

Spontaneous intrapapillary hemorrhage with adjacent peripapillary subretinal hemorrhage in adolescents

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Abstract

Purpose: The purpose of this study was to report the clinical features of seven eyes of seven adolescent patients with spontaneous intrapapillary hemorrhage with adjacent peripapillary subretinal hemorrhage (IHAPSH).

Design: This was a retrospective case series study.

Participants: A total of seven adolescent patients with IHAPSH were participated.

Methods: All the patients were seen at the Joint Shantou International Eye Centre, between 2006 and 2014. The patient demographics, clinical history, vision, funduscopy, and results of relevant investigations were collected.

Main Outcome Measures: This was a clinical history and fundoscopic appearance.

Results: All eyes (four males and three females) with IHAPSH had a median age at presentation of 15 years (mean 14.7, range 11–19). Four cases presented in the right eye and three cases in the left eye. All seven patients presented with blurring of vision and floaters upon awakening in the morning. No obvious underlying causes or predisposing factors were present. The presenting visual acuity ranged from 0.12 to 1.00. One eye had no refractive errors and the remaining six eyes had myopia ranging mild to moderate (−0.25 diopters (D)—−4.00 D). None had tilted discs. In four eyes, subretinal hemorrhage was <2 disc diameters. There was associated vitreous hemorrhage in five eyes. There were no posterior vitreous detachments in any of the eyes. Visual field test showed an enlarged blind spot in four cases. Optical coherence tomography of the disc revealed optic disc swelling in two cases. Fluorescein angiography demonstrated leakage in the late phase of three eyes. At 2–24 months follow-up, the hemorrhage resolved in all seven cases with vision recovering fully to 20/20. Visual field defect of enlarged blind spots improved with the resolution of intrapapillary and subretinal hemorrhages.

Conclusions: The condition of IHAPSH resolved spontaneously without intervention in all cases. There appeared to be no predisposing factors for the development of this condition. Previously reported risk factors such as myopia, a tilted disc, Valsalva maneuver, and posterior vitreous detachment were not seen in these cases. This is the 1st time a series of young adults have been reported with this condition, all of whom presented in the same manner and had the same clinical outcomes.

Introduction

A clinical syndrome of intrapapillary hemorrhage with adjacent peripapillary subretinal hemorrhage (IHAPSH) was first described in 1975.^[1] These cases were distinct due to the significant bleeding within the optic disc that extends into the peripapillary subretinal space; in some cases, vitreous hemorrhage

occurs too. It has been described more frequently in patients with myopia, small, mildly dysplastic, tilted optic discs^[2–8] and in the young Asian population.^[3–5,8] Despite the alarming appearance, a good visual prognosis has been documented where there is spontaneous resolution without intervention.^[1–8] The cause of this condition is unknown. Vitreous traction on the optic disc during posterior vitreous detachment has been proposed as one

possible cause for IHAPSH.^[1-3,6] It has also been recognized as a subtype and consequence of Valsalva retinopathy.^[5,6] The purpose of this study was to describe further the clinical characteristics associated with IHAPSH in adolescent Chinese.

Materials and Methods

Seven patients with IHAPSH who presented to Joint Shantou International Eye Centre in Shantou, China, between January 2006 and December 2014 were identified. Data were collected and evaluated retrospectively in this observational case series. The local ethical review board approved this study and informed consent was obtained from all patients.

Only those patients with acute bleeding within the optic disc and adjacent peripapillary subretinal hemorrhage surrounding the disc hemorrhage at presentation, as visualized with ophthalmoscopic evidence, were included. Those patients with hemorrhage at the optic disc only were not included. All patients with secondary causes for disc hemorrhage were excluded from our study, which include, bleeding diathesis, Terson's syndrome, peripapillary subretinal neovascularization, ischemic optic neuropathy, polypoidal choroidal vasculopathy, and Leber's idiopathic stellate neuroretinitis.

All seven patients included in the study had demographic data collected, refractive error recorded, and history of presenting complaint with possible precipitating events reviewed. All patients underwent best-corrected visual acuity, pupillary responses, and slit-lamp biomicroscopic examination.

Fundus photography was performed for all eyes. This included descriptions of disc anatomy, clarity of disc margins, multilayer hemorrhage location and extent, measured in disc diameters, and retinal features in the involved and fellow eye. Selected cases had further diagnostic testing. Five of the patients had fluorescein angiography done, optical coherence tomography (OCT) of the disc was performed for two cases, and B-scan ultrasonography was performed for one case. Finally, a Humphrey visual field assessment was done for four cases.

All patients were followed up until complete resolution of the hemorrhage and vision had fully recovered. The follow-up period ranged from 2 to 24 months with a mean of 10.5 months.

Results

All seven patients were adolescents with a median age at presentation of 15 years (mean 14.7, range 11–19). All patients were of Chinese ethnicity. There were three female and four male patients. All patients presented with unilateral eye involvement (four in the right eye and three in the left eye). The mean refractive error was -1.19 D with six mild to moderately myopic eyes (range from -0.25 D to -4.00 D). Using the definition of mild myopia as <3 D, moderate myopia 3–6 D, and severe myopia of >6 D, one patient had no refractive error, four had mild myopia, and two had moderate myopia. No patient had high myopia. They had no significant ocular or medical

history. There was no preceding history of straining or Valsalva maneuvers. All patients presented with similar symptoms, namely, blurring of vision and floaters in the involved eye on awakening in the morning. The presenting best-corrected visual acuity ranged from 0.12 to 1.00. Intrapapillary hemorrhage was observed nasally in two eyes and in 360° in one eye. Five patients had associated vitreous hemorrhage, in which two patients had a mild form of vitreous hemorrhage and three had severe vitreous hemorrhage that obscured further ancillary investigations. The adjacent subretinal hemorrhage was on the nasal side of the disc in all eyes in this series. Severe subretinal hemorrhage extending over two disc diameters was present in four eyes [Figure 1a]. The remaining three eyes demonstrated mild subretinal hemorrhage within one disc diameter in size [Figure 1b]. None of the patients had a posterior vitreous detachment, complete or incomplete, as defined by the presence of a visible, freely moving Weiss' ring on fundal biomicroscopy. B-scan ultrasonography was performed in one patient, revealing no evidence of a partially detached posterior vitreous detachment, vitreopapillary traction. One patient was confirmed having bilateral optic disc drusen and was excluded from this study.

All eyes revealed blurred disc margins, of which it was mild in five eyes and marked in two eyes. Similarly, blurred disc margins were seen in the fellow eye in five eyes, with a mild case in four eyes and marked in one eye. None of our patient had an unusual disc size or configuration and none were tilted.

Spectral-domain OCT (Cirrus HD-OCT; Carl Zeiss Meditec, Inc., Dublin, CA) of the optic nerve head and retinal nerve fiber layer was performed in two of seven eyes, at the time of presentation. They showed a thickened retinal nerve fiber layer in the nasal sector of the optic disc, which corresponded to the area of disc hemorrhage. In both eyes, a crowded disc was also demonstrated. No optic disc tilting presents. Furthermore, no bleeding occurred at the macula and the OCT scans of the macula were, therefore, normal.

We performed fundus fluorescein angiography (FFA) in five eyes. In early phase FFA, there was masking of fluorescein by the disc and subretinal hemorrhage at the respective area [Figure 2]. Late phase FFA showed fluorescein leakage in only two cases.

Visual field testing, using a full-threshold 24-2 Swedish interactive thresholding algorithm Humphrey field analyzer

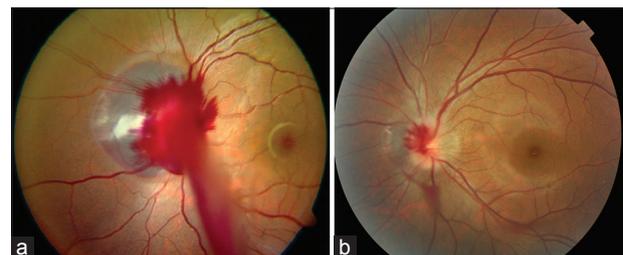


Figure 1: (a) Intrapapillary hemorrhage with adjacent peripapillary subretinal hemorrhage presenting with prominent hemorrhage at the optic disc and adjacent subretinal hemorrhage. (b) Mild hemorrhage at the disc with a smaller more localized hemorrhage in the adjacent subretinal space



Figure 2: Early phase fundus fluorescein angiography shows masking from the disc and subretinal hemorrhage

(Carl Zeiss Meditec, Inc., Dublin, CA), was performed in four of seven eyes. They all, acutely, showed an enlarged blind spot. The area of enlarged blind spot increased with the size and extent of subretinal hemorrhages. Three patients underwent magnetic resonance imaging (MRI) brain, without Gadolinium enhancement, which was reported as normal. These patients had an MRI brain to rule out other differential etiology, primarily to look for causes of intracranial hypertension that may lead to papilledema and disc hemorrhage.

All disc and adjacent subretinal hemorrhages resolved over a period of 1–9 months (mean of 3.5 months). Once the hemorrhages resolved the visual acuity recovered to 20/20 in all eyes and the visual field defect of enlarged blind spot also reduced in size.

Discussion

IHAPSH is a rare benign clinical syndrome with excellent prognosis, characterized by a sudden deterioration of vision and hemorrhage in and surrounding the optic nerve head. The previous reports on IHAPSH described patients from a diverse range of age and ethnicities.^[1-3] In this case series, findings were consistent with the previous reports in some respects but dissimilar in several key aspects, which will be discussed in the following passages.

First, the coexistence of a crowded, tilted disc,^[2,5,6,8] disc edema,^[2,5] and high myopia^[9,10] has been observed in previously published reports. Our series show similar results as crowded and edematous optic nerve head was found on fundoscopic examination and was further supported by similar findings on OCT in the cases this was performed in. This is in keeping with the notion that patients with a crowded optic nerve head are predisposed to IHAPSH. A structurally unique vascular anatomy of the prelaminar portion of the disc has been documented.^[11,12] This region is supplied by the peripapillary choroid and posterior short ciliary arteries and most of the venous drainage is to

the central retinal vein with the remaining drainage to the peripapillary choroidal veins. Since myopic, tilted, discs have an elevated superior and nasal margin, it has been suggested by Kokame^[5] that this leaves capillaries vulnerable to bleeding when retinal and choroidal tissues are dragged over and around the elevated edge.

Teng *et al.*^[8] proposed an additional theory that small scleral canal compression could play a major role in the pathogenesis of IHAPSH. They observed on OCT that nerve fibers reach the nasal retina by entering through a small scleral ring with a sharp nasal edge, along an acutely angled sclerol pathway, whereas temporal fibers gently bend over a rounded scleral margin. Thus, predisposing the choroidal arteries of the prelaminar nasal rim portion to vascular compromise and bleeding. It has been reported that increased myopia is associated with increasing tilting of the disc.^[10] However, one aspect of our results was contrary to previously published suggestion that this condition is predominantly in patients with high myopia or tilted disc, as exemplified when none of our seven cases had tilted discs or high myopia. It can be inferred that an alternate theory could exist. An interplay of ocular motor forces, scleral thinning, and vitreopapillary traction acting on a morphologically vulnerable optic disc has been suggested by Sibony *et al.*^[7]

It should be noted that this typical appearance can mimic papilledema which may lead to unnecessary investigations, brain imaging, and a misdiagnosis.^[6] Clinical judgment and caution are needed to differentiate other causes of disc hemorrhage from this benign syndrome with no systemic correlations. This is especially important as other conditions have been described to have hemorrhagic complications. These include optic disc drusen,^[6,13] bleeding diathesis,^[14] increased intracranial pressure,^[15] peripapillary subretinal neovascularization,^[16] and Leber's idiopathic stellate neuroretinitis.^[5] Knowledge of these conditions would be helpful on devising the most appropriate management plan.

Furthermore, all our patients with IHAPSH are adolescents with ages ranging from 11 to 19 and of Chinese origin. To exemplify the contrast, a case series conducted by Sibony *et al.*^[7] revealed all 10 of them to be of Caucasian ethnicity with a range of 8–63 years of age. A female predominance was noted in some studies^[4,7] and a male predominance in others.^[8] In keeping with a study of 16 eyes by Hwang and Lin,^[6] we similarly report no gender predominance. It can be inferred that gender differences may not be determining factor in the development of this rare condition.

The etiology of IHAPSH remains largely unknown; as mentioned previously, some believe that it is caused by traction of the vitreous at the optic disc.^[1-3,6] This was also suggested by Schepens^[17] who postulated that this vitreopapillary traction could lead to blurring of disc margins and elevation of the disc, causing tearing of superficial vessels which leads to hemorrhage in and around the optic disc. He described this as the “pseudo-papilledema” phenomenon, in which there was incomplete posterior vitreous detachment, due to firm vitreopapillary attachments, that has yet to be weakened by age proven by

anatomic evidence.^[18-20] Katz and Hoyt^[2] supported this theory when they found the posterior vitreous body separated from the retina but continued to be attached to the disc based on glial tags on the optic discs and B-scan ultrasonography results. Notably, none of the patients in this study had signs of posterior vitreous detachment, suggesting that this is an unlikely cause in our group of patients.

The previous studies also postulated that IHAPSH may be precipitated by the Valsalva maneuver. This phenomenon was first described in 1973 where there is an elevation of venous pressure due to the rapid rise in intra-abdominal or intrathoracic pressure against a closed glottis, which is transmitted to the eye and results in the rupture of typically, perifoveal capillaries beneath the internal limiting membrane, in the macular region.^[21] A plethora of Valsalva maneuver-induced retinal hemorrhage has been documented in literature; these include during a motorcycle ride,^[22] vigorous sexual activity,^[23] while inflating party balloons,^[24] weight lifting,^[25] and aerobic exercise.^[26] Hence, it has also been suggested that disc and subretinal hemorrhage could be a subtype of Valsalva hemorrhagic retinopathy.^[5,26] Although, the actual location of haemorrhage is variable, that is, subretinal, intraretinal or subhyaloid.^[21] Kokame *et al.*^[4] described a more direct mechanism where the sudden, shearing, stress causes rupture of susceptible choroidal capillaries on the nasal rim of the optic nerve head. To further support this hypothesis, it has been recorded on color Doppler imaging that reduced central retinal artery perfusion occurs following sneezing.^[26] However, our study findings refute this hypothesis as all seven patients in our study suffered from IHAPSH spontaneously, upon awakening in the morning, without a preceding history of exertion or straining. This suggests that, although Valsalva maneuvers can be a precipitant for IHAPSH, it can also occur without a precipitant, which raises the possibility of an alternate explanation.

The key characteristic that distinguishes our series from the previous reports is that all our patients presented similarly, namely, a sudden onset of visual blurring after awakening in the morning in one eye. To the best of our knowledge, there was no mention in literature of the exact timing of visual symptom onset. One factor that could contribute during the night to IHAPSH is the circadian periodicity in blood pressure which describes an extensive lowering of nocturnal levels of blood pressure, described as a nocturnal dip, with the largest surge on arousal in the morning. This morning peak has been associated with an increase in cardiovascular risk and several meta-analyses have reported the highest prevalence of myocardial infarction and stroke in the morning hours between 6 am and noon.^[27-29] Another factor could be due to the “non-dipping” pattern of blood pressure described as being more prevalent in patients with intracerebral hemorrhage, in which non-dipping is defined as a nocturnal systolic blood pressure dip of <10%.^[30] Further studies are required to ascertain whether these results can be extrapolated to retinal capillaries.

We acknowledge that the main limitation of this study is its small sample size due to its rarity, which limits our ability

to perform statistical analyses to control for potentially confounding factors.

Conclusions

It is likely that IHAPSH occurs in eyes with an anatomical predisposition to some extent, specifically, a crowded and edematous disc. However, the triggering event remains unknown as this study refutes the previous hypothesis that vitreopapillary traction and Valsalva maneuver at the disc lead to IHAPSH. It is also presented in both genders in a non-specific manner. This is the 1st time a series of young adults have been reported with this condition, all of whom presented in the same manner on awakening and had the same clinical outcomes.

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