Unilateral transient subretinal hyperreflective material in multiple evanescent white dot syndrome: A case report

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ABSTRACT

Introduction: The term subretinal hyperreflective material refers to the hyperreflective material located external to the retina and internal to the retinal pigment epithelium on optical coherence tomography. Multiple evanescent white dot syndrome is a white dot syndrome characterized by transient multifocal yellow-white fundus lesions. The case of a 77-year-old female with the diagnosis of multiple evanescent white dot syndrome and an unusual and transient unilateral subretinal hyperreflective material on optical coherence tomography is reported. Subretinal hyperreflective material may be observed in multiple evanescent white dot syndrome as well as many other retinal diseases. Case Report: A 77-year-old female presented with a history of photopsia and a decreased eyesight in the right eye for the past two weeks. Fundus examination revealed a yellow macular lesion and multiple subretinal deposits in the midperiphery in the right eye. There was a diffuse subretinal hyperreflective deposition on optical coherence tomography. An extensive work-up was done to exclude systemic causes. She was diagnosed with multiple evanescent white dot syndrome as there was a spontaneous and complete resolution of the funduscopic lesions and the optical coherence tomography findings within a four weeks’ time span. Conclusion: Subretinal hyperreflective material is due to a focal or generalized dysfunction of the retinal pigment epithelium and can be present in various retinal disorders, such as multiple evanescent white dot syndrome. The thick and extensive layer of subretinal deposition is an unusual finding in multiple evanescent white dot syndrome and reflects an extensive transient dysfunction of the retinal pigment epithelium in this disease.

Keywords: Adult-onset vitelliform macular dystrophy, Age-related macular degeneration, Multiple evanescent white dot syndrome, Retinal pigment epithelium, Subretinal hyperreflective material

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INTRODUCTION

The retinal pigment epithelium (RPE) constitutes a monolayer of cuboidal cells on the exterior of the neurosensory retina and has multiple functions [1, 2]. One of the most important functions of the RPE is the phagocytosis of photoreceptor outer segments (POS) that are being shed [1, 2]. When the phagocytosis of the POS is impaired, it leads to an accumulation of debris separating the photoreceptors (PR) from the RPE resulting in PR degeneration as seen in MERTK-mutated rodent models [3]. The subretinal debris of...
accumulated POS can be seen as hyperreflective material on optical coherence tomography (OCT) [3]. Other subretinal materials, deposits, and tissue components are also hyperreflective on OCT [4]. The term subretinal hyperreflective material is therefore used as catch-all term to describe all the hyperreflective material located external to the neurosensory retina and internal to the RPE on OCT [3–5]. It is a nonspecific finding present in various retinal disorders [4]. We report an unusual thick and extensive transient unilateral accumulation of subretinal hyperreflective material in a 77-year-old female with multiple evanescent white dot syndrome (MEWDS). MEWDS is a rare white dot syndrome that typically occurs in young to middle-aged healthy women presenting with a sudden, unilateral change in vision and is characterized by transient multifocal yellow-white fundus lesions [6]. The pathogenesis is unknown but might be infectious as viral prodrome is associated in up to 50% of the cases [6].

CASE REPORT

A 77-year-old female with a past medical history of Crohn’s disease and Graves’ disease presented with a history of photopsia and a decreased eyesight in the right eye for the past two weeks. The visual acuity at presentation was 6/24 in the right eye and 6/6 in the left eye. Slit lamp examination was normal. There was no anterior chamber or vitreous inflammation present. Fundus examination revealed a yellow placoid macular lesion and multiple yellow subretinal deposits in the midperiphery in the right eye (Figure 1). Fundus autofluorescence showed a low intensity mottled hyperautofluorescence of the lesions (Figure 1). Fluorescein angiography showed a maculate hypo- and hyperautofluorescence of the macular lesion (Figure 1). Indocyanine green angiography was also performed and showed a late hypofluorescence of the lesions temporally in the late phase (Figure 1). Optical coherence tomography imaging revealed a subretinal hyperreflective deposition (Figure 2). Multimodal imaging of left eye was normal. Infectious serology was negative for bacterial infections (Bartonella henselae, Mycobacterium tuberculosis, Borrelia burgdorferi, Coxiella, Treponema pallidum, Brucella abortus, and Brucella melitensis), viral infections (Hepatitis B and C, cytomegalovirus [CMV], human immunodeficiency viruses [HIV], Epstein–Barr virus [EBV], herpes simplex virus type 1 [HSV-1], herpes simplex virus type 2 [HSV-2], and varicella zoster virus [VZV]), and parasitic infections (Toxoplasma gondii, Toxocara canis, Toxocara cati, and Strongyloides stercoralis). Autoimmune serology (rheumatoid factor, antinuclear antibodies, antineutrophil cytoplasmic antibodies, and human leukocyte antigen B27) was also negative. Hematologic workup revealed a monoclonal gammopathy of unknown significance. Follow-up showed spontaneous resolution of the retinal deposits and the hyperreflective subretinal deposition on OCT (Figure 2) with recovery of the visual acuity to 6/6 without any systemic treatment over a period of four weeks. The subretinal hyperreflective material did not recur during a one-year follow-up period.
DISCUSSION

The pathological separation of the neurosensory retina and the RPE results in a subretinal space containing materials, deposits, and tissue components that are, with the exception of serous fluid, to some degree hyperreflective on OCT and which is referred to as subretinal hyperreflective material [4]. It may be present in age-related macular degeneration (AMD), myopia, central serous chorioretinopathy (CSC), adult-onset vitelliform macular dystrophy (AVMD), and other less frequent diseases, such as pseudoxanthena elasticum (PXE) and acute exudative polymorphous vitelliform maculopathy (AEPVM). The subretinal hyperreflective material may consist of neovascular tissue, fibrosis, exudate, hemorrhage, vitelliform material, and reticular pseudodrusen [4–6]. Other imaging modalities, i.e., autofluorescence imaging, fluorescein angiography, indocyanine green angiography, and optical coherence tomography angiography, may help resolve the differential diagnosis [4].

Subretinal hyperreflective material is frequently observed in AMD and is assumed to consist of fibrin, blood, and choroidal neovascularization with the composition changing over time [4, 5]. Presence of subretinal hyperreflective material in neovascular AMD is associated with a poorer visual outcome, in particular if persistent despite intravitreal anti-vascular endothelial growth factor therapy [5]. The pathogenesis of AMD is complex and remains poorly understood, but one of the underlying mechanisms seems to be a dysfunctional autophagy of the RPE [7].

CSC is characterized by a serous retinal detachment and/or an RPE detachment in the macular region and is associated with leakage of fluid through the RPE in the subretinal space due to a hyperpermeability of the choroid [8, 9]. Dysfunction of the RPE plays an important role in the pathogenesis of CSC: the loss of the RPE barrier and pumping function results in chronic subretinal fluid [9]. Optical coherence tomography imaging typically shows a neurosensory detachment, sometimes in combination with an elongation of the POS which can be seen as subretinal hyperreflective material [8–10]. The elongation of the POS represents a transient accumulation of waste products in the POS in the absence of phagocytosis by the RPE [8, 9].

Adult-onset vitelliform macular dystrophy is characterized by a solitary, yellow round to oval macular lesion [11, 12]. Optical coherence tomography imaging of those lesions shows a highly reflective fusiform thickening of the retinal pigment epithelium ultimately resulting in an atrophic outer retinal lesion [12]. Histopathological studies demonstrated massive accumulation of lipofuscin pigments in the macular retinal pigment epithelial cells with infiltration of pigment containing macrophages in the photoreceptor layer in those lesion [11–13]. Excess POS production and/or impaired POS phagocytosis by the RPE are likely the underlying pathophysiologic mechanisms in AVMD [11].

In our case, an elderly patient presenting with photopsia and a decreased eyesight in the right eye had a transient and diffuse subretinal hyperreflective deposition. A complete work-up revealed no systemic disorders except for a monoclonal gammopathy of unknown significance. A paraproteinemic maculopathy was not considered likely as the presentation was unilateral with spontaneous resolution of the fundus abnormalities and the subretinal hyperreflective deposition.

A wide spectrum of MEWDS has been described in the literature, with atypical features reported, such as a presentation in elderly patients, bilateral involvement, chronicity, and lack of white dots [14–16]. The typical lesions in MEWDS are a disrupted ellipsoid zone on OCT with accumulations of hyperreflective material variable in size and shape [6]. We describe in this case report a transient extensive and thick subretinal hyperreflective layer as yet another OCT manifestation in MEWDS.

CONCLUSION

An unusual and transient accumulation of subretinal hyperreflective material in MEWDS is described and considered to reflect a temporary and completely reversible dysfunction of the RPE in MEWDS. An accumulation of subretinal hyperreflective material may be identified in various other retinal conditions, often resulting in scarring of the outer retina.

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**Author Contributions**

Jean-Baptiste Frédéric Willemot – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Sylvie Vandelanotte – Analysis of data, Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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Authors declare no conflict of interest.

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All relevant data are within the paper and its Supporting Information files.

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