Central retinal vein occlusion as a presenting feature caused by Hypertensive crisis secondary to IgA nephropathy in a previously undiagnosed young patient.

ABSTRACT: We present a case of Ischemic CRVO caused by hypertension secondary to renal failure in IgA nephropathy. A 17 year old male reported to us with sudden painless diminution of vision in RE since 15 days. Fundus examination in the Right eye showed multiple superficial retinal haemorrhages in all 4 quadrents with dilated veins and cystoid macular edema suggestive of CRVO and Left eye showed superficial flame shaped haemorrhage suggestive of grade 3 hypertensive changes. On OCT (RE) cystoid macular edema is present.

Conclusion: Our case illustrates an interesting rare presentation of unilateral ischemic CRVO in a patient with IgA nephropathy. Close follow up, tight blood pressure are crucial to prevent similar scenario in the fellow eye.

INTRODUCTION

Immunoglobulin A nephropathy (IgAN), also termed Berger's disease, is one of the most common primary glomerulopathies worldwide. This immune complex- mediated disease typically affects males, in the second and third decades of life, and may be asymptomatic or manifest with hematuria and/or proteinuria. Renal biopsy is essential for its diagnosis, taking into account that IgA deposits may be observed even in patients without evidence of kidney disease[1]. Although IgAN is clinically restricted to the kidneys in most cases, there are associations with other conditions, particularly with a number of immune and inflammatory diseases, commonly rheumatic (i.e. ankylosing spondilytis, rheumatoid arthritis and Reiter syndrome), gastrointestinal (i.e. celiac disease), hepatic (i.e. alcoholic and non-alcoholic liver disease, and schistosomiasis), pulmonary (i.e. sarcoidosis), and cutaneous (i.e. dermatitis herpetiformis) [2]. Human immunodeficiency virus infection and hepatitis B (in endemic areas) have also been associated with IgAN [2]. Ocular involvement in patients with IgAN is infrequent, and the most common association occurs with uveitis [2]. Reports of the association between scleritis and IgAN are very scarce [3–5].

Case Report :- A 17 -year-old male presented with sudden diminution of vision in the Right eye since 15 days. On examination, her BCVA was 6/36 N6 in the right eye and 6/6 N6 in the left eye. color vision was 25/25 and Contrast sensitivity was 0.8 in both eyes Intraocular pressure (IOP) in the right eye was 20mmHg and in the left eye it was 21mmHg. CCT was 0.523 and 0.527 respectively.Anterior segment evaluation normal in both eyes and pupillary examination showed RAPD in right eye and normal in left eye. Fundus examination of the right eye showed a swollen and hyperemic disc, numerous retinal hemorrhages all over the fundus, tortuous and dilated retinal vasculature and macular edema (ME) (Figure 1), while fundus of the other eye showed flame shaped haemorrhage with generalised arteriolar attenuation (Figure 2). ÓCT showed increased central macular thickness (CMT) with intra retinal fluid and neurosensory detachment with irregular foveal contour in the right eye and Oct was normal for left eye.

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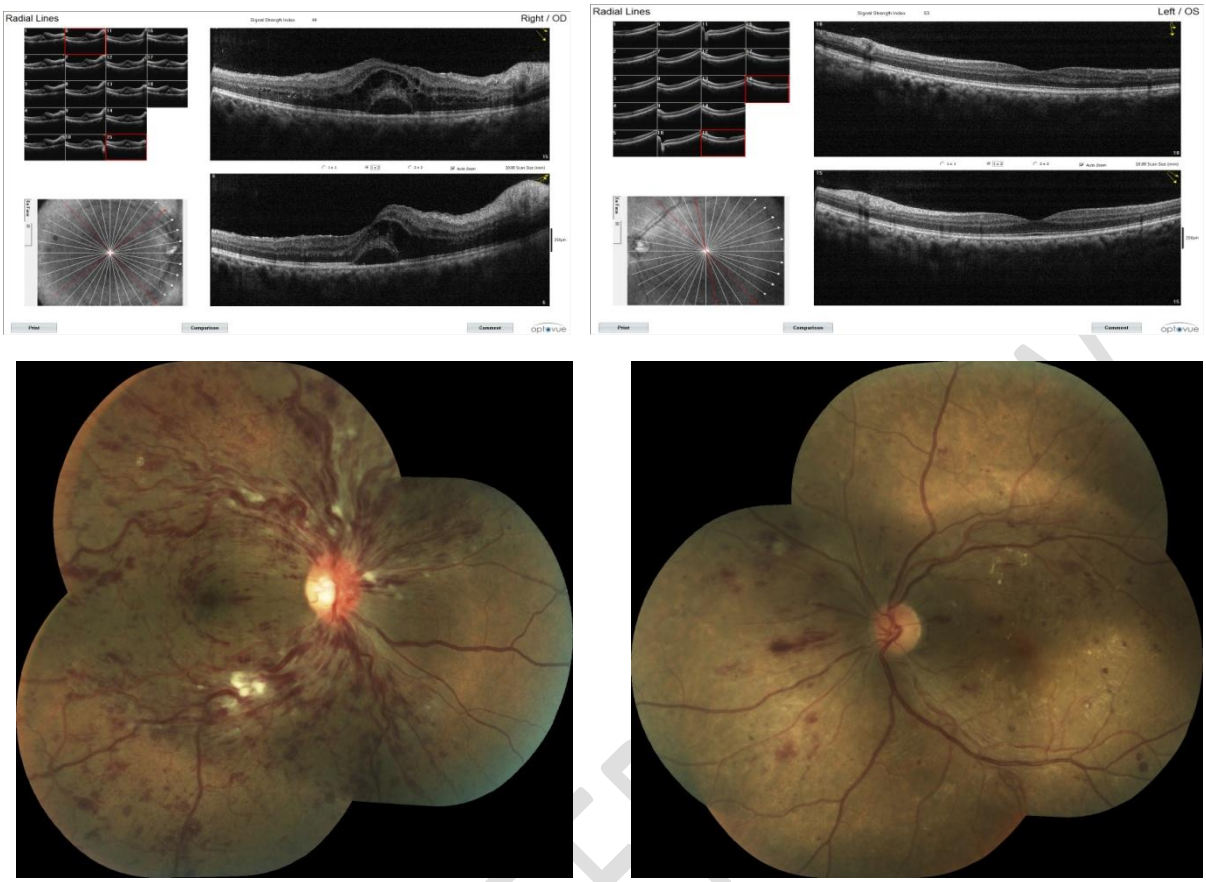

FIG 1,2: Intra retinal fluid and neurosensory detachment with irregular foveal contour

The workup included a normal complete hemogram (Hb- 6.6 gm percent, MCV-95.9 fl, MCH- 30.3 picogram, MCHC- 31.6 gm/dl, WBC- 5.4 1000/mul, Platelet count- 210 1000/mul), lipid profile (Total cholesterol 138 mg%, HDL Cholesterol 62.1mg%, Triglycerides 80 mg%) and The C- Reactive protein rate was normal 4.2 mg/l and Serum homocysteine level was 7.10 µmol/L,which was in the normal range of 6-15 µmol/L. His Montoux(tuberculin) Test and COVID antigen test was negative. His Sickling test came out to be positive, creatinine was 5.61 mg% and urea was 85.3 mg%, fasting and post prandial sugars were 73.7 mg% and 81.3 mg%. His BP was 171\81mmhg on presentation. On screening the family members his maternal uncle was postive for sickling test.Patient was referred to nephrologist and after multiple episodes of dialysis macular edema completly resolved and vision was RE 6/9 and LE 6/6.

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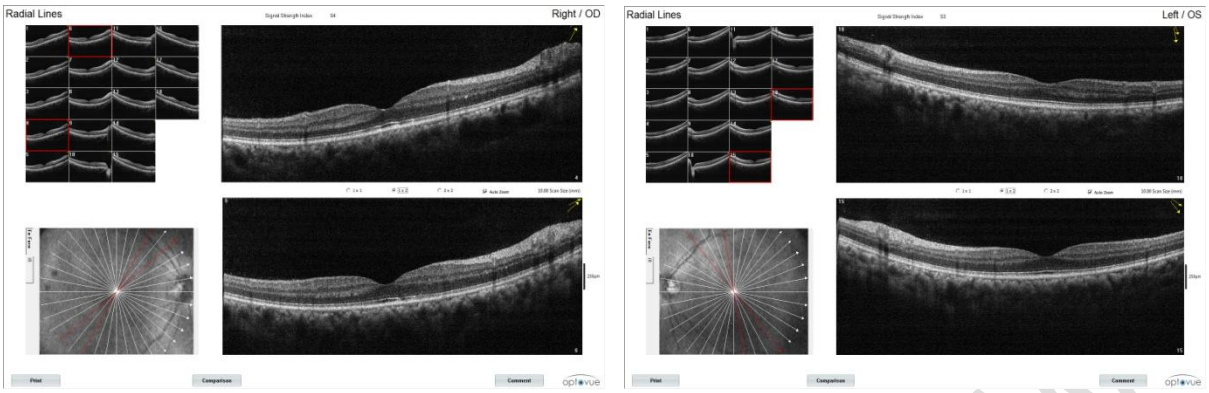

Figure 3: Radical lines

Discussion

In the presented case we report a case of CRVO secondary to hypertensive crisis in a patient with IgA nephropathy. Increased blood pressure is inherently associated with vascular damage. Mechanical stress caused by shearing forces causes endothelial dysfunction as measured by the reduced bioavailability of the nitric oxide, a potent vasodilatory factor. Concurrently, angiotensin II excess contributes to constriction of the vessels, which further confers risk of hypertension development. The examination of the retinal vessels may also show characteristic changes resulting from long-standing or uncontrolled hypertension. Narrowing of the vessels impairs capillary blood flow, which may lead to RVO, at extremes [6]. Tissue hypoxia promotes vascular endothelial growth factor (VEGF) production and release, which in turn leads to hyperperfusion. The VEGF excess is also claimed to be responsible for the initiation and progression of the oedema and ischaemia in RVO [6,7]. The reported patient also presented two other common RVO risk factors, i.e dyslipidaemia and impaired renal function. The discussed pathomechanisms contribute to the development of hypertension at an early age [8].The other factor which possibly added the risk of the development of the RVO in our patient was iron deficiency anemia. Iron functions to regulate platelet numbers and function by inhibiting thrombopoiesis [9]. In a state of iron- deficiency, there is a reactive thrombocytosis, thus leading to hypercoagulability. Red cell deformability is reduced in microcytic iron-deficient cells, resulting in an increased viscosity and furthermore contributing to the hypercoagulable state [10]. It has been hypothesized that anemic hypoxic injury to the retino-choroidal circulation causes endothelial cell dysfunction [11] and a weaker anti-oxidant defence in the IDA state, results in increased platelet aggregation [12].

Conclusion

We present this as an interesting case report of CRVO with previously undiagnosed IgA nephropathy where hypertension, dyslipedemia and iron deficiency anemia were the risk factors. The aetiology of CRVO in the presented case was multifactorial; however, hypertension was the most probable triggering factor. FFA was not performed due to impaired renal function; Ideally, future studies might be able to correlate the extent of macular flow loss seen on OCTA with severity and/or quantification of peripheral retinal ischemia, and allow for risk stratification.

Treatment includes multiple episodes of dialysis at regular intervals under a nephrologist and intra vitreal antivegf injections to reduce macular edema with frequent follow ups 3 months for the fellow eye and to look for neovascularization.

Declaration of patient consent

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The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given his consent for his their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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