Sudden Onset of Migraine Headaches as the First Symptom in a Pediatric Case of Multiple Evanescent White Dot Syndrome (MEWDS)
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Abstract
To present the transient, self-limiting nature of a pediatric case of multiple evanescent white dot syndrome (MEWDS) observed through non-invasive imaging and serial examinations, with spontaneous resolution at one month.

Case Report

I. Case History
- A 13 year old white male presents to clinic reporting of a sudden onset of migraine headaches for 3 days.
- Ocular and medical histories were unremarkable.
- Patient denied any associated symptoms including loss of vision, photopsia, scotomas, and paresis. Patient also denied any previous or recent illnesses.
- No medications or recent immunizations were reported.

II. Pertinent findings
- Entering unaided distance acuities were 20/20 OD, 20/25-2 OS, and 20/20-2 OU. Distance subjective refraction revealed low myopia OU. Confrontation visual field, color vision, and pupillary testing were all within normal limits OU. Intraocular pressures were normotensive OU. Normal accommodative and vergence findings. Normal and quiet anterior segment assessment findings OD, OS.
- Normal right fundus examination; however, the left fundus revealed evidence of a granular macular appearance with multifocal, deep whitish colored circular lesions scattered throughout the posterior pole and extending toward the mid-peripheral retina.
- Fundus autofluorescence revealed early multiple discrete hyperfluorescent spots without involvement of the optic nerve head OD, OS. Baseline Humphrey 30-2 visual field was reliable and revealed a few trace scattered paracentral defects OS which were consist with foveal granularity appearance, right eye was normal. Spectrum domain optical coherence tomography revealed mild disruption of the outer retinal layers and mild attenuation of the photoreceptor ellipsoid zone.
- A diagnosis of multiple evanescent white dot syndrome (MEWDS) was made.
- At the one week visit, the retinal appearance and clinical findings were stable. However, the patient reported that he began experiencing flu-like symptoms a few days prior to the follow up visit. The parent reported that a diagnosis of an upper respiratory infection was made by his pediatrician who believed that the migraine headaches were precursor to the virus.
- At the one month follow up visit, the fundus lesions OS resolved, vision recovered to 20/20 OD, OS, OU, and an improvement in migraine headache symptoms was reported.

III. Differential diagnosis
- Chorioretinal pathologies and posterior uveitic infectious and non-infectious diseases including birdshot retinochoroidopathy, acute posterior multifocal placoid pigment epipheliopathy, acute retinal pigment epithelitis, multifocal choroiditis, punctate inner choroidopathy, presumed ocular histoplasmosis syndrome, subretinal fibrosis and uveitis syndrome, diffuse unilateral subacute neuroretinitis, retinal pigment epithelitis and acute zonal occult outer retinopathy\(^1,4,5\).
IV. Diagnosis and discussion

- Multiple evanescent white dot syndrome (MEWDS) was confirmed as the diagnosis based on the acute onset, fundus appearance, clinical findings, and transient microstructural changes noticed over serial follow ups.
  - MEWDS is a rare white dot condition that often presents with unilateral multiple circular whitish colored lesions localized in outer retinal layers and concentrated mostly within the posterior pole, fas, and 12, 13.
  - Foveal granularity may occur simultaneously9.
  - Patient may present with symptoms including decreased vision, photopsia, visual field loss, flu-like symptoms or headaches 1, 9, 12, 13.
  - MEWDS often occurs more in young females than males 1, 9. However, these chorioretinal pathologies may manifest in children 13.
  - The prognosis is excellent although foveal changes may be residual 4, 9, 12.

- While the underlying pathophysiology of MEWDS is unknown, half of the patients with this inflammatory condition have a primary viral etiology, which occurred in this case 1, 9.
  - Other suggested causes have been infectious, neurological and autoimmune-mediated inflammatory mechanisms 2, 4, 10, 12.

- MEWDS is classified as a primary inner capillary choroidalopathy which is characterized as an acute inflammation of the outer retinal layers and choroid mainly confined within the macular zone 1, 4, 5.
  - During the initial onset of MEWDS, multifocal choroidal inflammation is thought to result in increased phagocytic activity of photoreceptor outer segments with increased production of lipofuscin, evident on fundus autofluorescence and angiography imaging 4, 11, 12.
  - As the inflammatory event resolves, regeneration of the outer segments photoreceptors occurs and a reduction in the release to the retinal pigment epithelium, resulting in decreased production of lipofuscin 4. Ultimately, the deep, white retinal lesions subside and symptoms improve 1, 4. Reoccurrence is uncommon 1, 2, 4, 12, 14.

- In this case, the patient initially presented with symptoms of migraine headaches and visual symptoms days prior to the onset of flu-related symptoms. A thorough case history is warranted when a child present with symptoms of headache 3, 6, 8. A common cause of headaches in children is an upper respiratory infection 7, 8, 14. While other life-threatening causes, such as intracranial tumors and hemorrhages, are minority, it is highly important to rule them out 8, 14.

V. Treatment & management

- There is currently no treatment for MEWDS 1, 4, 9. Most case reports and studies show improvement ranging from weeks to months 1, 4, 9.

- Over the course of follow ups, patients are closely monitored using diagnostic tests and imaging including fundus autofluorescence, fundus photography, optical coherence tomography and visual fields to evaluate the outer retinal regeneration process 4, 11.
  - Fundus fluorescein angiography, indocyanine green angiography, enhance depth imaging optical coherence tomography and multifocal electroretinography are other multimodal imaging techniques that can aid in MEWDS observation 4, 11.
  - Due to the inflammation of the outer retina, the orientation of these layers, particularly the photoreceptor outer segments, are disrupted, resulting in early hyperfluorescence visible on fundus autofluorescence and fluorescein angiography corresponding to hypoautofluorescent spots in the late phase of indocyanine green angiography 4, 11.
  - Attenuation of the ellipsoid zone and outer retinal disruption is evident on OCT 11.
  - Enlargement of the blind spot, central or paracentral scotomas are common visual field defects 4, 11.
Multifocal electroretinography during the active phase may show a reduced a-wave, consistent with inflammation at the level of the photoreceptors.

As the inflammatory event subsides and fundus appearance improves, these clinical findings should resolve as well. 1, 4, 11

VI. Conclusion

Chorioretinal pathologies can present with unusual secondary symptoms including headaches. Although MEWDS is a rare, benign disorder, diagnostic testing is critical to clinch a diagnosis and monitor the healing process.

When patients present with sudden onset of headaches, it is first important to rule out life-threatening pathologies including intracranial hemorrhages and space occupying lesions but also explore other possible causes such as stress, tension, hormonal and vision.

While the prognosis of MEWDS is excellent, you want to make an appropriate early diagnosis. If the clinical signs and tests reveal evidence of progression, consider another differential condition. In this case, using non-invasive imaging techniques with the pediatric patient yield successful compliance and management.

Bibliography


