Idiopathic Macular Ischemia: A Case Report
Kader Kasar¹, Burak Turgut*¹, Tamer Demir¹ and Orhan Aydemir¹

Abstract
We report here a case with idiopathic macular ischemia. A 46 year-old man presented to the university hospital with the complaint of blurred vision in his right eye for a period of one month. Fundus examination revealed the perivascular sheathings and attenuating arterioles, collaterals in macula in both eyes. Fluorescein angiography showed central macular ischemia in both eyes. Optical coherence tomography showed posterior vitreous detachment, macular edema, epimacular membrane in the right eye and macular atrophy in the left eye. Systemic examinations were negative for systemic vascular or vasculitic disease. We consider that macular ischemia might occur as idiopathic.

Keywords: Ischemic maculopathy; Macular ischemia; Idiopathic

Introduction
Ischaemic maculopathy occurs often as a complication of proliferative or non-proliferative diabetic retinopathy, conditions which cause retinal microvascular changes. However, it was also described in retinal vasculitis or macular vascular obstructive diseases including retinal venous or arterial occlusion, embolic phenomena such as talc retinopathy, the ocular ischemic syndrome, sarcoidosis, pars planitis, retinopathy of prematurity, familial exudative vitreoretinopathy and Eales’ disease. Ischaemic maculopathy is a sight threatening condition which does not respond to laser therapy. In ischaemic maculopathy, capillary loss affects the fovea and causes severe visual loss in colour and sharp vision [1-6]. We report here a case of idiopathic macular ischemia.

Case Report
A 46 year-old man presented to the university hospital with the complaint of blurred vision in his right eye for a period of one month. He had no remarkable history of any systemic and ocular disease or drug usage or exposure to radiation. On initial examination, his corrected visual acuity was finger counts from 3 meters in the right eye and 0.8 (with spherical -2.25 Diopters) in the left eye. The intraocular pressures were 11 mmHg in right eye and 14 mmHg in the left eye. Direct and indirect light reflexes were intact, without a relative afferent pupillary defect. The extracocular movements were full. The anterior segment examination with slit-lamp biomicroscopy was normal without iris or cristalline lens pathology and cellular reaction in both anterior chamber and anterior vitreous. Fundus examination revealed the perivascular white sheathing on major arterioles, attenuated arterioles, hard exudate plaques, and cotton wool spots and a few retinal hemorrhages in macula in the right eye, and perivascular white sheathing on major arterioles, attenuated arterioles, a few retinal hemorrhages and collaterals in macula in the left eye (Figures 1A and 1B). Late phase fluorescein angiograms showed central macular capillary non-perfusion areas in both eyes; dye leakage, retinal edema, aneurysmal dilatations in vessels and macular edema in the right eye and retina pigment epithelial window defects in the left eye (Figures 2A, 2B, 3A and 3B).

On initial examination optical coherence tomography (OCT) (Spectral OCT/SLO, GTI/OPKO Inc, Toronto, Canada) revealed...
incomplete posterior vitreous detachment, macular edema, epimacular membrane, the shadowing lines belonging hard exudates in the right eye and macular retinal atrophy in the left eye (Figures 4A and 4B).

In order to detect any ocular pathology or systematic disease to be causing macular ischemia and vasculitis such as diabetic maculopathy, retinal vessel occlusion, t alc retinopathy, sickle cell anemia, idiopathic juxtafoveal retinal telangiectasia, radiation maculopathy, central Eales disease and idiopathic retinal vasculitis aneurism and ne uroretinitis (IRVAN) and other systemic vasculitis, the comprehensive research was performed. However, it failed to reveal any underlying etiology.

Systemic examinations, including erythrocyte sedimentation rate, C-reactive protein, serum angiotensin converting enzyme, serum cholesterol and triglycerides, chest X-ray, and ECG, s alerting test; plasma protein, and urea level, complete blood cell count, serum immunoglobulins, coagulation factors and bleeding factors; transaminases and electrolytes; tumor markers; anti-cardiolipin and anti- mitochondrial antibodies; and titers for cytomegalovirus (CMV), human immune deficiency virus (HIV), toxoplasmosis and syphilis, brucella, and tuberculosis were negative for systemic vascular or vasculitic disease.

With these tests, the imaging examinations including computerized tomography thorax and abdomen, positron emission tomography and cranial magnetic resonance scanning revealed no obvious abnormalities. In systemic examinations, no pathological finding was obtained.

Consultations including Rheumatology, Heamatology, Infectious Diseases, Endocrinology and Internal Medicine clinics did not reveal any systematic inflammatory disease, systemic vasculitides, coagulopathies, protein S and C deficiency, hiperhomocysteinemy and infective diseases such as tuberculosis, AIDS or syphilis; diabetes mellitus and any endocrine disease.

Based on fundus fluorescein angiography (FFA) and OCT findings, intravitreal anti-VEGF or triamcinolone acetonide (TA) injection was recommended. However, the patient did not accept to be performing any intravitreal injection and he did not come in the next follow up visit.

On the examination 8 months after first presentation, there was no significantly change in visual acuities, fundus findings and FFA findings (Figures 1C and 1D). However OCT showed that macular edema in right eye was increased (Figures 4C and 4D). Additionally, the patient was started oral azathioprine 100 mg/day by an ophthalmologist in another hospital which he was referred two months after the presentation to us. It was recommended again an intravitreal TA injection for macular edema which was performed.

On the examination 4 weeks after intravitreal TA injection, there was no improvement in visual acuity and macular edema in right eye. In this time, intravitrealbevacizumab injection was recommended for refractory macular edema. Intravitrealbevacizumab injection was performed in dose of 1.25 mg/0.05 mL. Control examination 4 weeks following injection showed that visual acuity increased to 0.1 Snellen level and that macular edema with evidence of OCT scans reduced partially but not completely (Figure 4E and 4F). As a response to bevacizumab in macular edema was occurred, once more intravitrealbevacizumab injection was performed. Although visual acuity remained as stable in 0.1 levels, it was observed that macular edema increased in OCT (Figures 4G and 4H). The last fundus examination revealed that hard exudates reduced while other finding remained (Figures 1E and 1F). The third bevacizumab injection was recommended, however the patient did not accept more injection. The follow-up visits have been continued in our clinic.

Along follow up duration, informed consents for all diagnostic and therapeutic interventions were taken from the patient.

**Discussion**

Ischaemic maculopathy is characterized by capillary non-perfusion areas in macula and enlargement or irregularity of foveal avascular zone (FAZ) and the presence of microaneurisms at the border of FAZ due to perifoveal capillary occlusion in FFA. It often occurs with some degree of macular edema [2,3]. Visual loss usually occurs gradually and is painless. There is no proved treatment of ischaemic macular oedema and ischaemicmaculopathy does not respond to laser therapy [1-3]. Ischaemicmaculopathy is a complication of proliferative or non-proliferative diabetic retinopathy, retinal vasculitis or vascular obstructive diseases, t alc emboli, the ocular ischemic syndrome, sarcoidosis, pars planitis, retinopathy of prematurity, familial exudative vitreoretinopathy and Eales’ disease involving the macula [1-10].

At the onset of case management, we considered that macular ischemia and associated retinal findings occurred as the complication

![Figure 3: Early and late phasefluorescein angiographic frames of right and left eyes at the follow up visit in 8th month (A-D).](image1)

![Figure 4: Optical coherence tomographic scansat the presentation (A and B) and follow up visits in 8th (C and D), 10th (E and F) and 11th (G and H) months.](image2)
due to retinal vasculitis caused with systemic vasculitis. However, systemic examinations revealed no systemic vasculitic disease including polyarteritis nodosa, giant cell arteritis, and Wegener granulomatosis. Retinal vasculitis may also be a part of Eales’ disease [7], frosted branch angiitis [8], IRVAN [9] and Susac syndrome [10]. Although Eales’ disease is a form of peripheral periphlebitis in patients who have skin test reactivity to tuberculin, it may also occur in macular area. However, tuberculoprotein test and infective disease investigations for our case revealed no pathological finding. The possible causes of macular ischemia in our case were eliminated with detailed investigations.

In a recent study, it has been reported that the patients with retinal vasculitis were rarely suffered from systemic vasculitis [11]. In case of that we failed to find any possible cause for macular ischemia at detailed systemic and ophthalmologic investigations, we considered that macular ischemia in our case was idiopathic type.

References