ISSN: 2573-9573

Case report

Journal of Ophthalmology & Clinical Research

Optic Disc Drusenoid Deposits with Hemifield Defects in a Patient with IgA Nephropathy - New Findings

SK Prabhakar* and PV Priyamvada

JSS Academy of Higher Education and Research, JSS Medical College Hospital, Mysore, Karnataka, India

***Corresponding author**
Dr. SK Prabhakar, JSS Academy of Higher Education and Research, JSS Medical College Hospital, Mysore, Karnataka, India, Tel: 9880074529

Submitted: 18 Dec 2020; **Accepted**: 29 Dec 2020; **Published**: 09 Jan 2021

Citation: *SK Prabhakar and PV Priyamvada (2021) Optic Disc Drusenoid Deposits with Hemifield Defects in a Patient with IgA Nephropathy - New Findings. J Ophthalmol Clin Res 5: 53-57.*

Abstract

To describe emerging new OCT and Humphrey visual field findings is the objective of the study. A previously treated 20-year old female patient for skin rashes, hematuria, and pain abdomen 8 months ago reported with visual disruption in the right eye for the last one week. Features of leukocytoclastic vasculitis on skin biopsy and glomerular mesangial cell hyperplasia on renal biopsy inferred a clinical diagnosis of IgA nephropathy.

Consequently, proteinuria manifested one month after administration of prednisolone 50 mg/day tapered for 8 months, Visual acuity of 6/9 in the right eye and 6/6 in the left eye with normal color vision recorded in addition to early signs of papilledema. Classically, on OCT, hyperreflective intense echoes discovered in the optic disc, and accordingly inferior hemifield defects established predominantly in the right eye on HFA.

Total leucocyte count at11720 cells/cu mm and ESR 60 mm/hr by the Wintrobe method with the presence of albumin, RBCs, and pus cells on urine analysis in addition to positive occult stool blood detection. C3, C4, and ANCA panel revealed negative results and the USG abdomen disclosed fatty liver with mesenteric nodes. In conclusion, Optic disc drusenoid deposits corresponding with hemifield visual field defects detected in the current case study is possibly the associated early fundus manifestation of IgA nephropathy.

Keywords: Drusenoid deposits, Humphrey field analyzer (HFA), IgA nephropathy (IgAN), Ocular coherent tomography (OCT), Optic nerve head (ONH)

Introduction

The purpose of the study is to describe the new fundal manifestation and the mechanisms involved in the causation of visual disturbances. The significance of the disease is that it is the most common type of glomerulonephritis that can lead to symptomatic disturbance in the vision. Meta-analyses performed on 1,619 publications revealed greater specificity of the disease in the Asian population affecting 45 cases per million population/y in Japan and 31 cases per million populations per year in France accordingly [1].

The first description of IgA nephropathy characterized by recurrent macrohematuria or asymptomatic microhematuria, proteinuria, and nephrotic syndrome obtained from Jean Bergers who reported this clinical entity as the most common chronic glomerulonephritis worldwide [2]. Nevertheless, definitive diagnostic serological tests are unavailable to confirm the existence of IgA Nephropathy. A brief review of the literature disclosed a previously reported case of subretinal drusenoid like deposition associated with IgA Nephropathy [3]. Scleritis, episcleritis, keratoconjunctivitis, anterior uveitis, and retinal vasculitis characterize the ophthalmological findings related to IgAN [4].

Objective

Notwithstanding the associated perimacular and subretinal drusenoid deposits described in the literature, however, currently, to our knowledge association of optic disc drusenoid deposits and visual field defects are unavailable. Therefore, we present a case of IgA nephropathy patient presenting with bilateral drusenoid deposits in the optic disc with inferior visual field defects in addition to macular deposits.

Case report

General data of the case consists of a 20-year old female patient with a medical history of previously undergone treatment for skin rashes, haematuria, and pain abdomen 8 months ago with an insig-

nificant family history now presenting with disturbed vision predominantly in the right eye for the last one week. The presenting illness comprised of bilateral gradual and insidious onset of visual disturbance, obscuring the field of vision as described by the patient.

Clinical manifestation comprises of visual acuity of 6/9 in the right eye and 6/6 in the left eye with normal colour vision noted with unremarkable anterior segment examination. Bilateral optic disc hyperemia with indistinct margins, venous engorgement, and absent spontaneous venous pulsation observed on direct ophthalmoscope in addition to irregular widened macular reflex and yellowish raised lesions more-conspicuous noted in the right eye.

Methods of diagnosis included OCT analysis of optic nerve head and macular area in addition to visual fields plotting on HFA. Optic disc at the temporal and nasal aspects disclosed hyper echoic reflections that probably suggested drusenoid like deposits predominantly in the right eye. Consecutively inferior hemi field defects discovered on HFA progressed prominently in the right eye. Conversely, on OCT of the macula, similar hyper echoic reflections found specifically in the outer retinal layers and Bruch's membrane that plausibly implied drusenoid like deposits.

Laboratory investigations composed of total leukocyte count at 11720 cells/cu mm, Hb 10.8 Gm/ dL, and ESR 60 mm/hour by Wintrobe method in addition to peripheral smear showing normocytic normochromic anaemia. Skin and renal biopsy revealed features of leukocytoclastic vasculitis and glomerular mesangial cell hyperplasia that confirmed the clinical diagnosis of IgA nephropathy. Albumin, RBCs, and pus cells found on urine analysis with positive results acquired for occult stool blood.

Negative results obtained with ANA, ANCA, and pANCA with a normal level of C3 and C4, and the USG abdomen and pelvis showed fatty liver and few mesenteric lymph nodes. Normal results observed for RBS, RFT, LFT, serum electrolytes, and coagulation profile. Although no medical treatment was recommended for the visual disturbances as improvement in the visual outcome occurred within two months and as a part of systemic therapy, an immunosuppressive dose of prednisolone 20 mg per day administered.

Discussion

This case study describes new information of scientific observation with unique features not previously described that contribute to medical literature and teaching point of view. This is the first case study of naive fundus manifestations characterized by drusenoid like deposits in the optic disc correlated with hyper reflectivity on OCT and accordingly manifesting with visual field defects on HFA. This young 20-year-old female patient presented with disturbed vision essentially in the right eye with typical symptoms of IgA nephropathy [5]. Direct ophthalmoscope findings were well correlated with fundus photography and OCT images.

OCT of optic nerve head disclosed peripapillary hyper reflectivity with significant elevation at the temporal and nasal aspects mostly implying drusenoid deposits (Figure 1).

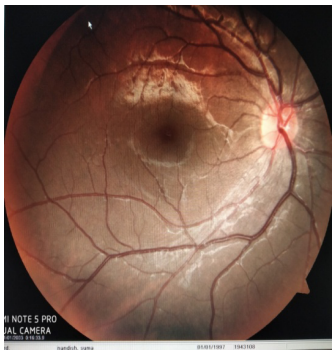

Figure 1: Right eye fundus camera picture showing peripapillary hyperreflective echos with irregular reflection at the superior macular area

Right inferior visual field defects observed predominantly with blind-spot enlargement on HFA (Figure 2).

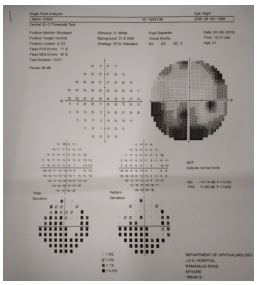

Figure 2: showing prominent right inferior visual field defects with blind-spot enlargement on HFA

Fundus imaging revealed yellowish globular elevations at the superior macular area in addition to the deformation of macular reflexes (Figure 3).

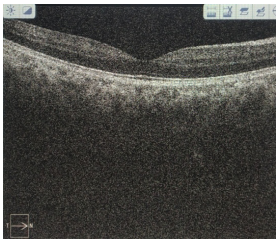

Figure 3: Right eye OCT macula showing drusenoid deposits in the outer retinal layers and Bruch's membrane

Hyper reflectivity at the outer retinal layers and Bruch's membrane revealed on OCT macula probably signifies the existence of drusenoid like deposits similar to the findings described in the previous case report. In contrast to a young female patient in the present study, one case of a 42-year-old male patient with IgA nephropathy reported in the published literature [4].

OCT optic disc abnormalities significantly correlated with field defects observed on HFA probably is the cause for diminished vision. Right field analysis by HFA revealed predominantly inferior field defects, arcuate-shaped defects in the upper nasal field, and two depressed areas noted within 20 to 30 degrees in the upper temporal quadrant. Despite the symmetrical clinical picture observed

in both the eyes however the magnitude of the lesions appreciated inconspicuously in the left eye.

Although reliability indices were within acceptable range however there were 40% false-negative errata. Accordingly, the mean deviation of -13.16 dB, p<0.5%, and pattern standard deviation at 11.65 dB, p < 0.5% found in addition to GHT interpreting the fields as outside normal limits in the right eye (Figure 2). Regardless of acceptable indices in the left eye, the GHF test showed lower patient reliability and interpreting the fields as outside normal limits with a mean deviation of -4.02 DB, p <2%, and pattern standard deviation at 8.75 DB, p <0.5%.

A 30-year-old male patient reported an acute bilateral loss of vision presented with retinal haemorrhages, cotton-wool spots, macular oedema, and optic disk hyperaemia in previously unknown IgAN with a diagnosis of grade IV Hypertensive retinopathy confounding with clinical manifestation of IgAN [5,6].

Comparison, analysis, and discussing the similarities and differences between the reported cases and present case study depicted in Table 1.

Sl No.	Author	Year of publication	Clinical features	Clinical association/Laboratory investigations	OCT	HFA	Visual recovery
1.	Lally et al	2014 vol 132	A 42-year-old asymptomatic woman manifested with proteinuria and stage III kidney disease secondary to IgA nephropathy diagnosed two years ago.	Renal biopsy demonstrated mesangial IgA deposition, expansion of the mesangial matrix, and positive direct immunofluorescence for complement C3 andC1q.	Perifoveal, hyperreflective and convex shaped deposits observed at RPE–Bruch membrane	-	Recovery observed.
2	Cetin et al	2018 vol 53	A 9-year-old girl reported with recurrent gross hematuria.	Increased serum IgA levels at 459 mg/dL [70–453 mg/dL]) detected with proteinuria and hematuria. Fundus revealed small, sharp-edged, and multiple hypopigmented areas on the macula. Kidney biopsy discovered increased mesangial matrix, cellularity, and congestion.	Bilateral hyper-reflectivity between the ellipsoid layer and RPE noted. Inner retinal layer abnormalities not detected.	-	-
3.	Ehrhardt J et al	2018 vol 115	A 30-year-old male patient reported with acute bilateral loss of vision.	Bilateral retinal hemorrhages cotton-wool spots, macular edema, and optic disk hyperemia observed on fundoscopy.	-	-	-
4.	Taban et al	2006 vol 43	Loss of vision	Developed cotton-wool spots and serous RD. Fluorescein angiography demonstrated choroidal infarcts in both eyes from hypertensive retinopathy in acute on chronic renal failure.	-	-	Vision recovered after plasmapheresis and hemodialysis
5.	Rueda-Rueda et al	2017 vol 92	A 42-year-old man manifested with episodes of unilateral uveitis in the right eye.	Anterior uveitis associated with vitritis discovered and renal biopsy revealed features of IgA nephropathy.	-	-	-
6.	Sirbat et al	1983 vol 6	Episcleritis and scleritis	Large numbers of dimeric IgA secreting plasma cells observed.	-	-	-
7.	Endoh et al	1981 vol 6	A 29-year-old male patient with IgA nephropathy developed scleritis and myasthenia gravis.	An immunofluorescent study on muscle biopsy showed deposits of immunoglobulin A in muscular vessels.	-	-	-

8.	Pavlin et al	1993 vol116	A 47-year-old woman an reported with recurrent macroscopic hematuria with red-eye.	B-scan ultrasound disclosed diffuse choroidal thickening with a completely closed-angle detected on Ultrasound biomicroscopy.	-	-	Vision recovered after Iridectomy.
9.	Hegde V et al	2009 vol 32	A 39-year-old man manifested with recurrent episodes of the red and painful left eye diagnosed as episcleritis.	IgA nephropathy detected on kidney biopsy.	-	-	-
10.	Present study		A 20-year-old female manifested with skin rash, hematuria and pain abdomen, and visual disruption in the right eye since one-week	Features of leukocytoclastic vasculitis on skin biopsy and glomerular mesangial cell hyperplasia on renal biopsy detected.	Hyperreflective intense echoes in the optic disc and outer retinal layers with parafoveal thickening predominantly observed in the right eye.	Inferior hemifield defects	Vision recovered within 6 to 8 weeks.

Symptomatic disturbance in the visual fields secondary to the optic disc and macular area exhibit a good prognosis with a brief spontaneous resolution and possibly prevented or picked up early with renal related symptoms as described, subjecting the patient for OCT and HFA analysis in addition to laboratory serological tests. In the differential diagnosis, the importance of specificity of ocular manifestation of IgAN relates to the development of drusenoied deposits that appear similar to age-related macular degeneration, diabetes retinopathy, hypertensive retinopathy, and glaucomatous visual fields. However, the aforementioned clinical entities differ significantly in their clinical appearance and presentations.

The pathogenesis of IgA nephropathy involves an error in IgA1 glycosylation resulting in IgA1 secretion into the systemic circulation. The IgA1 forms complex deposits attached to the extracellular matrix and mesangial cells within the glomerulus. This induces mesangial cells to release pro-inflammatory mediators and activate the complement system via lectin and alternative pathways [3]. The previous study reported the presence of cotton wool spots, serous RD and choroidal infarcts on FFA Rueda-Rueda T, et al. reported an association of unilateral uveitis in IgA nephropathy [7,8]. IgA nephropathy in younger age group tends to manifest with macroscopic hematuria, compared to the older age group presenting with proteinuria, microscopic hematuria, and/or deposits in the glomerular mesangium with complement C3 deposition.

Conclusion

The new findings demonstrated in the present case study include optic disc drusenoid like deposits corresponding to the visual field defects significantly add on to the previous reports that discovered paramacular drusenoid-like characterizing the ocular manifestation of IgA nephropathy. Therefore in patients manifesting with visual disturbances and renal related symptoms, we recommend investigation of the optic disc and macular area by OCT and visual field evaluation on field analyzers [9-11].

Acknowledgment: We are thankful to the head of the department and institution for the continuous support and encouragement.

Disclosure: Authors declare that no disclosures exist.

References

1. Schena FP, Nistor I (2018) Epidemiology of IgA Nephropathy: A Global Perspective. Semin Nephrol 38: 435-442.
2. Berger J, Hinglais N (1968) Intercapillary deposits of IgA-IgG. J Urol Nephrol (Paris) 74: 694-695.
3. Lally DR, Baumal C (2014) Subretinal Drusenoid Deposits Associated With Complement-Mediated IgA Nephropathy. JAMA Ophthalmol 132: 775-777.
4. Cetin Nuran, Hikmet Basmak, Aylin Gencler, Mustafa Fuat Acikalin (2018) Perimacular drusenoid deposits in a child with IgA nephropathy. Canadian Journal of Ophthalmology 53: e71-e74.
5. Ehrhardt J, Gelisken F (2018) Acute, bilateral visual loss in a patient with IgA glomerulonephritis. Ophthalmologe 115: 222-225.
6. Taban M, Chand D, Sears JE (2006) Ocular findings in IgA nephropathy with renal failure and hypertension. J Pediatr Ophthalmol Strabismus 43: 378-380.
7. Rueda-Rueda T, JL Sánchez-Vicente, A Moruno-Rodríguezet, FE Molina-Socola, M Contreras-Díaz, et al. (2017) Unilateral uveitis associated with IgA nephropathy. Arch SocEsp Oftalmol 92: 598-601.
8. Sirbat D, Saudax E, Hurault de Ligny B, Bene MC, Raspiller A (1983) A new etiology of episcleritis: nephropathies with IgA and/or isolated C3 deposits. J Fr Ophtalmol 6: 921-925.
9. Endoh M, H Kaneshige, Y Tomino, Y Nomoto, H Sakai, et al. (1981) IgA nephropathy associated with myasthenia gravis and scleritis. Tokai J Exp Clin Med 6: 421-425.
10. Pavlin CJ, Easterbrook M, Harasiewicz K, Foster FS (1993)

An ultrasound biomicroscopic analysis of angle-closure glaucoma secondary to ciliochoroidal effusion in IgA nephropathy. Am J Ophthalmol 116: 341-345.

11. Hegde V, Mitrut I, Bennett H, Singh J (2009) Episcleritis: an association with IgA nephropathy. Cont Lens Anterior Eye 32: 141-142.