Intrapapillary Hemorrhage with Adjacent Peripapillary Subretinal Hemorrhage in Myopic Chinese Adolescents

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Abstract

Background: To report three cases of Intrapapillary Hemorrhage with Adjacent Peripapillary Subretinal Hemorrhage (IHAPSH) in Chinese adolescents, and to review the published literature on IHAPSH.

Case presentation: Three patients were diagnosed with IHAPSH in a tertiary ophthalmic centre in Hong Kong between 2007 and 2019. All patients were young Chinese with moderate to severe myopia, and crowded/tilted discs, presenting with unilateral optic disc hemorrhage. Their disc hemorrhages gradually resolved over 20 weeks after initial presentation, without recurrence or long-term sequelae identified during a follow-up period of up to 134 months.

Conclusion: To the best of our knowledge, this is the first case series of IHAPSH from Hong Kong, illustrating the typical benign course of the disease that usually leads to complete resolution. Diagnosing the disease can be a challenge as the clinical signs often mimics other serious pathologies such as papilloedema, optic neuritis or even Terson’s syndrome. In Hong Kong where the incidence of myopia is one of the highest in the world, ophthalmologists should be aware of the existence of this condition and keep an open and vigilant mind in order to make the correct diagnosis. Careful history taking and appropriate ophthalmic examinations are prudent in establishing the diagnosis, while other additional investigations should be arranged if there is clinical suspicion of more sinister diseases.

Keywords: Disc hemorrhage; Optic disc; Myopia; Intrapapillary hemorrhage; Optic disc hemorrhage; Subretinal hemorrhage; Adolescents

Introduction

Despite being first described in the literature in 1975 [1], Intrapapillary Hemorrhage With Adjacent Peripapillary Subretinal Hemorrhage (IHAPSH) has seldomly been reported worldwide, with only small case series reported in the United States of America (USA) [2], Japan [3], Taiwan [4] and the Mainland China [5]. There is a recognized association with crowded and tilted myopic discs [3,6] and given the high prevalence of myopia in Hong Kong (70-80%, one of the highest in the world [7-9]), it is perhaps surprising that IHAPSH has not been reported in Hong Kong previously. The appearance of bilateral crowded discs with disc hemorrhage, especially in young people, may be alarming and raise the suspicion of increased intracranial pressure or even Terson’s syndrome. The lack of awareness of this condition may often lead to costly radiological studies and invasive procedures, resulting in unnecessary financial burden and patient’s anxiety. The case series aims to illustrate the clinical characteristics and diagnostic pathway of IHAPSH and compare them with other cases reported elsewhere.

Case presentation

We have retrospectively reviewed the records of patients attending the neuro-ophthalmology clinic in a tertiary ophthalmic centre in Hong Kong from 2007 to 2019, and identified three case of IHAPSH.

Patient I

A 9 year-old Chinese boy with good past health and no family history of ocular diseases, was referred to our unit for incidental finding of hemorrhage over the nasal aspect of the right optic disc on fundus photography during routine optometrist check-up. The patient was asymptomatic, and had no history of head injury but recalled previous roller coaster ride in an amusement park.

The ocular history was unremarkable except for rapidly progressing myopia from Plano 2 years before presentation. On presentation, his Best-Corrected Visual Acuity (BCVA) was 1.0 for both eyes (Right eye: -3.50/-0.50 x 5 and left eye: -3.25/0.50 x 170). No Relative Afferent Pupillary Defect (RAPD) was detected. Color vision, Extraocular Movement (EOM) and Visual Field (VF) by confrontation were unremarkable. Anterior chamber examination and intraocular pressure were normal. Dilated fundus examination showed right myopic tilted disc with small patch of nasal subretinal hemorrhage, blurred disc margin without definite Retinal Nerve Fiber Layer (RNFL) swelling. (Figure 1) The left eye showed myopic tilted disc with clear margins.

Both plain Computed Tomography (CT) scan of the brain and Magnetic Resonance Tomography (MRI) brain and orbit with gadolinium had been performed with normal findings. Ultrasound scan only demonstrated Optic Nerve Head Drusen (ONHD). The peripapillary RNFL thickness of the right eye and the left eye were 108 um and 98 um respectively as detected by Cirrus Optical Coherence Tomography (OCT). Humphrey VF was done for both eyes but the test results were of low reliability due to unsteady fixation.

The retinal hemorrhage completely resolved at 5 months of follow up. The patient remained asymptomatic with no further recurrence disc hemorrhage during 23 months of follow up.

Patient II

An 11 year-old Chinese boy, with unremarkable perinatal, medical and family history was referred for incidental finding of left optic disc hemorrhage on fundus photo during optometric check-up. The patient has been previously followed up for pseudo-squint from 10 months to 6 years old, with otherwise unremarkable ophthalmic history.

The patient was asymptomatic all along. He had a recent history of increased myopia of approximately 0.5 diopters in 6 months but denied recent trauma. His BCVA were 1.0 for both eyes (Right eye: -6.25/-0.25 x 15 and left eye: -7.25/-1.00 x 155).

On examination, no RAPD was detected. Colour vision and visual fields by confrontation were full for both eyes. Anterior chamber examination and intraocular pressure were unremarkable.

Dilated fundus examination showed normal fundus with bilateral small slightly tilted pink discs. A small disc hemorrhage with blurring of disc margin at 8 o’clock was noted over the left eye (Figure 2). Ultrasound B-scans showed bilateral ONHD.

Blood results were unremarkable. There was no leakage identified on fluorescence angiography.

The patient achieved a complete resolution of retinal hemorrhage at 16 weeks of follow up. At the latest follow up at 11 years (134 months) after initial presentation, there had been no recurrence of disc hemorrhage and the patient remained asymptomatic.
**Patient III**

An 18 year-old Chinese young lady, with good past health was referred for 4-day history of right eye scotoma. She had no other eye or neurological symptoms.

She recalled a history of minor head trauma 1 day before symptom onset when she hit the temporal and occipital region of her head against a handrail.

On examination, BCVA was 1.0 for both eyes (Auto-refraction right eye: -3.75/-0.25 x 92 and left eye -4.75/-0.50 x 61). Intraocular pressures and colour vision were normal. Pupils were equal and reactive with no RAPD. EOM were full with no diplopia or pain.

Anterior chamber examinations were unremarkable. Dilated fundus examination showed bilateral myopic crowded and tilted discs with dense flame shaped hemorrhage over the right superior disc (Figure 3).

![Figure 3: Right disc morphology at the time of presentation under 90D non-contact slit lamp lens view. Dense flame shaped hemorrhage over right superior disc was evident.](image)

Blood tests were unremarkable. Ultrasound scan did not reveal any calcification suggestive optic nerve head drusen. CT scan of brain and orbit without contrast was unremarkable. Lumbar puncture was performed with normal opening pressure and unremarkable biochemical findings. MRI brain and orbit with gadolinium was non-contributory.

Patient was referred to neuro-ophthalmology clinic 2 weeks later after initial presentation. By then, there had already been resolution of symptom and marked reduction in disc hemorrhage (Figure 4). Complete resolution of flame-shaped disc hemorrhage was noted at 10 weeks of follow up. Patient remained asymptomatic over 38 months of follow up.

![Figure 4: Fundus photo taken 2 weeks after initial presentation. Gradual resolution of right superior disc hemorrhage over the tilted crowded disc was evident. The macula morphology was normal.](image)

**Table 1: Summary of the Patients’ demographics and clinical course.**

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age at presentation (years)</th>
<th>Gender</th>
<th>Affected Eye</th>
<th>Refractive Error of the affected eye</th>
<th>Symptom</th>
<th>Duration between presentation to diagnosis (week)</th>
<th>Duration between presentation and hemorrhage resolution (week)</th>
<th>Duration of Follow up (month)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>9</td>
<td>Male</td>
<td>Right</td>
<td>-3.75/-0.25 x 180</td>
<td>Asymptomatic</td>
<td>3</td>
<td>16</td>
<td>23</td>
</tr>
<tr>
<td>2</td>
<td>11</td>
<td>Male</td>
<td>Left</td>
<td>-7.25/-1.00 x 155</td>
<td>Asymptomatic</td>
<td>3</td>
<td>20</td>
<td>134</td>
</tr>
<tr>
<td>3</td>
<td>18</td>
<td>Female</td>
<td>Right</td>
<td>-3.75/-0.25 x 92</td>
<td>Central scotoma</td>
<td>2</td>
<td>10</td>
<td>38</td>
</tr>
</tbody>
</table>

**Discussion**

It is speculated that the unique architecture of the elevated superior and nasal margin of a tilted myopic disc predisposes to the pathogenesis of the IHAPSH as the optic nerves inserts into the sclera in an oblique manner [10]. The physical torsional force of the myopic disc structure acting onto the corresponding capillaries would ultimately lead to intrapapillary as well as peripapillary subretinal hemorrhage[11,12]. Teng et al. and Hwang et al. both reported unique disc finding in eyes suffering from IHAPSH of which the affected eyes tend to have smaller optic discs and scleral canal with a higher level of nerve fiber crowding [4,5].

**Patient demographics**

Compared to other countries, the patients’ characteristics of our series were similar to those previously reported with IHAPH. While the case series in USA reported a relatively older age of onset from age 8-64, our patients were similar to those in Taiwan, with the mean age of 15 +/- 2.6 years old. The relatively younger age of presentation could be attributed to the fact that Asian has a higher incidence of high myopia, leading an increased incidence of myopic change of the optic disc with tilted, crowded appearance.

The patients in our case series are all myopics, with two of...
the three patients with high myopia of over-6 diopters. The finding correlated with the general findings of the published case reports, which patients of IHAPH are myopic, ranging from -1.5D to -10.5D internationally [2-5].

Disease presentation

The presentation of IHAPH varies among patients. While Kokame et al. [3] reported all patients with IHAPH present with acute visual symptoms; while the cases gathered by Sibony et al. [2] were all asymptomatic. In our case series, 2 patients were asymptomatic and one noted a central scotoma. Thus the suspicion of the IHAPH should be raised in patients with disc hemorrhage with unremarkable investigation findings, irrespective of whether patient is symptomatic or not.

It is also worth noting that in addition to the sign of crowded and tilted discs, a history of rapid myopic progression is evident in IHAPH. Both patient 1 and 2 were noted to have rapidly progressive myopia before presenting to an ophthalmologist. Existing literature addressed the relationship between a crowded myopic tilted disc and IHAPH, but the rate of progression of myopia in the pathogenesis of the disease was not well defined.

Investigations

There has yet to be a consensus on the gold standard of investigations to diagnose IHAPH. As IHAPH is a diagnosis by exclusion, the degree and extent of investigations should depend on whether other sinister diagnoses have been adequately excluded. In the setting of bilateral suspected disc swelling with disc haemorrhage, increased intracranial pressure and Terson’s syndrome will need to be considered. Table 2 summarized the investigations performed in our three patients.

In our case series, Humphrey VF testing was arranged for all of the patients as a part of the complete functional assessment, in addition to visual acuity and colour vision. The VF is expected to be normal, except for possible slightly enlarged blind spot caused by the hemorrhage directly. The lack of optic nerve dysfunction makes an intrinsic optic neuropathy, like optic neuritis, less likely. For non-invasive structural examinations, Teng et al. advocated the use of OCT of optic nerve head and peripapillary RNFL, as patients with IHAPSH tend to have smaller optic disc and a higher level of nerve fibre crowding comparing with control subjects; as well as a larger rim area of the affected eye as compared with the contralateral unaffected eye in the same patient [5]. While the size and crowding of optic disc can be appreciated clinically, the subtle difference in rim area between the affected and fellow eyes would not be readily appreciated without the use of the OCT. Patient 1 and 2 had OCT examination of their optic discs. Patient 1 was demonstrated to have a larger rim area in the affected optic disc area compared to that of the contralateral eye (affected eye: 1.35 mm² versus fellow eye 1.33 mm²). Although OCT of patient 2 demonstrated a contradictory finding (affected eye rim area 1.10 mm² versus fellow eye 1.77 mm²), it is worth noting that the affected eye had a significantly smaller total optic disc area, therefore a direct comparison of the rim areas of the two eyes may not be appropriate. OCT of the optic nerve head (in particular extended-depth OCT), as well as B-scan ultra-sonography and fundus auto fluorescence may help demonstrate ONHD as cause of crowded discs.

Perhaps a more definitive, but invasive investigation would be fluorescein angiography, which can help confirm the presence of true disc swelling by demonstrating leakage, and help rule out peripapillary choroidal neovascularization as a cause of disc haemorrhage. Once the diagnosis of true papilledema has been excluded, costlier and invasive investigations (e.g. MRI and lumbar puncture) may be avoided.

Table 2: A Summary of the Investigations Performed in Patients with IHAPH.

<table>
<thead>
<tr>
<th>Investigation</th>
<th>Patient number</th>
<th>Fundus photo</th>
<th>Humphrey Visual Field Test</th>
<th>Blood taking</th>
<th>OCT RNFL</th>
<th>B-scan ultra-Sonography</th>
<th>MRI brain and orbit (contrast)</th>
<th>Fluorescein angiography</th>
<th>Auto-Fluorescence Imaging</th>
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</thead>
<tbody>
<tr>
<td>1</td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
</tr>
<tr>
<td>2</td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
</tr>
<tr>
<td>3*</td>
<td>✓</td>
<td>✓</td>
<td>Patient did not attend appointment</td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
<td>✓</td>
</tr>
</tbody>
</table>

*: Other investigation performed by Patient 3: Lumbar puncture; CT brain and orbit with contrast.

While more negative investigation results would make a diagnosis of IHAPH more likely, ophthalmologists should be aware of the importance of balancing the potential risk of harm bought by the invasive testing as well as the potential benefit of diagnosing sinister cause of disc hemorrhage, where prompt treatment may reduce morbidity or even mortality.
Disease course

All patients in our case series had the disease diagnosed within 3 weeks after presentation, which were at the first encounter at the neuro-ophthalmology subspecialty clinic after initial presentation to the general eye clinic. All patients had resolution of disc hemorrhage at 10 to 20 weeks after presentation, which was similar to the findings by the Japanese group (ranging 1-7 months) [3]. None of the subjects suffered from a recurrence of the disease during their follow up period.

Conclusion

In conclusion, this is the first case series of IHPH in Hong Kong illustrating the typical benign course of the disease with eventual complete resolution. Diagnosing the disease can be a challenge as the clinical appearance often mimics that of other more sinister diseases such as optic neuritis and papillodema. In Hong Kong where the incidence of myopia is one of the highest across the world, ophthalmologists should be aware of this condition and keep an open and vigilant mind for early and correct diagnosis.

Declarations

Ethics approval and consent to participate: The patients mentioned in the manuscript are anonymised.

Consent for publication: The patients mentioned in the manuscript are anonymised.

Availability of data and materials: Not Applicable

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