major pattern of blood spillage was present in the right subarachnoid space and although angiographic vasospasm signs were evident in the M1 segment and the anterior circulation of the right hemisphere.

Ethical approval

The Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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How to cite this article: Marrone S, Pizzo C, Paolini F, Giovannini E, Crea A, Cinquemani G, *et al*. Atypical Terson syndrome after subarachnoid hemorrhage from middle cerebral artery aneurysm rupture during coitus. Surg Neurol Int. 2024;15:291. doi: 10.25259/SNI_287_2024