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Retinopathy in Blood Dyscrasias: Case-Series And Review of Literature.

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Abstract
Purpose: To study incidence of retinopathy in various blood dyscrasias, in patients attending GGH, Guntur, for a period of 6 months.
Study design: Hospital based prospective study.
Methodology: *This is a study of 100 patients of various age groups with different types of hematological disorders for a period of 6 months from November 2015 to April 2016 at Government general hospital,Guntur.*
Results and Observations: *Out of 100 patients studied,with various blood dyscrasias, 47(47%) patients had hematological malignancies; 48 (48%)patients with anemia and 5(5%) patients had platelet disorders. Among 100 patients only 42 patients had retinopathy, out of which, leukemic retinopathy in 24 patients, anemic retinopathy in 15 patients and retinopathy of bleeding disorder in 3 patients.*
Conclusion: *In study conducted in GGH, Guntur, for a period of 6 months , Patients with different blood dyscrasias of different types viz. anemia,leukemia,lymphoma and bleeding disorders were examined.Complete ophthalmic examination and blood counts were made.Amongst 100 patients, 42 patients showed retinopathy.Out of which, leukemic retinopathy was seen in 24 patients, anemic retinopathy identified in 15 patients and retinopathy of bleeding disorder accounts in 3 patients.Retinopathy had higher incidence in males with 61% compared to 39% in females. Patients with age <18 years had lower rates of retinopathy than those above 18 years. Patients with leukemic retinopathy had hemorrhages predominant than those with anaemic retinopathy. While,anaemic retinopathy patients had pale discs, cotton wool spots predominat than those of leukemic retinopathy. As incidence of bleeding disorders is low in our study 3%, reliable changes could not be commented.*
Keywords: *flame shaped hemorrhages, subhyaloid hemorrhages,white centered haemorrhages, cotton wool spots.*

I. Introduction

Blood dyscrasias are group of disorders indicating pathology of blood like increase or decrease in redblood cells number or size, variation in protein content of blood. Interestingly, disorders of the various cellular components of the blood often overlap. Retina as a part of systemic disorders is involved in these disorders. So,ocular fundus provides an unequaled direct view of the hematologic effects of blood dyscrasias in the living patient.

Materials And Methods:

In a period of 6 months, we have studied 100 patients, attending Out-Patient Department of ophthalmology, in patient department of General medicine, Pediatrics, and Obstetrics and Gynaecology.
After taking detailed history, every patient is being examined in the following way, general physical examination, visual acuity, slit lamp examination for anterior segment, IOP measurement by Goldmann applanation tonometry,gonioscopy, posterior segment examination using +90D, +78D lens, direct ophthalmoscope,indirect ophthalmoscope, for visualization of retina for any haemorrhages, cotton wool spots, retinal edema, disc abnormalities, and fundus camera for documenting the changes seen. Estimation hemoglobin level by colorimetric method, peripheral blood smear for complete blood picture.

Inclusion Criteria:
1.Males, females and children of all ages.
2.All blood dyscrasias.

Exclusion Criteria:
1. Associated systemic diseases like diabetes,hypertension,renal disease etc..
2. Patients with debilitating diseases
3. Associated other ocular diseases, like eale's, CRVO, CRAO etc..

4. Associated trauma.

Results: 200 eyes of 100 patients with various blood dyscrasias were seen.
Among 100 patients 31 were females, 47 were males, 22 were below 18 years of age.

	Males	Females	total
<18 years	14	8	22
Above 18	47	31	78
	61	39	100

Incidence Of Blood Dyscrasias:
Hematological malignancies:47 patients (47%)
Anemia:48(48%)
Bleeding disorders:5(5%)

Out of the total of 47 patients with hematological malignancies, maximum number suffered from acute leukemias (26 patients),out of which Acute Myeloid Leukemia was the largest group, with 15 patients, 11 patients suffered from Acute Lymphoblastic Leukemia. Chronic leukemia was found in 16 patients, out of which 12 patients were with CML, 4 were with CLL . Lymphomas were diagnosed in 5 patients.

Disease	No: of patients
AML	15
ALL	11
CML	12
CLL	4
lymphoma	5
Total	47 patients

Amongst the 48 patients belonging to the group of anemia, 23 patients were diagnosed with iron deficiency anemia. 10 patients suffered from different thalassemias, whereas 4 patients were diagnosed with aplastic anemia. Megaloblastic anemia accounted for 4 patients while the diagnosis for 5 patients was hemolytic anemia and another 2 patients suffered from anemia of chronic disease(AOCD).

Disease	No:of patients
Iron Deff.Anemia	23
Thalassemia	10
Aplastic anemia	4
Megaloblastic anemia	4
Hemolytic anemia	5
AOCD	2

Among 5 patients with platelet disorders, 4 patients are with ITP(idiopathic thrombocytopenic pupura) and a single patient is with hemophilia.

Disease	No:of patients
ITP	4
Hemophilia	1

Incidence Of Retinal Findings In Blood Dyscrasias: Out of 47 patients with hematological malignancies 24 patients showed retinopathy.Among 48 anemia patients,15 had retinopathy.In 5 patients of bleeding disorders 3 showed retinopathy.

Disease	No:of patients examined	No:of patients with retinopathy
Hematological malignancies	47	24
Anemia	48	15
Bleeding disorders	5	3
Total	100	42

Distribution Of Changes In Retinopathies.

Out of 24 patients of retinopathy of malignancies , 6 patients had pale disc, 17 patients had hemorrhages, 10 patients had cotton wool spots, 8 patients showed hard exudates, 11 patients showed tortuosity of vessels. All these findings often overlaped.

Among 15 patients with anemic retinopathy 8 patients had disc pallor,6 patients had hemorrhages, 11 patients showed cotton wool spots, 6 patients showed hard exudates,9 patients showed vessel tortuosity. All these findings often overlaped.

Among 3 patients with retinopathy of bleeding disorders, all three of them showed hemorrhages, one patient showed vessel tortuosity and hard exudates. None of them showed pale disc or cotton wool spots.

Disease	No:of pt's with retinopathy	Pallor of disc	Heamorrhages	Cotton wool spots	Hard exudates	Dilated tortuous viens
Hematological Malignancies	24	6	17	10	8	11
Anemias	15	8	6	11	6	9
Bleeding disorders	3	-	3	-	1	2

*Hemorrhages- pre-retinal, intra-retinal, flame shaped, deep, white centered hemorrhages, hemorrhages without white centre.

The typical ophthalmoscopic findings in the various blood dyscrasias are not pathognomonic and may actually be observed in many different local and systemic diseases involving the eye (i.e., diabetes, hypertension, collagen vascular disease). However, the pattern and distribution of the retinal findings in blood dyscrasias are characteristic. Identification of these characteristic findings on ophthalmoscopy,further ancillary testing may reveal a blood dyscrasia and allow for early referral for treatment by the appropriate medical subspecialist.

II. Discussion

The word *dyscrasia* comes from the Greek language and means "bad temperament." In the older medical literature, the term dyscrasia was used to indicate disease. Currently, we use the phrase *blood dyscrasia* to indicate a pathologic condition of the blood, usually when referring to disorders of the cellular elements of the blood. An increase or decrease in the total number of red blood cells in a given patient is referred to as polycythemia or anemia, respectively. A large population of atypical or neoplastic white blood cells within the blood constitutes leukemia. A subnormal number of platelets in the circulating blood (thrombocytopenia) or loss of normal platelet function can lead to bleeding disorders or coagulopathies. Interestingly, disorders of the various cellular components of the blood often overlap (i.e., leukemia, anemia, and thrombocytopenia).

Anemia:

Retina reveals haemorrhages,cotton woolspots, retinal edema. Haemorrhages may be Superficial flame shaped ,white centered, Pre retinal, Vitreous haemorrhage, deep haemorrhages without white centre. Retinal vascular changes like pale arterioles, tortuous dilated veins. FFA may show increased arm- retinal transit time.Cotton wool spots, macular star. Vision is usually normal, (decreased only in lesions at macula). White centered haemorrhages are more common in pernicious anemia[(1).] preretinal haemorrhages, white-centered haemorrhages more common in aplastic anemia[(2).] Moderate to severe iron deficiency anemia reveals retinal arterial occlusive events, branch retinal artery occlusion. Fanconi anemia presents with widespread vasculopathy, areas of central and peripheral leakage, areas of nonperfusion and neovascularization. Angioid streaks more commonly found familial dyserythropoietic anaemiatype III[(3).] Malaria when associated anemia,have high incidence of retinopathy, cerebral malaria without anemia also shows fundus changes, and the severity is the predictor of worse outcome. Retinopathy is more significant with Hemoglobin concentration < 6 g/dL, hematocrit value <45. For given hemoglobin concentration ,retinal hemorrhages are more common with more severe thrombocytopenia. more profound the anemia more cotton wool spots are found than hemorrhages. Hemorrhages are more common in advancing age, and in males for given hemoglobin concentration. Optic neuropathy may occur which presents as centro-ceacal scotoma in visual field analysis.

Leukemia

- **Acute lymphocytic**
- **Acute myelocytic**
- **Chronic lymphocytic**
- **Chronic myelocytic**

Retina in leukemia shows vascular changes including venous dilatation,tortuosity, irregularity, abnormal colour and sheathing.Haemorrhages may be of various types like flame shaped, white-centered,

preretinal and Vitreous haemorrhage.cotton wool spots, leukemic infiltrates which are white clumps or masses are found in the retina. Microaneurysms, venous occlusions, neovascularisation are more common in chronic leukemia. Serous detachment of retina occurs due to choroidal infiltration.Choroidal deposits in chronic leukaemia may give rise to a 'leopard skin' appearance. Changes are more common with Myeloid leukemia than lymphoid leukemia. Microaneurysms are found in mid periphery due to hyperviscosity. Neovascularization is seen in chronic leukemia associated with capillary closure.The white centers in hemorrhages consists of *leukemic cells and debris, platelet-fibrin aggregates, septic emboli*[(4).] Alterations in RPE include pigment clumping or a reticular pattern of pigment change. On Pathologic examination, leukemic infiltration of the choroid and retina, with destruction and hyperplasia of RPE were identified. Central Serous Retinopathy is more common in ALL[(5).] Leukemia associated with HTLV-1 shows pigmentary retinopathy predominantly. Progressive, widespread retinal degeneration resembling that of retinitis pigmentosa. sectoral or regional atrophy of the retina and choroid were found which is nonprogressive. Deep retinal and subretinal infiltrates, anterior uveitis,vitreous infiltration, and uveitis with retinal perivasculitis were also found[(6).]Optic nerve infiltration causes swelling and visual loss.

III. Polycythemia

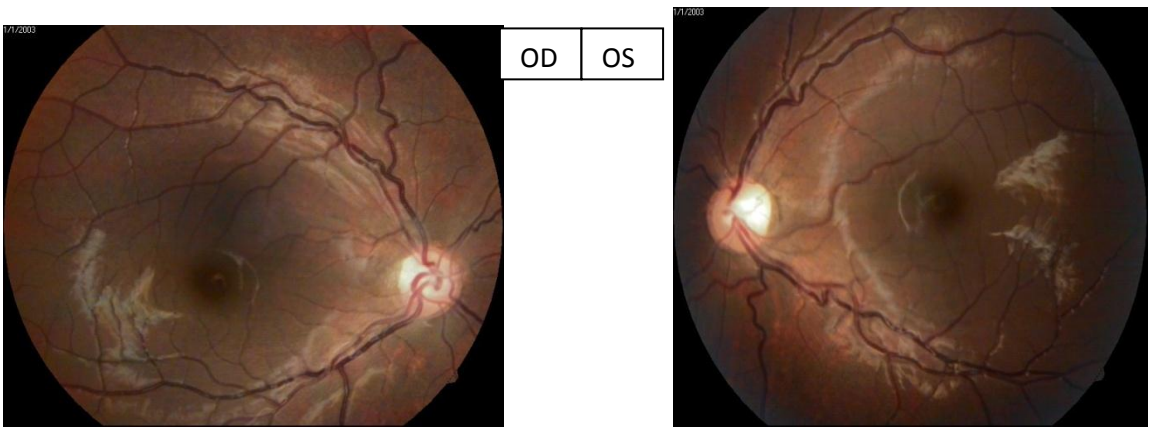
clinical signs of hyperviscosity are more predominant with HCT> 50%. Haematocrit maintains linear relation with viscosity upto 50%, with further increase in HCT, viscosity increases exponentially.retinal changes more common in viens dark,dilated,tortuous. Optic disc is usually hyperemic and swollen.Intra retinal hemorrhages,retinal edema are predominant. Bilateral vein occlusion in polycythemia secondary to Eisenmenger syndrome has been reported[(7).] vision is affected due to retinal edema or vein occlusion.

IV. Dysproteinemias

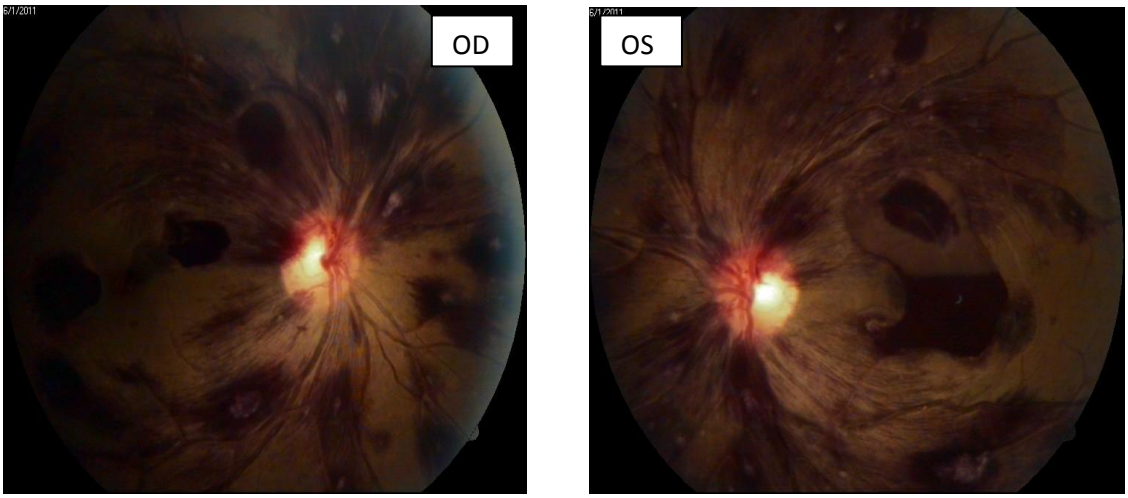
Dysproteinemias include cryomacroglobulinemia, hypergammaglobulinemia, Waldenström's macroglobulinemia, CLL with macroglobulinemia, multiple myeloma with hyperglobulinemia. Waldenström's macroglobulinemia may be the most likely dysproteinemia to produce retinopathy,possibly because the large size of the protein molecules in this condition lead to very high viscosity levels.[(8)]retinal veins- dark, dilated, and tortuous,intraretinal hemorrhages, cotton wool spots ,disk -hyperemic. Retinal edema ,Retinal microaneurysms are seen in chronic cases.Neovascularization of the retina or iris with VH, NVG,fibrous proliferation.Paraneoplastic retinopathy in a patient with Waldenström's macroglobulinemia has been reported with findings of deteriorating vision, declining electroretinogram and presence of serum antibodies against photoreceptor proteins[(9)].Cases of serous detachment of the retinal pigment epithelium and neurosensory retina have been reported in patients with paraproteinemia. [(10,11)] .Purtscher-like retinopathy is seen with hepatitis C-associated cryoglobulinemia.[(12)]

V. Hemorrhagic Disorders

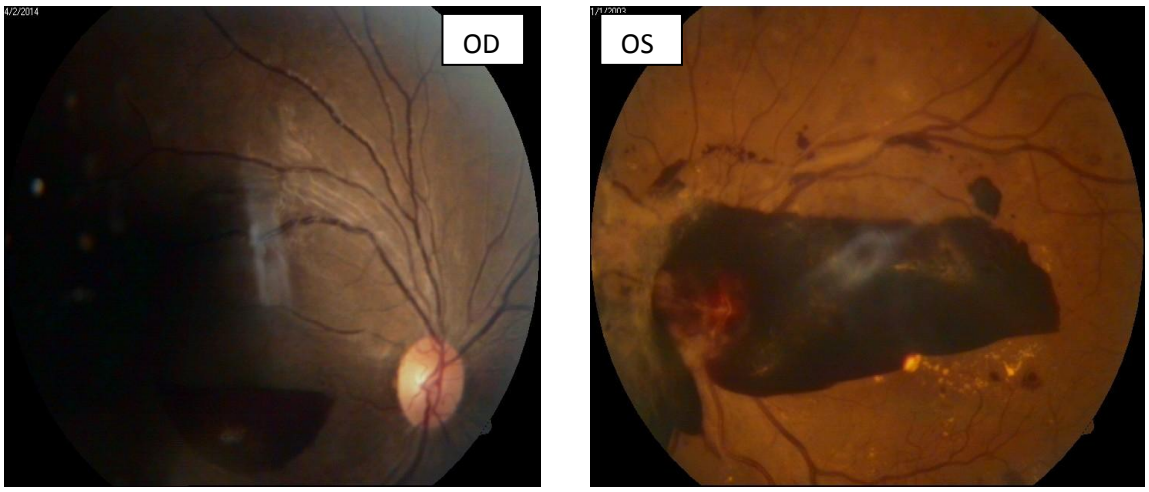
Spontaneous hemorrhage is more common after ocular trauma. Thrombocytopenia is an exception spontaneous with,Idiopathic thrombocytopenic purpura,rarely results in retinopathy.Thrombotic thrombocytopenic purpura presents with retinal hemorrhages and serous retinal detachment. Retinal changes are predominant when ITP is associated with thrombosis, hypertension,and renal disease,fundus shows Purtscher's like retinopathy in thrombotic thrombocytopenic purpura[(13)].



Retinal changes in anemia, showing pale optic disc,dilated tortuous viens, ischemic areas along superior and inferior arcuate vessels,edema of background retina.



Retina in patient with chronic myeloid leukemia showing normal optic disc,with vessel tortuosity in right eye, background retina showing multiple hemorrhages viz, flame shaped, deep retinal, white centered hemorrhages, subhyaloid hemorrhage in left eye.



Retina of patient with idiopathic thrombocytopenic purpura, showing subhyaloid hemorrhage in right eye inferior to macula,diffuse subhyaloid hemorrhage in left eye involving optic disc and macula in left eye,multiple deep retinal hemorrhages, with hard exudates in left eye.

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