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ORIGINAL ARTICLE

Profile of retinal vasculitis in a tertiary eye care hospital

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ABSTRACT:

Aim: To provide a fact file on the etiology, clinical presentations and management of retinal vasculitis. **Materials and Methods**: A cross sectional Study was conducted in the Department of Ophthalmology. Demographic variables, risk factors, symptoms, clinical signs and visual acuity at presentation were studied. Examination tools used were Log MAR chart, slit lamp, direct & indirect ophthalmoscope. **Results:** 120 eyes of 100 patients with retinal vasculitis were studied. Among the 100 patientsstudied, 60(60%) were females and 40(40%) were males. The most common presenting symptom was defective vision seen in 70eyes (58.33%), followed by floaters in 45 eyes (37.5%). Other symptoms were pain reported in 23 eyes (19.17%), redness in 34 eyes(28.33%), photophobia in 24 eyes (20%) and flashes in 8 eyes (6.67%). Vascular sheathing was noted in 103(85.83)eyes making it the most common finding in retinal vasculitis eyes. Vitritis 65(54.17) and vascular sclerosis 50 (41.67) were other common findings. Vitreous hemorrhage 32 (26.67)was the most common type of hemorrhage noted in vasculitic eyes. Retinal neovascularization was seen in 42(35)eyes. Capillary nonperfusion (47%) was the most common angiographic finding followed by collaterals 24%. Macula was normal in 61 (50.5) eyes while it was not possible to comment on the macular status in 20 (16.67)eyes. Cystoid macular edema 10(8.33) epiretinal membrane 8 (6.67) and internal limiting membrane striae 7(5.84) were most common macular abnormalities noted. Majority of patients 37%) did not have any systemic illness. 47 %) patients had received oral steroids for retinal vasculitis. Out of 100 patients with retinal vasculitis, Mantoux test was positive in 40 (40%) but tuberculosis could be confirmed with X-ray chest and sputum examination for acid fast bacilli in only 8(8%) individuals. Serum angiotensin-converting enzyme (ACE) level was found to be raised above normal levels in 7(7%) patients and antinuclear antibody (ANA) was found in an equal 7 (7%) patients. **Conclusion**: Retinal vasculitis cases had similar clinical presentations and common treatment plan. There was no systemic disease association with vasculitis warranting a careful approachin prescribing investigations.

Key words: Angiotensin-converting enzyme, antinuclear antibody, laboratory investigation, retinal vasculitis

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INTRODUCTION

Retinal vasculitis may occur as a primary syndrome called idiopathic retinal vasculitis, which affects the eye vasculature without evidence of any systemic, or eye disease.[1] More commonly, it is seen as a manifestation of systemic diseases including sarcoidosis, collagen vascular diseases, malignancy, neurologic conditions and systemic diseases. It also occurs in ocular conditions like parsplanitis or birdshot retinochoroidopathy. The various stages of disease can be described as stage of inflammation, stage of ischemia, stage of neovascularization and stage of complications.[2]

Inflammation of peripheral retinal vessels may be completely asymptomatic even in patients with associated systemic disease. They often complain of painless loss or blurring of vision. Areas of retinal infiltrates or haemorrhage can cause scotomata or floaters. [2]

The major hurdles in the management of retinal vasculitis are its nonspecific clinical manifestation and obscure etiology.[3]Apart from infective, obstructive and neoplastic retinal vasculitis, which can be diagnosed based on serial ophthalmological examinations and systemic features; most of the cases of retinal vasculitis which are secondary to systemic inflammation and those of primary category have indiscriminate clinical presentations making it difficult to pinpoint the etiology based on clinical examination alone.[3]Tailored laboratory investigations have been propounded as the only way to find out the etiology of such cases of retinal vasculitis.[4] Retinal vasculitis also shows considerable geographical variation.[5] While Eales' disease is reported in one in 200 to 250 ophthalmic patients in Indiait is a rarity in developed world.[6] Similarly Behcet's disease which is uncommon in Indian population is seen predominantly in Mediterranean region and Japan.[7] Present study was done to analyze the patients with retinal vasculitis in a tertiary care center. We have tried toprovide a fact file up on the clinical manifestations, diagnosis and treatment of retinal vasculitis in the eastern region of country.

MATERIAL AND METHODS

A cross sectional study was conducted in the Department of Ophthalmology, after taking the approval of the protocol review committee and institutional ethics committee. After taking informed consent detailed history was taken from the patient or

the relatives if the patient was not in good condition. The technique, risks, benefits, results and associated complications of the procedure were discussed with all patients

INCLUSION CRITERIA FOR DIAGNOSIS OF RETINAL VASCULITIS

1. Predominantly peripheral retinal venous dilation, tortuosity, discontinuity or sheathing along with leakage of dye on fluorescein angiography
2. Predominantly peripheral retinal nonperfusion on fluorescein angiography along with venous tortuosity, dilation, discontinuity and sheathing
3. Retinal neovascularization along with predominantly peripheral venous dilation, tortuosity,discontinuity and sheathing
4. Recurrent vitreous hemorrhage along with predominantly peripheral venous dilation, tortuosity, discontinuity or sheathing

Age, gender, age of onset of disease, age at presentation and history of prior or present systemic illness were noted. Detailed scrutiny of presenting symptoms was done with regard to laterality. Best corrected visual acuity at presentation was noted from the records for eachpatient. Previous ocular treatment for vasculitis or other diseases was also recorded. Any ambiguity or missing information in the records was a criterion for exclusion from the study. Slit-lamp examination was done to look for anterior uveitis. Any other obvious finding like rubeosis, band shaped keratopathy was noted when ever encountered. Slit-lamp biomicroscopy was used to assess the macular status. Signs of retinal vasculitis were notedfor each patient from the standard fundus drawing made at the first visit. The parametersnoted were, vascular sheathing, sclerosis of vessels, vitritis, neovascularization, vitreous hemorrhage, status of the macula and choroidal pathologies. On fluorescein angiography the presence of capillary nonperfusion, collaterals, neovascularization and status of the macula were recorded. The positive results of the tailored laboratory investigations advised on the basis of history and clinical findings were recorded for each patient.

RESULTS

120 eyes of 100 patients with retinal vasculitis were studied. Among the 100 patients studied, 60(60%) were females and 40(40%) were males. most of the patients below 20 years age group and followed by above 40 years. Range of age of the patients was below -50 years (Table 1) with mean age at presentation as 30±10.85 years.

Table 1: Age and sex distribution of study participants

Age group(yrs.)	Males n=13	Females n=15	Total
Below -20	28	18	46
20-30	12	4	16
30-40	6	6	12
40-50	14	12	26
Total	60	40	100

Retinal vasculitis was bilateral in 65 (65%) and unilateral in 35 (35%) cases. 40 (61.53%) maleshad bilateral retinal vasculitis and 20 (30.77%) had unilateral disease; whereas in female group,30 (75%) and 10 (25%) subjects had bilateral and unilateral disease, respectively.

Table 2: Distribution of symptoms

Symptom	No. of eyes N=120	%
Defective vision	70	58.33
Floaters	45	37.5
Pain	23	19.17
Redness	34	28.33
Photophobia	24	20
Flashes	8	6.67%

The most common presenting symptom was defective vision seen in 70eyes (58.33%), followed by floaters in 45 eyes (37.5%). Other symptoms were pain reported in 23 eyes (19.17%), redness in 34 eyes (28.33%), photophobia in 24 eyes (20%) and flashes in 8 eyes (6.67%).

Table 3: Clinical findings in retinal vasculitis eyes

Signs	No. of eyes. N=120	%
Vascular sheathing	103	85.83
Vitritis	65	54.17
Sclerosed vessel	50	41.67
Neovascularization else where	42	35
Vitreous hemorrhage	36	30
Retinal hemorrhage	32	26.67
Anterior uveitis	14	11.67
Branch retinal vein occlusion	9	7.5
Subhyaloid hemorrhage	9	7.5
Pars plana membrane	5	4.17
Cataract	3	2.5
Rubiosis iridis	3	2.5
Glaucoma	2	1.67
Total retinal detachment	2	1.67
Band-shaped keratopathy	1	0.84

Vascular sheathing was noted in 103(85.83) eyes making it the most common finding in retinal vasculitis eyes [Table 3]. Vitritis 65(54.17) and vascular sclerosis 50 (41.67) were other common findings. Vitreous hemorrhage 32 (26.67) was the most common type of hemorrhage noted in vasculitic eyes. Retinal neovascularization was seen in 42(35) eyes. Capillary nonperfusion (47%) was the most common angiographic finding followed by collaterals 24%.Macula was normal in 61 (50.5) eyes while it was not possible to comment on the macular status in 20 (16.67) eyes [Table 3]. Cystoid macular edema 10(8.33) epiretinal membrane 8 (6.67) and internal limiting membrane striae 7(5.84) were most common macular abnormalitiesnoted.

Table 4: Macular findings

Within normal limits	61 (50.5)
No view	20 (16.67)
Cystoid macular edema	10(8.33)
Epiretinal membrane	8 (6.67)
ILM folds	7(5.84)
Macular edema	5(4.17)
Fibrovascular proliferation	2(1.67)
RPE defect	2 (1.67)
Others (Macualr hole, chorioretinal atrophy, choroidal neovascular membrane, scar and hard exudates plaque)	1(0.84%) each

Majority of patients 37%) did not have any systemic illness. 47 %) patients had received oral steroids for retinal vasculitis.

Table 4: Previous treatment for retinal vasculitis

Parameter	No. of patients=100	%
Oral corticosteroids	47	47
None	37	37
Laser photocoagulation	25	25
Periocular steroids	14	14
Surgery	11	11
Immunosuppressive#	5	5
Cryotherapy	2	2
Anti-VEGF	2	2
Antituberculous treatment	2	2

Out of 100 patients with retinal vasculitis, Mantoux test was positive in 40 (40%) but tuberculosis could be confirmed with X-ray chest and sputum examination for acid fast bacilli in only 8(8%) individuals. Serum angiotensin-converting enzyme (ACE) level was found to be raised above normal levels in 7(7%) patients and antinuclear antibody (ANA) was found inan equal 7 (7%) patients. Normal X-ray and computerized tomography scan of chest, normal serum lysozyme and serum and urinary calcium levels combined with evaluation by a pulmonologist refuted the diagnosis of sarcoidosis in patients with raised serum ACE levels.

Similarly negative anti-double stranded DNA

antibody and anti-Smith antibody along with assessment by a rheumatologist excluded systemic lupus erythematous in patients with positive serum ANA. 2 (2%) patients reported to us with a positive result for human leukocyte antigen B5 (HLA B5) marker in absence of oral, genital or cutaneous manifestation of Behcet's disease.

Corticosteroids were the mainstay of management of retinal vasculitis. 47 patients were treated with oral corticosteroids and 5 patients were administered immunosuppressives in the form of oral azathioprine 3, cyclosporine (1 and methotrexate 1 each. Out of 120 eyes, retinal laser photocoagulation was required in 25 eyes.

The mean follow-up period of retinal vasculitis cases was 17.4±5.2 months with a range of 12-24 months. 57 (47.5%) eyes gained one or more lines on Snellen's distant visual acuity chart whereas 25 (20.84%) eyes lost one or more lines. 38 (31.67%) eyes maintained their initial visual acuity through the available follow-up period.

DISCUSSION

Retinal vasculitis has always been an uncommon eye disease which has the potential of inflicting significant visual morbidity.[8] Complicating the successful management of these cases is the fact that most of the cases of retinal vasculitis have elusive etiology.[9,10] The main dilemma in management of retinal vasculitis is to identify whether the etiology was infectious or non-infectious, as their managements are completely different.[8] Control of the intraocular inflammation is sufficient in noninfectious cases but infectious retinal vasculitis needs an appropriate antimicrobial therapy alongside anti-inflammatory and/or immunosuppressive therapy.[11] On the other spectrum of etiology of retinal vasculitis are the cases associated with systemic immunological disease conditions. Onset of retinal vasculitis in these cases heralds worsening of the systemic disease making identification of the systemic vasculitic entity necessary.[11] Still there is another subgroup of retinal vasculitis patients who do not provide any positive clue on history and clinical examination and have negative laboratory investigations. Such cases of primary retinal vasculitis are the majority and are often administered multitude of laboratory investigations, yielding no confirmatory result.[9,10,12] We have found that all the patients included in this study were cases of primary retinal vasculitis

Majority of patients with retinal vasculitis visiting our center had bilateral disease at presentation. This finding is in keeping with that of Saxena et al., who have studied 159 cases of Eales' disease in India.[13] Male preponderance and clustering of cases in below 20 years, similar to previous reports.[12,14] We have found that dimness of vision was the most common symptom of retinal vasculitis in our study population. Vascular sheathing was found to be the most common sign of retinal vasculitis in contrast to vitreous hemorrhage which was found tobe the most common presenting feature by Saxena et al. [13] This difference might be due to the fact that all cases of primary retinal vasculitis are not Eales' disease, which has been considered as a specific disease entity.[13] In our group of patients vitreous hemorrhage was the less common than vitreitis, vascular sclerosis and neovascularization else where Capillary nonperfusion was the most common angiographic marker of vasculitis. Assessment of the systemic history, clinical examinations and evaluation of the tailored laboratory investigations revealed that none of patients had any incriminating systemic etiology for retinal vasculitis. In our study population Mantoux test positivity n=40(40%) was the foremost finding as far as positive results were concerned. However none of these cases had signs or symptoms of active pulmonary tuberculosis. Habibullah et al. have studied the significance of Mantoux positivity in tuberculous retinal vasculitis and have found no statistically significant association between them.[15] Similarly 7 cases which reported to us with raised serum ACE levels had normal chest X-ray and serum and urinary calcium and serum lysozyme levels. None of these patients had keratic precipitates, snow ball opacities or chorioretinal nodules needed for diagnosis of ocular sarcoidosis as elaborated in the International Criteria for Diagnosis of Ocular Sarcoidosis.[16] Single positive laboratory finding in absence of compatible uveitis was insufficient for the diagnosis of probable or possible ocular sarcoidosis which require at least two positive laboratory findings and compatible uveitis in absence of lung biopsy and bilateral hilar lymphadenopathy for diagnosis.[16,17] Patients with raised ANA levels neither had anti-double stranded DNA antibody or anti-Smith antibody nor were positive for Hepatitis B or C which could have confirmed the diagnosis of lupus vasculitis or hepatitis, respectively. 2 patient had reported to us with positive HLA B5 marker in absence of oral, genital or cutaneous manifestation of Behcet's disease.[18] HLA B5 marker has been reported to be present in about 6% healthy Indians.[19] Such positive laboratory findings in absence of clinical features and confirmatory markers of the suggested disease were assigned as false positive results by George et al., who reported it to be over 20% of all retinal vasculitis cases.[12] They have followed 25 such patients of retinal vasculitis for 4-year duration, only to find that barring one patient, who had developed systemic lupus; none of them had developed the disease, initially pointed out at retinal vasculitis work-up. This may hold true for present study also. It also puts emphasis on the fact that prescription of laboratory investigation for retinal vasculitis should always be backed by positive leads on systemic history and clinical examination.[12]

In face of elusive etiology and the fact that there is no well defined guidelines for the management of retinal

vasculitis, the treatment of retinal vasculitis in present study was mainly palliative.[9] Corticosteroids were the mainstay of treatment and were used to control intraocular inflammation in eyes with vascular sheathing and vitritis. As suggested by Saxenaet al., in context with Eales' disease, laser photocoagulation was used for neovascularization at the disc and elsewhere and to the fibrovascular proliferations[13].

CONCLUSION

Present study has found that all the cases of retinal vasculitis visiting our center were primary retinal vasculitis in which no systemic disease association or infectious etiology could be ascertained after detailed history, clinical examination and tailored laboratory work-up. The finding that retinal vasculitis cases were primary in nature may lead to an approach where laboratory investigations are advised sparingly, based mainly on previous systemic history and clinical judgment. It also calls for a larger population-based study to know the prevalence of etiological factors associated with retinal vasculitis in this part of country.

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