Case Report

An Unusual Case of Unilateral Maculopathy

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Keywords
Medical retina · Unilateral acute idiopathic maculopathy · Solar retinopathy · Optical coherence tomography · Submacular fluid

Abstract
A 44-year-old male presented with unilateral sudden onset reduced visual acuity. The optical coherence tomography (OCT) scan demonstrated submacular fluid with thickening and hyper-reflectivity of the outer retinal layers, together with subfoveal retinal pigment epithelial hyper-reflectivity corresponding to a small area of foveal interdigitation zone/ellipsoid zone (IZ/EZ) loss in the detached retina. An OCT 4 months later showed resolution of the submacular fluid, but the IZ/EZ loss persisted with thinning of the outer nuclear layer, resulting in a poor visual outcome. The clinical findings most likely represent a case of unilateral acute idiopathic maculopathy; however, the OCT features and poor visual outcome are not typical. Differential diagnoses include acute solar maculopathy, central serous chorioretinopathy, poppers maculopathy, whiplash maculopathy, and acute retinal pigment epitheliitis.

Introduction
This is a case of acute unilateral maculopathy in which the clinical and optical coherence tomography (OCT) features do not correlate with any particular previously described diagnosis. The most likely diagnosis is unilateral acute idiopathic maculopathy (UAIM). The clinical findings and other possible differential diagnoses are discussed.
Case Report

A 44-year-old Caucasian male noticed sudden onset of a blurred central circle in his left eye while on holiday skiing at 1,800 m above sea level. He gave a history of using his laptop on a balcony where he was not wearing his sunglasses shortly before the visual loss and had no systemic symptoms. The patient was fit and well with no medical or recreational drug use and no recent vaccinations.

Visual acuity (VA) on presentation 1 week later was 85 EDTRS letters OD and 45 EDTRS letters OS, and examination of the fundus showed a pale circular area surrounding the left fovea (Fig. 1a). OCT demonstrated submacular fluid, with an area of increased hyper-reflectivity of the subfoveal retinal pigment epithelium (RPE) corresponding to an area of ellipsoid zone (EZ) and interdigitation zone (IZ) loss on the detached macula (Fig. 1b). In addition, there was thickening and increased hyper-reflectivity of the detached EZ, IZ, and outer plexiform (OPL) layers and thinning of the outer nuclear layer (ONL). The right eye was unaffected (Fig. 2a, b), and there were no inflammatory cells seen in the anterior or posterior chamber. The patient
was diagnosed in eye casualty with central serous chorioretinopathy (CSCR), and follow-up was arranged for 4 months later in the medical retina clinic.

At follow-up, symptoms and VA were no better and fundal examination demonstrated a fine hypopigmented dot at the central fovea. OCT showed resolution of submacular fluid with reduced thickness of the EZ, IZ, and OPL. The area of subfoveal EZ and IZ loss persisted however, as well as the ONL thinning (Fig. 1c). OCT 14 months post-presentation demonstrated a similar pattern but with slight enlargement of the EZ and IZ loss (Fig. 1d) and three new areas of RPE disturbance situated at what was the inferior edge of the previous macular elevation (Fig. 1e).

Further imaging in the form of fundus fluorescein angiography, indocyanine green angiography, and fundus autofluorescence at presentation could have contributed important information in making a definitive diagnosis; however, this was not done in the acute phase as the case was misdiagnosed as a typical case of acute CSCR. In addition, OCT angiography was not available, and the resolution of the OCT machine used at the time was not sufficient to differentiate the choroidal-scleral border; therefore, there are no choroidal thickness measurements for this case.

Discussion

UAIM typically affects Caucasian young adults with an equal predilection between the sexes; however, older patients have been reported with a range of ages from 15 to 57 [1–3]. Patients experience sudden unilateral visual loss caused by exudative detachment of the macula, commonly with a preceding flu-like viral illness in which some cases have tested positive for Coxsackie virus [1, 4]. This form of maculopathy has also been reported following an influenza vaccine [5]; however, this patient had no recent vaccinations. The neurosensory macular detachment typically has a pale appearance with irregular margins acutely followed by pigmentary disturbance after resolution of the acute phase [1, 3]. Some patients may also have posterior vitreous cells or intraretinal hemorrhages [1, 3]. Other rarer findings have also been reported including bilateral, eccentric macular lesions, and papillitis [2]. OCT in UAIM demonstrates that the photoreceptor outer segment/RPE complex is predominately involved with outer retinal thinning, irregularity of the IZ, EZ, and RPE layers often with preservation of the external limiting membrane [1]. Typically, UAIM has spontaneous recovery within 3–6 weeks with a return to near-normal vision [1, 3]; however in some cases, additional improvement may occur up to 6 months [3]. On OCT, there is normalization of the outer photoreceptor layers with time but often some persistence of the RPE disruption [1, 6]. This is in contrast to our case in which there was no recovery of the IZ and EZ layers and no recovery of vision.

An alternative cause for unilateral, sudden maculopathy is acute solar maculopathy, caused by direct sun exposure [7]. It is usually bilateral; however, there are examples of unilateral maculopathy occurring most typically in the dominant eye. Its effects are isolated to the fovea, and on examination, a yellow spot can be seen at the foveal center in the acute phase [8]. After several weeks, the OCT demonstrates a characteristic outer retinal defect in the subfoveal EZ and IZ layers with a surrounding hyper-reflective ring [8]. Acute submacular detachment has not been previously reported, and the hyper-reflectivity and thickening of the outer retinal layers at the perifovea in our case are far more severe than would be expected in solar maculopathy. The history prior to the visual loss in our case suggests sun exposure, and the final OCT appearance supports the diagnosis of acute solar maculopathy. However, acute submacular detachment with thickening and increased hyper-reflectivity of the perifoveal IZ, EZ, and OPL layers has not previously been reported as a feature.

Acute CSCR typically occurs in young and middle aged men, but there is no upper age limit, and it can also occur in women [9]. Risk factors include stress, type A personality, steroid
use, and pregnancy [9]. It is unilateral in 60% of cases and causes disturbances of the RPE with secondary serous detachments of the neurosensory retina [9]. Abnormalities of the RPE can be seen as yellow spots of hypopigmentation or pigment epithelial detachments, which may be persistent after resolution [9]. If CSCR persists longer than 4–6 months, there is a risk of permanent photoreceptor atrophy and visual loss [9]; however, the majority of cases are acute and the serous detachments resolve with restoration of near-normal visual function. Our case may fit the demographic for acute CSCR, but the OCT changes and natural history are not consistent with this diagnosis.

Popper maculopathy is a bilateral maculopathy caused by inhalation of poppers, which are volatile alkyl nitrite compounds used recreationally for their psychoactive effects [10]. OCT findings in the acute phase include disruption of the subfoveal EZ/IZ layers with varying levels of subfoveal elevation on OCT [10]. Symptoms and signs recover completely within a few weeks in the majority of cases; however, there may be persistence of symptoms in a small number of cases [10]. There was no history of popper abuse in our case, and there was no visual recovery. In addition, the hyper-reflectivity and thickening of the perifoveal outer retinal layers have not been described.

Whiplash maculopathy from sudden deceleration, most commonly seen in motor vehicle accidents, usually causes a bilateral maculopathy; however, asymmetrical cases have been described [11]. To our knowledge, there are no reported cases of deceleration maculopathy from skiing. Patients with deceleration maculopathy demonstrate intraretinal cysts within the ONL with or without subfoveal detachment. They may also develop other features of trauma such as retinal hemorrhages, commotio retinae, or a macular hole [11]. VA and OCT findings commonly fully resolve in whiplash maculopathy within a few weeks [12]. In one case report of whiplash retinopathy, the OCT did have some features similar to our case in the acute phase, with significant submacular fluid and separation of the IZ subfoveally; however, on resolution there was only a mild persistence of IZ irregularity and VA returned to near normal in 2 weeks [13]. Our patient did not give any history of deceleration accident, his VA did not recover, and the OCT features were not in keeping with previously described cases of whiplash maculopathy.

Acute retinal pigment epitheliitis (ARPE) is a transient macular disorder affecting healthy adults from the 2nd to 4th decade with unknown pathophysiology which is predominantly unilateral, but can be bilateral [14]. In the initial phase, the fundus shows retinal pigmentary macular stippling and hypopigmented halos, and OCT demonstrates areas of hyper-reflectivity from the RPE to the ONL [14]. It is usually self-limiting with good VA 2–3 months later without treatment; however, some patients develop a persistent disruption of the EZ which results in incomplete visual recovery [14]. One case of ARPE reported in 1972 in one of the first descriptions by Krill and Deutman [15] had bilateral subretinal and intraretinal serous fluid. This patient was 46 at presentation, and he required treatment with systemic steroid as there was no resolution of the edema after 3 months and was the only patient of the six described in the paper to have fluid at the macula, so it is possible this patient had an alternative diagnosis. Apart from this case described in 1972, subfoveal fluid has not been described in ARPE, and patients with ARPE invariably improve, so this diagnosis is unlikely in our case.

**Conclusion**

The acute clinical features in this case support UAIM as the diagnosis; however, there was no visual recovery and the EZ/IZ layers are not usually as severely affected. Taking other possible causes of acute maculopathy into consideration, UAIM appears to be the most likely diagnosis, and this case represents a severe form of the disease.
Statement of Ethics

This research was conducted ethically in accordance with the World Medical Association Declaration of Helsinki. There was no change in the patient’s management as a result of this case report, so ethical approval was not required. The NHS Health Research Authority decision tool was used and stated that NHS Research Ethics Committee approval is not required. Written consent to publish the clinical images was obtained from the patient and verbal consent from the patient for publication of the details of their medical case. This report does not contain any personal information that could lead to the identification of the patient.

Conflict of Interest Statement

Both authors have no conflicts of interest or financial disclosures.

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Author Contributions

All authors attest that they meet the current ICMJE criteria for authorship. Chloe J. Shipton: acquisition and interpretation of data, drafting the work, final approval of the version to be published, and agreement to be accountable for all aspects of the work to ensure accuracy and integrity. David F. Gilmour: conception and design of the work and analysis of data, critically revising the work for important intellectual content, final approval of the version to be published, and agreement to be accountable for all aspects of the work to ensure accuracy and integrity.

Data Availability Statement

No additional data are available.

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