Scholars Journal of Medical Case Reports (SJMCR) **ISSN 2347-6559 (Online)**
Abbreviated Key Title: Sch. J. Med. Case Rep. **ISSN 2347-9507 (Print)**

A United of Scholars Academic and Scientific Society, India

Atypical Inflammatory Pigmented Paravenous Retinochoroidal Atrophy

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Article History
Received: 12.03.2018
Accepted: 23.03.2018
Published:30.03.2018

DOI:
10.36347/sjmcr.2018.v06i03.025

Abstract: A 28-year- old Malay male with 6/9 OU vision was incidentally found to have hyper pigmented clumps at end retinal vascular arcades bilaterally with surrounding chorioretinal thinning and diagnosed to have pigmented parvenus retinochoroidal atrophy. He presented a year later with left eye blurring of vision of two weeks with vision of 6/12 OS, occasional fine anterior chamber and anterior vitreous cells with inferior exudative retinal detachment. Fluorescein angiography showed vasculitis at terminal vessels. Blood investigations for inflammatory markers and tests for tuberculosis were negative. Herpes simplex virus, Epstein-Barr virus and cytomegalovirus IgG were positive with a negative IgM. Based on a similar case found in literature, the patient was started on anti-tubercular medication. He showed initial improvement but anterior segment inflammation persisted and oral Azathioprine was started. Upon completion of anti-tubercular medication, vision remained at 6/24 OS. The patient's inferior exudative retinal detachment persisted and anterior segment inflammation was 3+. We then considered a viral aetiology and twice daily dose of 900mg oral valganciclovir was started. His inflammation improved with occasional residual anterior chamber cells. The patient's subretinal fluid became more shallow and vision improved to 6/12 OS. PPRCA occurs bilaterally, more often found in young men and is usually an incidental finding. PPRCA with inflammation may present atypically, and viral aetiology should be entertained for PPRCA with active inflammation even when serological evidence is not suggestive.
Keywords: vasculitis, retinal detachment, antiviral agents, uveitis, viruses.

INTRODUCTION

Pigmented paravenus retinochoroidal atrophy (PPRCA) is identified by characteristic retinochoroidal atrophy with pigment clumps along the retinal veins. Few cases of PPRCA with active inflammation have been reported. We describe an inflammatory PPRCA with unusual presentation.

CASE REPORT

Following a motor vehicle accident, a 28-year-old Malay male with 6/9 OU vision was incidentally found to have hyperpigmented clumps at end retinal vascular arcades bilaterally with surrounding chorioretinal thinning (Fig 1).

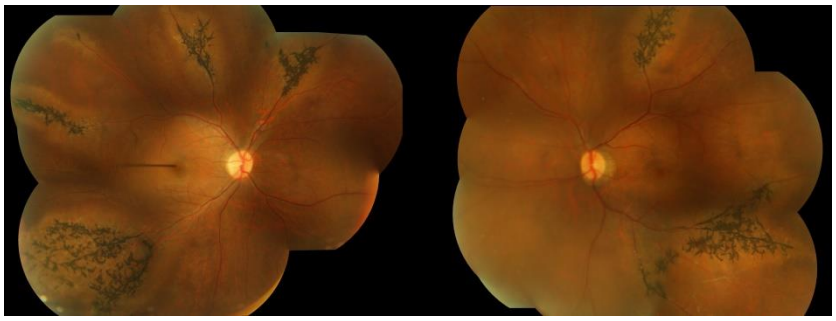

Fig-1: Fundus photograph showing hyper pigmented clumps

The patient presented a year later with left eye blurring of vision of two weeks with vision of 6/12 OS. Occasional fine anterior chamber and anterior vitreous cells with exudative retinal detachment involving the inferior vascular arcade were found. Fluorescein angiography showed hypo fluorescence corresponding to the pigmentation with vasculitis changes at terminal vessels (Fig 2).

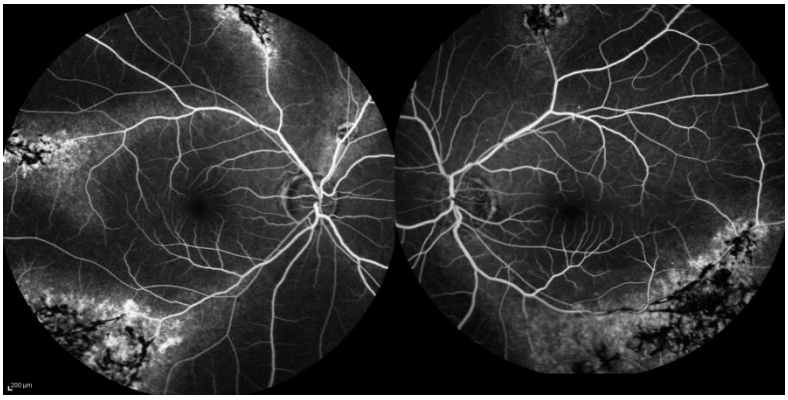

Fig-2: Fluorescein angiography

Blood investigations for inflammatory markers and tests for tuberculosis were negative. Herpes simplex virus, Epstein-Barr virus and cytomegalovirus IgG were positive with a negative IgM. The patient was started on oral prednisolone. His vision deteriorated to 6/60 OS and inflammation worsened with anterior chamber cells of 3+ and retinal detachment increased to involve the fovea.

Literature review led us to a case of PPRCA with anterior uveitis and vasculitis treated with anti-tubercular medication. Our patient was started on oral isoniazid, rifampicin, pyrazinamide and ethambutol. The patient's vision showed initial improvement to 6/18 OS and the exudative retinal detachment showed slight reduction. Anterior segment inflammation persisted and oral Azathioprine was started. Upon completion of anti-tubercular medication, vision remained at 6/24 OS. The patient's inferior exudative retinal detachment persisted and anterior segment inflammation was 3+.

We then considered a viral aetiology (however, the patient refused anterior chamber tapping for viral polymerase chain reaction). Upon discussion, the patient was agreeable to a trial of twice daily dose of 900mg oral valganciclovir. The patient developed no adverse effects from treatment and after 5 months, inflammation improved with occasional residual anterior chamber cells. The patient's subretinal fluid became more shallow and vision improved to 6/12 OS.

DISCUSSION

The first case of PPRCA was reported in 1937, but it was not until 1962 that the term pigmented paravenous retinochoroidal degeneration was coined by Francheschetti [1]. PPRCA occurs bilaterally, more often found in young men and is usually an incidental finding [2]. It is suggested that PPRCA is scarring as an end result of a degenerative disease or inflammatory insult [2,3].

PPRCA cases in which exudative retinal detachment, anterior segment inflammation and vasculitis found occurring concurrently has yet to be reported. Francois *et al*. reported a case of PPRCA with unilateral inferior retinal detachment associated with vitreoretinal degeneration and peripheral retinoschisis [4]. There are few reported cases of PPRCA presenting with active uveitis [5,6]. Haustrate *et al*. has described a case with panuveitis and periphlebitis but with no evidence of peripheral vasculitis [7].

Nagaraj *et al*. described a patient with PPRCA with a positive tuberculin skin test who had anterior uveitis and vasculitis improving with anti-tubercular medication [8]. However, a trial of the same treatment for our patient was unsuccessful. We then considered a viral cause; however none of the reported cases with active inflammation that we found had viral aetiologies or treatment with antiviral medication.

Oral valganciclovir, an L-valyl ester prodrug of ganciclovir, has vitreous concentrations comparable to its in vitro inhibitory concentrations for viral replication [9]. Valganciclovir at a dose of 900mg twice daily has been shown to be effective in treating anterior uveitis due to cytomegalovirus infection [10]. In our patient, his exudative retinal detachment and anterior segment inflammation both improved upon treatment with Valganciclovir.

CONCLUSION

PPRCA can manifest with atypical active inflammation. A viral aetiology should be entertained even when serological evidence is not suggestive.

Disclosure: The authors have no conflict of interest.

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