Case Report

Bilateral Idiopathic Retinal Vasculitis, Aneurysm And Neuro-Retinitis (IRVAN): A Case Report

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Abstract:

A teenage girl presented with posterior uveitis with retinal vasculitis, aneurysm and neuro-retinitis and a gradual decrease in vision bilaterally. There was peripheral vascular non-perfusion the aetiology of which was not found. Optical coherence tomography (OCT) and fundus fluorescein angiography (FFA) pictures are presented. The treatment included intravitreal bevacizumab injection (IVBI), systemic steroid, cyclosporine and pan retinal photocoagulation. Subsequent prevention of relapse was achieved by mycophenolate mofetil (MMF). The vision improved to 30/20 and was stable 7 years after management. To the best of our knowledge, a case with seven years follow up has not been previously described.

Key Words: IRVAN, Retina, Steroid, Uveitis

الملخص

يصف هذا المقال حالة لمريضة في سن المراهقة مصابة بالتهابات في العينين حيث تبين بعد الفحص الإكلينيكي وجود تغيرات في شبكية العينين بصورة التهابات في الشبكية والأوعية الدموية لها وكذلك تغير في قوام الأوعية وذلك بصورة تمدد وضعف في اجزاء من الأوعية الدموية، كما بين الفحص وجود ترشح في مركز البصر في الشبكية مما أدى الى انخفاض في حدة الإبصار لدى المريضة، كذلك وجود نقص في التروية في أطراف الشبكية. وبعد عمل فحص التصوير المقطعي بالتماس البصري (OCT) وتصوير أوعية قاع العين بالفلوريسنت (FFA) شخصت الحالة: التهاب الأوعية الدموية بالشبكية مجهول السبب مع تمدد الأوعية الدموية والتهاب الشبكية العصبي

(Bilateral Idiopathic retinal vasculitis, aneurysm and neuro-retinitis (IRVAN

تم اعطاء المريضة حقن بيفاسيزوماب (IVBI) والستيرويد والسيكلوسبورين وتم التخثير الضوئي للشبكية

intravitreal bevacizumab injection (IVBI), systemic steroid, cyclosporine and pan retinal photocoagulation

بعد أن تلقت المريضة العلاج المناسب تحسن حدة النظرالى ٣٠/٢٠ وظلت الحالة مستقرة ما يقارب سبع سنوات

Introduction:

The prevalence of non-infectious uveitis in paediatric population in USA was 29 per 100,000 in 2012. The magnitude and the underlying causes differ in different parts of the world [1]. Even after extensive investigations, the cause remains obscure in nearly 25% of cases [2]. A study from western Saudi Arabia reported that 40% of cases had bilateral presentation and 46% were idiopathic anterior uveitis [3]. Various management mo-

dalities have been used for idiopathic posterior uveitis, however, the long-term benefit of systemic and local treatment is not very encouraging [4].

We present a case of idiopathic retinal vasculitis, aneurysm and neuro-retinitis (IRVAN) that was managed by intravitreal bevacizumab injection (IVBI), systemic steroid and cyclosporine and pan retinal photocoagulation (PRP). The case was managed with mycophenolate mofetil (MMF) without relapse for seven years.

Case report:

A 14 years old Saudi female presented with complaints of gradual dimness of vision bilaterally. She had no history of similar complaints, ocular surgery or ocular trauma. The vision was assessed with a Snellen chart at 6-meters distance and was recorded at 20/200 without correction and 20/100 in right eye and 20/160 in left eye with pinhole. Slit lamp examination (Topcon Corp, Tokyo, Japan) indicated a normal anterior segment. There was no evidence of corneal or lenticular pathology. The pupils of both eyes were normal in size, and responded briskly to light. Applanation tonometry measurements were 16 and 18 mmHg in the right and left eyes respectively. The posterior segment was evaluated using a binocular fundus camera (Canon Inc., Tokyo, Japan) after dilation of pupil with 0.5% tropicamide eye drops.

(Fig. 1 a & b) The patient was diagnosed with posterior uveitis. Subsequently, laboratory investigations ordered including, complete blood count (CBC), urea and electrolytes, liver function tests (LFT), purified protein derivative (PPD) skin test, venereal disease research laboratory (VDRL) test, angiotensin-converting enzyme (ACE) test, lysozyme, toxoplasmosis titre, Brucella titre, erythrocyte sedimentation rate (ESR), C reactive protein (CRP), rheumatoid factor and antinuclear antibody (ANA) tests. All these investigations were within normal range. Computed tomography (CT) of the chest and magnetic resonance imaging (MRI) of the brain were normal. Optical coherence tomography (OCT) of both macula and fundus fluorescein angiography (FFA) indicated chorio-retinal inflammation and choroidal neovascular membranes (CNVM) at the macula, vasculitis and aneurysms in macular area and ischaemic changes in peripheral retina. (Fig. 1 c, d)

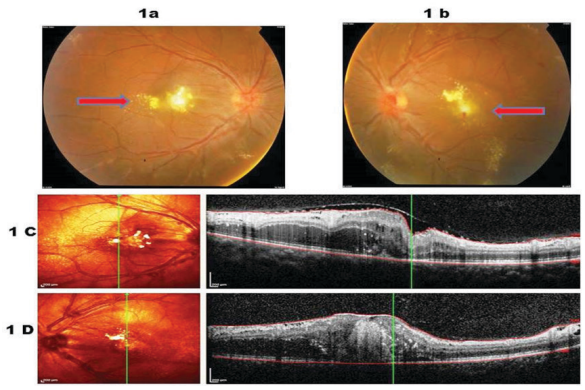


Fig. 1: Fundus photo of right eye (a) and left eye (b) shows elevated area of CNVM in the macula, also shows vasculitis with vascular sheathing. (c, d) Optical Coherence Tomography (OCT) of the same lesion both eyes.

The patient was diagnosed with IRVAN and treated with 35 mg oral prednisolone four times a day and cyclosporine 50 mg twice a day. Additionally, 0.05 mL solution contain-

ing 1.25 mg of bevacizumab was injected intravitreal.

At follow up, 6 weeks after treatment, vision was 20/160 in RE and 20/200 in the LE without correction and 20/80 in RE and 20/160 in LE with correction. FFA indicated an active leak in the macula of the RE and resolution of the macular lesion in LE. The OCT studies indicated signs of inflammation bilaterally. Hence, a second dose of intravitreal bevacizumab was delivered to both eyes and the systemic steroid was tapered. The patient was also prescribed mycophenolate mofetil (MMF) 500 mg tablets twice a day. The patient was monitored for side effects and this medication was continued as prophylaxis to prevent relapse.

At last follow up in 2017, the vision in both eyes was 20/30. The anterior segment was normal. There was evidence of PRP in both eyes. Macular scar was visible. There was no active lesion in the macula or other regions of the retina. The patient continued 500 mg MMF twice a day and lubricant eye drops four times a day. The retinal image of both eyes at last follow up is presented in Figure 2.

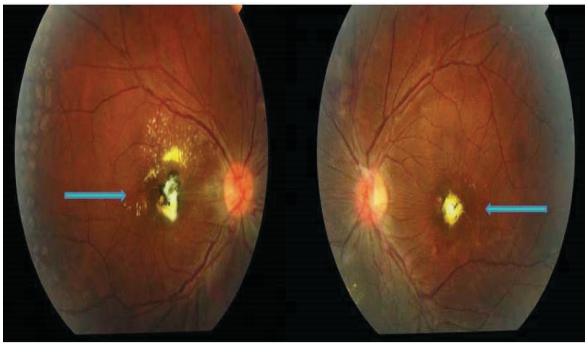


Fig. 2: Fundus photo of both eyes shows scar of cicatrized CNVM in the macula

Fundus exam shows bilateral diffuse retinal exudation more in the macular area, abnormal arterial structure in form of looping, kinging and saccular dilatation presenting aneurysmal dilatation, area of vascular sheathing in periphery (Fig. 3). Sequential fluorescein angiogaphy images of the left eye shows in the early arteriovenous phase. An early hyper fluorescence, however the vessel loops are not identified (Fig. 3a1, a2).

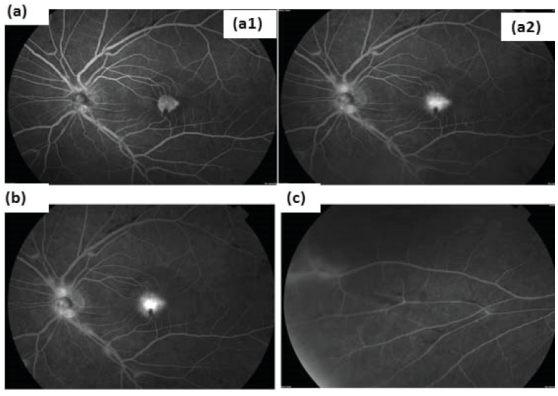


Fig. 3: Sequintional flurosceine angiogaphy images of the left eye. (a) Early hyperfluorescence; (b) Late phase hyperfluorescence; (c) Peripheral capillary drop out consisting with a wide area of non vasculrization.

In the late phase a clear leakage from the choroidal neovascularization were seen (Fig. 3b). Figure 3c shows the peripheral capillary drop out consisting with a wide area of non-vascularization.

In the early arteriovenous phase, the vessel network of the classic CNVM appears as an early well defined hyperflurescence, later there is a marked increase hyperflurescence in the area of the CNVM later the fluorescein is flowing out of the vessels and the area of increasing leakage extends beyond the vessel network that was visible in the early phase (Fig. 4).

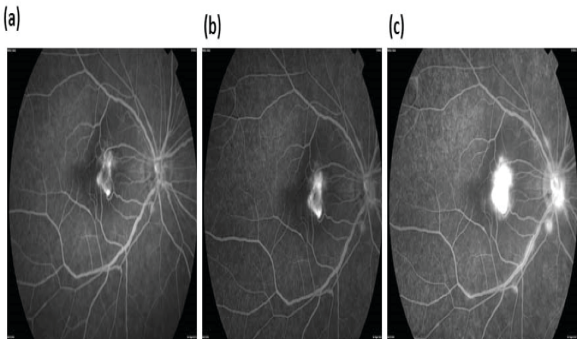


Fig. 4: (a) Early arteriovenous phase the vessel network of the classic CNVM; (b) Later arteriovenous phase of the CNVM; (c) The area of increasing leakage extends beyond the vessel network

Discussion

Reports of IRVAN have been published as early as in 1973 [5]. Although it is rare in the Asian population, there are cases reported in India and China in the last three years [6]. Hence, clinician should remain vigilant for cases of IRVAN and in order to detect it in the early stages.

Samuel et al have described the staging and criteria for the clinical diagnosis of IRVAN (7). Our case was classified as stage II IRVAN. Notably, a good outcome was achieved even 7 years after initial treatment. We believe this outcome can be attributed to early detection and management.

The laboratory investigations were normal in the current case. The patient did not have any systemic disease that could be associated to IRVAN. A case with similar presentation but associated with fungal sinusitis was described in Saudi Arabia [8]. A case of IRVAN in an adult was associated with antinuclear antibodies against WBCs [9]. However, the exact pathogenesis for the development of ocular changes in IRVAN remains unknown. Over time more investigative tools have become available (eg. OCT), but the underlying cause of IRVAN is a challenge for researchers.

The management of IRVAN has changed by time. A case in 2001 was treated with steroid and PRP and was reported as stable without remission after one year [10]. PRP in early stages is effective for addressing peripheral non-perfusion and preserving long-term vision in cases of IRVAN [11]. The successful use of intravitreal anti-vascular endothelial growth factor for treating IRVAN was documented by Karagiannis et al [12]. Slow release steroid implants in the vitreous cavity and systemic antimetabolites have been attempted for treating IRVAN [13].

MMF is used for immunosuppression mainly in cases of organ transplantation. However, it is also useful in managing uveitis as it reduces the dose of other antimetabolites used in combination and is considered a safe and effective treatment if used in suitably low doses for a prolonged time [14]. We found MMF was effective for managing IRVAN and preventing relapse without any side effects to date.

Periodic FFA and OCT is recommended of similar cases to monitor for retinal aneurysms [13]. All ocular investigations were repeated every 6 months in our patient.

The blood, renal and liver function tests were repeated at least every three months. No side effects were noted during 7 years of follow up. Use of systemic antimetabolites has been found to negatively affect renal func-

tions and the patient's quality of life especially mental health. Hence periodic investigations are recommended to rule out the side effects and a periodic mental health assessment is warranted [15].

This case of IRVAN was successfully treated with MMF for preservation of vision and halting active inflammation without relapse.

Conflict of interest:

The author declares that there is no conflict of interest regarding the publication of this paper.

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