HKJO Quiz

Answer

1. Right eye recurrent multiple evanescent white dot syndrome (MEWDS).
2. Fluorescein angiography/indocyanine green angiography findings are shown in **Figures 1** and **2**.
3. Optical coherence tomography is shown in **Figure 3**.

(Questions on page 40)

Discussion

Multiple evanescent white dot syndrome (MEWDS) was originally described in 1984 as an acute, multifocal, usually unilateral chorioretinopathy affecting young adults.[1] Patients may report photopsias, sudden blurred vision, a field defect, and sometimes an enlarged blind spot. At fundal examination, the cardinal signs of MEWDS are multiple white dots located at the level of the deep retina or retinal pigment epithelium (RPE). The white dots are mostly concentrated in the paramacular area, usually sparing the fovea itself, and are less prominent and numerous beyond the major vascular arcades.[1] Vitreal cells, retinal venous sheathing and blurring of the disc margins are often seen. The macula usually shows orange granularity or yellow dots, distinct from the large white dots which are approximately 100 to 200 μm in size. In practice, the macula granularity is a more persistent and reliable sign than the 'multiple white dots', which are largely subtle or transient.[2] Although MEWDS is usually a unilateral disease, bilateral involvement could occasionally be observed, either as simultaneous or sequential entities.[3]

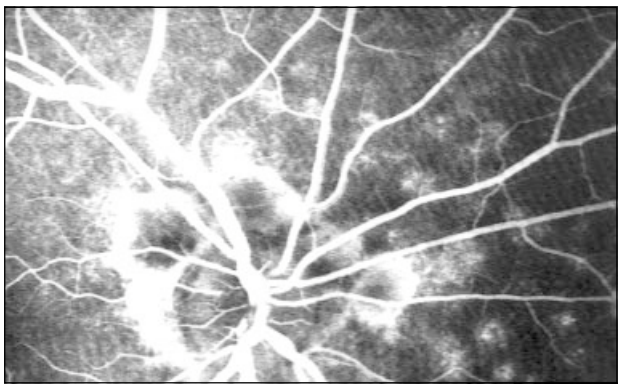

Figure 1. Fluorescein angiography of the superonasal quadrant.

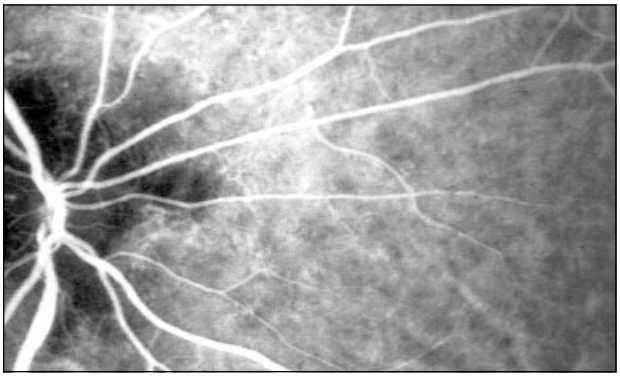

Figure 2. Indocyanine green angiograpy of the superonasal quadrant.

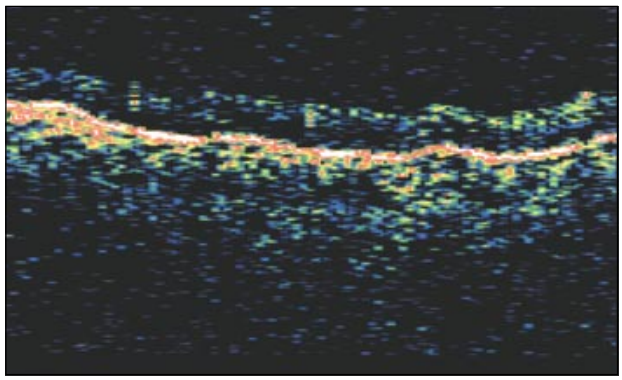

Figure 3. Optical coherence tomography of the lesions.

Fluorescein angiography (FA) and indocyanine green angiography (ICG) show typical changes in this disease. At FA, early and late hyperfluorescence of the white dots with diffuse, but patchy, late staining at the level of the RPE, retina, and disc can be detected (**Figure 1**). After resolution, these window defects may also become less prominent.[2] ICG shows no abnormality in the early phase, although in the late phase, hypofluorescent dots appear, corresponding to the white dots (**Figure 2**).[4] In the acute stage of MEWDS, the hypofluorescent areas in ICG may even outnumber the 'white dots', especially in the peripapillary area and the posterior pole. In a recent study in Japan,[5] it was shown that scotoma correlated well with the peripapillary hypofluorescent area in ICG, and it was postulated that the inflammatory changes of MEWDS involve the choroid and all layers of the retina, which block the weak background fluorescence in the late phase of ICG and cause these hypofluorescent spots.

Optical coherence tomography (OCT) is a new technology for diagnosing retinal diseases. OCT imaging in MEWDS could show up the 'white dots' in the fundus as hyper-reflective spots during an acute attack (**Figure 3**).

Electrophysiologic studies in patients with acute MEWDS have found that the electroretinogram (ERG) and the early receptor potential (ERP) amplitudes are profoundly

decreased and the ERP regeneration times are prolonged. Besides, patients with acute attack also demonstrate focal depression in multifocal electroretinogram (MERG) corresponding to the visual fields defects, with subsequent near total recovery of the MERG to baseline.[6] These results suggest a transient metabolic disturbance at the level of the RPE-photoreceptor complex.

With regard to the clinical course of MEWDS, it is usually a self-limiting disease, and the recovery of visual function occurs over several weeks with a concurrent, dramatic improvement of the ERG and ERP amplitudes. Therefore, reassurance to the patients about the benign nature of the disease is often sufficient treatment. However, recurrent disease is occasionally reported, and a recent case report from Spain documented a trial of cyclosporin to combat recurrent attacks, and showed promising control of the disease.[7]

The cause of MEWDS is still unknown. One patient with MEWDS from China with no previous history of concurrent illness exhibited increased serum immunoglobulin (Ig) M and IgG values.[8] Recovery of vision in 3 weeks was coincident with the normalization of IgM. Hence, a parainfectious disorder was suggested. Recently, human leukocyte/lymphocyte antigen (HLA) typing in patients with MEWDS found a 3.7-fold increased frequency of HLA-B51 than in a healthy Caucasian group.[9] This may point to a genetic predisposition for patients with MEWDS, but the exact mechanism of the disease is still veiled.

References

1. *Jampol LM, Sieving PA, Pugh D, Fishman GA, Gilbert H. Multiple evanescent white dot syndrome. I. Clinical findings. Arch Ophthalmol 1984;102:671-674.*
2. *Davis JL. Posterior uveitis. The University of Miami Bascom Palmer Eye Institutes Atlas of Ophthalmology. Philadelphia: Butterworth-Heinemann; 1999:360-363.*
3. *Jost BF, Olk RJ, McGaughy A. Bilateral symptomatic multiple evanescent white dot syndrome. Am J Ophthalmol 1986;101:489-490.*
4. *Obana A, Kusumi M, Miki T. Indocyanine green angiographic aspects of multiple evanescent white dot syndrome. Retina 1996;16:97-104.*
5. *Ikeda N, Ikeda T, Nagata M, Tano R, Mimura O. Location of lesions in multiple evanescent white dot syndrome and the cause of the hypoflurescent spots observed by indocyanine green angiography. Graefes Arch Clin Exp Ophthalmol 2001;239:242-247.*
6. *Oh KT, Folk JC, Maturi RK, Moore P, Kardon RH. Multifocal electroretinography in multifocal choroiditis and the MEWDS. Retina 2001;21:581-589.*
7. *Figeroa MS, Cianca E, Mompean B, Quereda C. Treatment of multiple evanescent white dot syndrome with cyclosporine. Eur J Ophthalmol 2001;11:86-88.*
8. *Chung YM, Yeh TS, Liu JH. Increased serum IgM and IgG in the MEWDS. Am J Ophthalmol 1987;104:187-188.*
9. *Boruat FX, Herbort CP, Spertini F, Desarnaulds AB. HLA typing in patients with MEWDS. Ocul Immunol Inflamm 1998;6:39-41.*