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SHORT COMMUNICATION

Acute retinal necrosis presenting as central retinal artery occlusion with cilioretinal sparing

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PURPOSE. *To report a case of acute retinal necrosis presenting as central retinal artery occlusion with cilioretinal sparing.*
METHODS. *Single interventional case report. The findings of the ophthalmic examination, MRI, blood parameters, biopsy results and clinical course are reported.*
RESULTS. *A forty two year old gentleman reporting sudden loss of sight, ophthalmic examination revealing uveitis, central retinal artery occlusion with cilioretinal sparing and peripheral necrotizing retinitis.*
CONCLUSIONS. *Central retinal artery occlusion can be an early feature of acute retinal necrosis (ARN). (Eur J Ophthalmol 2005; 15: 287-8)*

KEY WORDS. *Acute retinal necrosis, Central retinal artery occlusion, Cilioretinal sparing, Varicella-zoster*

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INTRODUCTION

Acute retinal necrosis (ARN) was initially reported 30 years ago by Urayama et al as a distinct ocular inflammatory syndrome, the definition of which has subsequently been refined by the American Uveitis Society.

We report for the first time to our knowledge a case in which central retinal artery occlusion (CRAO) occurred with cilioretinal sparing presenting as ARN.

Case report

A 42-year-old man with a past medical history of Hodgkin's lymphoma (successfully treated 6 years previously) presented with a photophobic, painful left eye. Visual acuity was 6/5 and 6/9 in the right and left eyes respectively and after being diagnosed with left anterior uveitis, topical steroids were commenced. A few days later, he noticed acute deterioration of vision. Examination revealed visual acuity of counting fingers (CF), a relative afferent pupillary defect, inferior mutton fat keratic precipitates, with anterior and posterior uveitis. Fundoscopy revealed CRAO with attenuation of the arterioles and cilioretinal sparing (Fig. 1A). Peripherally there was perivascular infiltration of the retina with areas of necrotizing retinitis (Fig. 1B).

Blood parameters were normal and MRI of the orbit and brain revealed no recurrence of lymphoma. He underwent an urgent vitreous biopsy with intravitreal injection of 2mg foscarnet. Varicella-zoster virus DNA was identified by polymerase chain reaction analysis of the vitreous sample. He was commenced on intravenous acyclovir 10 mg/kg TDS for 7 days and then switched to 3-month course of oral valaciclovir 1g TDS. The right eye remains unaffected. To date visual acuity has remained CF with no retinal detachment.

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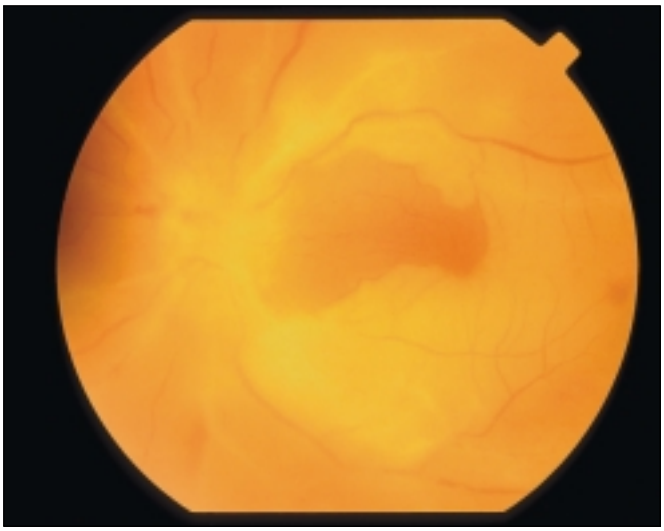

Fig. A - *Left ARN with central retinal artery occlusion and cilioretinal sparing.*

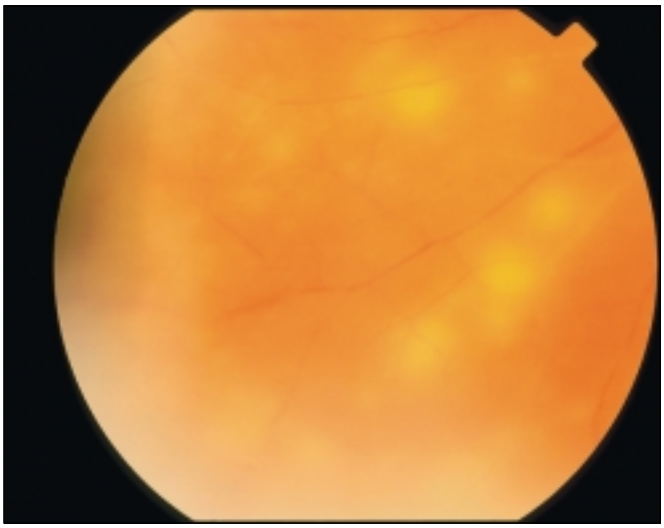

Fig. B - *Left inferonasal fundus photograph showing areas of peripheral retinal necrosis*

DISCUSSION

The diagnosis of ARN is based primarily on the clinical picture and by the exclusion of other pathologies. Clinical features are initially characterized by the development of anterior uveitis, followed by vitreous inflammation and a rapidly progressive outer necrotizing retinitis. The retinal whitening begins in multiple areas that become confluent in the peripheral fundus and branch retinal artery occlusions are common. The major cause for poor visual prognosis is secondary retinal detachment. Acute severe central vision loss is a rare complaint since the posterior pole is typically spared, however, optic nerve involvement caused by inflammatory swelling or arteritis (1) has been reported. Pathologically, there is mononuclear cell infiltration with perivasculitis and vasculitis of both retinal and choroidal vessels and indications of vascular compromise have been confirmed with colour Doppler studies (2). Hayreh (1), reporting a major critical review of 107 eyes of 78 patients, concluded that peripheral widespread retinal arteriolar occlusion and sheathing were the major vascular changes and Duker et al recognized that branch retinal artery occlusion, but not CRAO, as a sign of ARN (3). Kang et al described a patient presenting with a combined central retinal artery and vein occlusion who then subsequently developed ARN in the fellow eye (4). To our knowledge CRAO with ARN has been reported in one other patient who also presented with a subtotal retinal detachment (5). We stress dilated fundus examination of every patient presenting with uveitis and recommend that ARN is included in the differential diagnosis of an inflammatory CRAO.

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