Metamorphopsia as an Initial Complaint of Idiopathic Intracranial Hypertension

We examined 4 patients in the past 6 years who were first seen at our institution because of metamorphopsia and who were eventually diagnosed as having idiopathic intracranial hypertension (IIH). Idiopathic intracranial hypertension is a condition of elevated intracranial pressure that occurs primarily in young women. Neuroimaging findings and cerebrospinal fluid (CSF) indices are normal. Its cause is unknown, but recent weight gain is nearly universal. Symptoms include headache, pulse-synchronous tinnitus, visual obscurations, and diplopia. Most patients have papilledema, and they may have sixth nerve palsies. Optic nerve dysfunction can occur if the papilledema is severe or prolonged.

Report of Cases. Case 1. This 46-year-old woman's medical history was notable for several sinus surgeries. Her ophthalmic history was notable for myopia (−5.00 OU). Fundus photographs taken in 1993 showed normal optic nerves, with a small choroidal hemorrhage in the right eye. At a routine follow-up visit in 1995 she complained of blurred, distorted vision in the right eye during the preceding 10 months. She noted “twinkling lights” in her vision on waking. She denied headaches, except for those associated with sinus problems. She weighed 106 kg, was 157 cm tall, and had gained 15 kg during the preceding 4 years.

Her visual acuity was 20/15 OU. She described distortion of the Amsler grid temporally in the right eye (Figure 1A). Automated perimetry showed enlargement of the physiologic blind spot in the right eye. She had Frisen stage 2 papilledema in the right eye and stage 1 papilledema in the left eye (Figure 2A). There were no abnormalities of the maculae. A fluorescein angiogram showed no abnormalities except for late staining of both optic nerves. Magnetic resonance imaging (MRI) results were normal, and lumbar puncture showed an opening pressure of 280 mm CSF, with normal indices. Treatment with acetazolamide resulted in improvement of the twinkling lights and gradual regression of the papilledema. The metamorphopsia improved slightly (Figure 1A).

Case 2. This 43-year-old woman had had metamorphopsia in the right eye for 4 months. Her medical history was unremarkable. Her ophthalmic history was notable for myopia and contact lens wear (−3.50 OU). She weighed 63 kg, was 152 cm tall, and had gained 7 to 9 kg during the past year.

Best-corrected visual acuity was 20/20 OD and 20/15 OS. She described distortion of the Amsler grid in the right eye. Her examination findings were normal except for stage 2 papilledema in the right eye (Figure 2B) and mild relative enlargement of the blind spot on automated perimetry in the right eye. There were tiny macular drusen in the right eye. A fluorescein angiogram showed only late hyperfluorescence of both optic nerves. The MRI results were normal. Lumbar puncture showed an opening pressure of 310 mm CSF, with normal indices. A multifocal ERG showed no abnormalities. Treatment with acetazolamide resulted in improvement of the papilledema, but no change in the metamorphopsia.

Case 3. This 48-year-old woman had had distortion of her vision bilaterally for 6 to 12 months. Her medical history was notable for asthma and hypercholesterolemia. Review of systems was notable for non–pulse-synchronous tinnitus and rare headaches. Her ophthalmic history was notable for myopia (−4.25 OU). She weighed 106 kg, was 157 cm tall, and had lost 7 kg in the past few months.

On examination, her visual acuity was 20/20 OD and 20/25 OS, with distortion on the Amsler grid bilaterally (Figure 1B). Examination findings were normal except for stage 2 papilledema in both eyes (Figure 2C). A fluorescein angiogram showed only late hyperfluorescence of both optic nerves. The MRI results were normal. Lumbar puncture showed an opening pressure of 310 mm CSF, with normal indices. A multifocal ERG showed no abnormalities. Treatment with acetazolamide resulted in improvement of the papilledema, but no change in the metamorphopsia.

Case 4. This 36-year-old woman noticed spots missing in the vision immediately after a hysterectomy 2 months earlier. Her medical history was notable for asthma, depression, and an episode of carbon monoxide poisoning 16 years earlier without sequelae. Review of systems was notable for a few episodes of pulse-synchronous tinnitus in the immediate postoperative period. She also noted a pressure sensation around the eyes and in the occipital region. She had recently gained 12 kg, weighing 120 kg at a height of 165 cm. Her ophthalmic history was notable for mild myopia (−1.00 OU). On examination, her visual acuity was 20/20 OU. She had no abnormalities of her left eye. Her right eye had stage 2 papilledema. Automated perimetry showed enlargement of the physiologic blind spot in the right eye. A fluorescein angiogram showed no abnormalities except for late staining of both optic nerves. Results of ocular coherence tomography (OCT) of the maculae were normal. A multifocal electroretinogram (ERG) showed no focal abnormality corresponding to her metamorphopsia.
Figure 1. A, Amsler grids drawn by case 1 at initial examination and at final follow-up showing paracentral distortion of the grid temporally in the right eye. B, Amsler grids drawn by case 3 at initial examination showing paracentral distortion of the grid temporally in both eyes. C, Amsler grids drawn by case 4 at initial examination showing paracentral distortion of the grid superotemporally in both eyes.
Figure 2. Fundus photographs at initial examination. A, Case 1 showing stage 2 papilledema in the right eye and stage 1 papilledema in the left eye. B, Case 2 showing stage 2 papilledema in the right eye and stage 0 papilledema in the left eye. C, Case 3 showing stage 2 papilledema in both eyes. D, Case 4 showing stage 2 papilledema in both eyes.
acuity was 20/20 OU with correction. She described distortion without scotoma on the Amsler grid bilaterally (Figure 1C). Examination findings were otherwise normal except for stage 2 papilledema in both eyes (Figure 2D) and moderate enlargement of the physiologic blind spots on automated perimetry in both eyes. The MRI results were normal. Lumbar puncture showed an opening pressure of 410 mm CSF, with normal indices. Results of OCT of the macula were normal. No follow-up data are available at this time.

Comment. Metamorphopsia is a visual distortion in which straight lines appear curved, and it is commonly seen in disorders that disrupt the normal orientation of the macular photoreceptors. It may remit or it may become permanent. Metamorphopsia has not specifically been mentioned in reports of IIH, but by implication it occurs in the many patients with IIH who have severe papilledema, resulting in a macular star or “fan” due to extravasation of fluid from the edematous nerve between the nerve fibers into the macula. Severe papilledema may also be associated with Paton lines (concentric retinal folds surrounding the optic nerve that may also induce metamorphopsia). A syndrome of acquired choroidal folds and hyperopia has been described with and without IIH. The incidence of metamorphopsia in patients with IIH may be underestimated because of the other, more debilitating symptoms experienced by these patients, or because physicians fail to document its presence.

The cases we describe are all unusual in that the metamorphopsia was associated with very mild papilledema (stage 2 or less). None of the patients had visible distortions of the maculae, choroidal folds, macular edema, or hyperopic shifts. We speculate that the metamorphopsia was due to traction on the retina from nerve elevation, with resultant disruption of photoreceptor orientation, or from fluid within the retina or nerve fiber layer that was not apparent by ophthalmoscopy, angiography, or OCT. Although OCT is a sensitive indicator of retinal abnormalities, it probably lacks the resolution necessary to detect subtle misalignments of the photoreceptor layer. Patients with more severe metamorphopsia associated with more severe papilledema do have distortions in their retinal anatomy discernible on OCT (J.E.A.W., unpublished data, 2004).

Despite the resolution of papilledema, none of these patients had complete relief of their metamorphopsia. We speculate that these patients did not experience complete relief of their metamorphopsia because of the permanent anatomical disruption of photoreceptor alignment associated with prolonged papilledema. Three of the women had moderate myopia, and 1 had mild myopia; it is possible that the anatomy of their myopic globes and the topography of their retinas somehow predisposed them to the development of metamorphopsia. Two patients had peripapillary hemorrhages. We considered that the metamorphopsia could be a manifestation of subtle myopic degeneration or of some other condition of the peripapillary retina (such as acute zonal occult outer retinopathy [AZOOR]). However, none of these patients had refractive errors greater than –6.00, and none had ophthalmoscopic or angiographic evidence of myopic degeneration. In addition, 2 patients (cases 2 and 3) had normal photoreceptor function when evaluated by multifocal ERG. It remains to be seen whether these women harbor an occult form of macular degeneration that will become apparent in the future.

We describe 4 middle-aged women with initial complaints of metamorphopsia coinciding with the development of mild papilledema. Because no other cause of metamorphopsia was discovered and because of the temporal association between the onset of metamorphopsia and the finding of papilledema, we believe that these patients experienced metamorphopsia as an initial symptom of IIH. Physicians should consider the diagnosis of IIH in patients with metamorphopsia and optic nerve edema.

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Optical Coherence Tomographic Findings in X-linked Juvenile Retinoschisis

X-linked juvenile retinoschisis is a progressive bilateral disease that is probably present at birth and has been documented as early as 7 weeks of age. It was first reported by Haas in 1898 and has recently become better understood as a mutation of the XLRS1 gene on the short arm of the X chromosome (Xp22). This mutation results in an abnormal retinal protein that participates in intercellular spaces. Cystoid changes arranged in a stellate pattern with radial striae projecting from the fovea are seen in all patients, along with a peripheral schisis in 50% of cases, and variable findings are well described by Gass and others. In this study, we used optical coherence tomography (OCT) (Stratus OCT, Carl Zeiss Meditec AG, Jena, Germany) to examine the foveal areas in 2 patients with juvenile retinoschisis. Our OCT findings suggest that the foveal schisis is probably located in the outer plexiform layer, not in the nerve fiber layer (NFL), as de-