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IDIOPATHIC RETINITIS, VASCULITIS, ANEURYSMS AND NEURORETINITIS- A DIAGNOSTIC CHALLENGE

Ophthalomology

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ABSTRACT

Diagnostic dilemma between Retinal Vasculitis in the young due to causes like Eale's disease or Coat's disease and Idiopathic Retinitis, Vasculitis, Aneurysms and Neuroretinitis (IRVAN) confuses clinicians. The case report highlights two such cases of IRVAN Syndrome. Two 30-34 years Non smoker males with no history of diabetes, hypertension and Connective tissue disorders presented with sudden onset dimness of vision in both eyes for 3-4 months. Fundoscopy revealed vascular sheathing of large vessels and macular exudates. The OCT showed macular edema. Comprehensive clinical examination only demonstrated positive tuberculin test. Initial clinical diagnoses was Eale's disease and were treated with oral steroids and posterior subtends injection of triamcinolone acetate. Follow Up examinations detected macroaneurysms and subsequent fluorescein angiography demonstrated various aneurysm. One patient was found to be pANCA positive. Patients were diagnosed as having IRVAN. Multiple doses of Ranibizumab and oral steroid reduced macular edema and recovered vision. Laser photocoagulation also has a role.

KEYWORDS

Irvan, Idiopathic Retinitis, Vasculitis, Aneurysms And Neuroretinitis, Macroaneurysms, Ranibizumab, Exudates, Retinitis

INTRODUCTION

Idiopathic retinal vasculitis, aneurysms, and neuroretinitis (IRVAN) syndrome is a rare clinical entity of unknown etiology. Arterial involvement is a common finding in cases of IRVAN, which is associated with multiple aneurysmal dilatations of the retinal and optic nerve-head arterioles. These peculiar vascular anomalies measure 75–300 μm in diameter, and are present at or near major branching sites on retinal arterioles.[1,2]

Other features include peripheral capillary nonperfusion, retinal neovascularization, and macular exudation, which leads us to believe that this disease, which was once thought to be a benign self-limiting condition, is not so, and could lead to severe vision-threatening complications if not treated in time.[2]

Various therapeutic regimens have been advocated for treatment of IRVAN, such as panretinal photocoagulation (PRP), corticosteroid therapy, administration of monoclonal antibodies, such as ranibizumab, and immunomodulatory therapy. [3,4,5,6,7]

Samuel et al conducted the largest cohort study of IRVAN, and proposed functional staging to improve treatment paradigms. They studied 44 eyes of 22 patients, and advocated PRP in cases of widespread retinal nonperfusion and before or shortly after the development of neovascularization.

He also provided for three major criteria are retinal vasculitis, aneurysmal dilations at arterial bifurcations, and neuroretinitis, while minor criteria are peripheral capillary nonperfusion, retinal neovascularization, and macular exudation.[2]

The staging was proposed to quantify the degree of retinal ischemia. Stage 1 is the presence of macroaneurysms, exudation, neuroretinitis, retinal vasculitis, stage 2 is when there is angiographic evidence of capillary nonperfusion, stage 3 is when there is posterior-segment neovascularization of the disk or elsewhere and/or vitreous haemorrhage, stage 4 is the presence of anterior-segment neovascularization, and stage 5 is neovascular glaucoma.[2]

CASE REPORT

CASE-1

A 34 year Non-smoker male, chauffer by profession presented with sudden onset dimness of vision in both eyes for 3-4 months(LE>RE).The condition progressed gradually and had no associated aggravating or relieving factors. He had no history of diabetes, hypertension, asthma and Connective tissue disorders. No medical history or family history was recorded.

General examination revealed normal parameters. Visual acuity was reduced to 6/9 in the Right eye and 6/18 in the left eye. Intraocular pressure was 16mmHg and 18mmHg in the right and left eye respectively (as measured using tonoapplanation). The anterior segment showed no abnormalities. The fundal glow was altered.

The posterior segment could be clearly visualised, CDR was bilaterally 0.3 with sharp optic disc margins; diffuse exudates with perivascular sheathing. In the left eye, exudates were centred around the peripapillary region and macula. Optical Coherence Tomography shows presence of macular edema in both eyes. The case was provisionally diagnosed as a case of retinal vasculitis.

The patient underwent blood investigations for HIV, HbsAg, Anti HCV, Serum ACE, TORCH Profile and VDRL and the investigations were found to be negative. Mantoux test yielded a positive induration >10mm.

Clinical diagnoses was (BE) Eale's disease. Patient was treated with oral steroids. This continued for 2 months.

Since there was no resolution of macular edema, two doses of posterior subtenon triamcinolone acetate injections were given in the (LE).The doses were given 1 month apart followed by serial OCT. Patient showed resolution of macular edema. Vision improved to 6/12 in the (LE). Oral steroids were gradually tapered.

Within 3 months, patient reported back with another episode of dimness of vision. Fundoscopy now revealed presence of macroanerysms along with diffuse exudates in the background of vasculitis. On OCT, Macular edema was noted.

After thorough review of literature, the diagnosis of IRVAN was established.

An OCT-Angiography was performed, showing two 'saccular' aneurysms, at the level of superficial vascular complex. A Fundus Fluoroscence Angiography, showed a large macroaneurysm, with 2 more small aneurysms, along the branching points within the equator. Leaks were noted in the venous phase.

However, no new vessels and CNP areas were noted. Patient was now planned for Intravitreal Anti VEGF. Patient received 2 doses and has been started on Oral steroids. After 1 dose, there was slight reduction in Central Macular thickness and no improvement in Visual acuity. Patient has been planned for subsequent doses and Grid laser.

Figure 1a (Right eye showing peripapillary and macular exudates, with vessel thickenings focally) and 1b (Left eye showing macroaneurysm and diffuse periarteriolar exudation)

CASE 2

A 31 year old male with no history of hypertension and diabetes presented with sudden onset dimness of vision which showed mild gradual progression in the last 8 days.He had no significant known family history, history of allergies and autoimmune disease. He had normal bowel and bladder habits and was a non smoker.

Visual Acuity in the Right eye was 6/9, while that in the lefty eye was noted to be 6/24 with affected colour vision. The anterior segments were normal. The fundus showed Clear view, CDR 0.3 with blurred optic disc margins; diffuse peripapillary exudates. Patient was diagnosed as a case of (LE) Optic neuritis and treated as per the ONTT regimen.

After 3 weeks (LE) VA resolved to 6/12 (P). OCT RNFL and AP-HFA suggested nerve fibre loss. VEP was WNL and no neurological associations were noted. However 2 months later , Patient now complained of floaters in (BE) which was more evident in the (RE). The right eye fundus showed a clear view, VCDR 0.3 with sharp optic disc margins, diffuse exudates and aneurysms Fundus fluorescein angiography suggested leakage, mainly peripapillary in location. Immunological and diagnostic workup only yielded p-ANCA positivity. After elimination of systemic causes, the diagnosis of IRVAN was elicited.

Patient was started on Oral steroids and intravitreal triamcinlone acetate. Patient was however lost to follow-up and returned after 1 year with retinal ischemia and new vessels. Vision dropped to 6/60 in both eyes. Patient was planned for urgent PRP and has currently been kept on follow up. Vision was 6/24 after treatment.

Figure 2 a and b showng presence of disc leakage, macular exudation-star aooerance with features of advanced disease and evident aneurysmal swelling in right eye

DISCUSSION

The syndrome is however not well understood since not all clinical features are present at a time and develop as the course progresses. Khairullah et al suggest that the typical stellate exudative maculopathy seen in IRVAN syndrome is primarily caused by leakage from retinal arteriolar aneurysms at or near the optic disc, and it could be misinterpreted as a feature of neuroretinitis. Patients with IRVAN syndrome usually do not present with visual function alterations consistent with optic neuropathy or neuroretinitis. Thus the acronym IRVARE (Idiopathic Retinal Vasculitis, Aneurysms, and Retinal Exudates) is proposed to better characterize this syndrome. In our cases too, neuroretinits component was not well pronounced.[8]

IRVAN's clinical presentation varies widely. Affected individuals often experience sudden vision loss, typically due to macular involvement, retinal ischemia, or neovascular complications. Imaging techniques, including fluorescein angiography and optical coherence tomography, are essential for diagnosis, allowing detailed visualization of aneurysmal changes and ischemic progression.

Asad et al affirm that management strategies focus on reducing ischemia and inflammation. Panretinal laser photocoagulation is the cornerstone of therapy, aiming to mitigate neovascular complications. Corticosteroids, immunosuppressive agents, and biologic therapies play roles in controlling inflammatory components. Anti-Vascular Endothelial Growth Factor (Anti-VEGF) treatments have shown promise, particularly in advanced disease stages.[9]

A trial of all were given, and it was noted that it can control the disease progression but not stop it. However, without therapy, the prognosis is grave. Also owing to its bilateral and subsequent affection, both eyes must be treated. Also, in the second case, a need for laser treatment was seen, due to progression of the disease. It was seen in a study Rouvas et al that early PRP may be considered in the presence of angiographic evidence of peripheral retinal non-perfusion. However, treatment could be withheld until the patient develops retinal ischemia in more than two quadrants.[10]

Differentials for this disease were Eales disease, Autoimmune Neuroretinitis (especially the second case), Adult onset Coats disease, Retinal Artery Macroaneurysm.

Even though features were similar, along with positive tuberculin test in first case, presence of macroaneurysm is a feature consistent with IRVAN. Coats disease is also a close differential, specially the Leber's military aneurysm variant. Leber's miliary aneurysms are grouped as idiopathic retinal telangiectasias, along with idiopathic juxtapapillary retinal telangiectasias and Coats' disease. The disease is usually male-dominant, unilateral, adult-onset, and slow-progressing. Aneurysms are surrounded by hard whitish exudates and are most frequently found in the mid-to-remote peripheral temporal retina. However there is not affection specific predilection for peripapillary area.[11]

Absence of hypertension and early aged presentation rule out Retinal Artery Macroaneurysm.

IRVAN has favourable outcomes when treated with a combination of PRP and intravitreal injections of antivascular endothelial growth factor. The effectiveness of this combination treatment can be seen in a case of IRVAN with both posterior and anterior neovascularization. In our case to some response was noted.

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