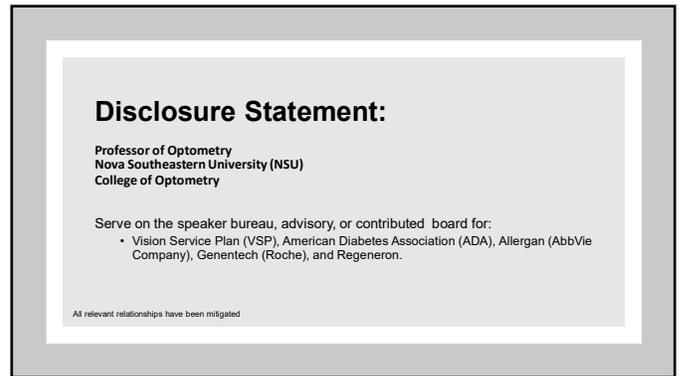


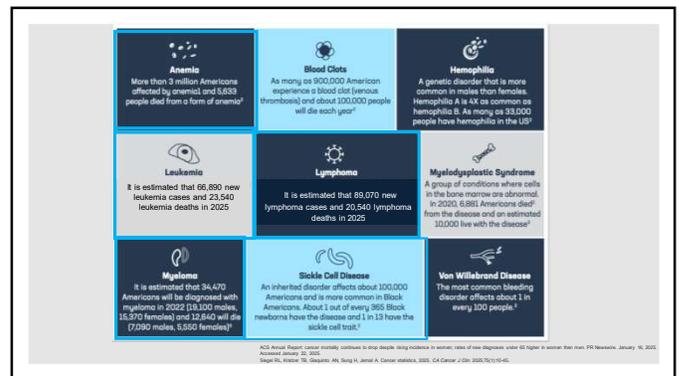
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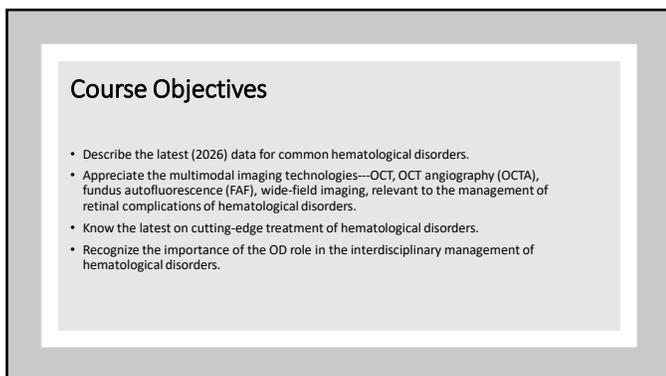
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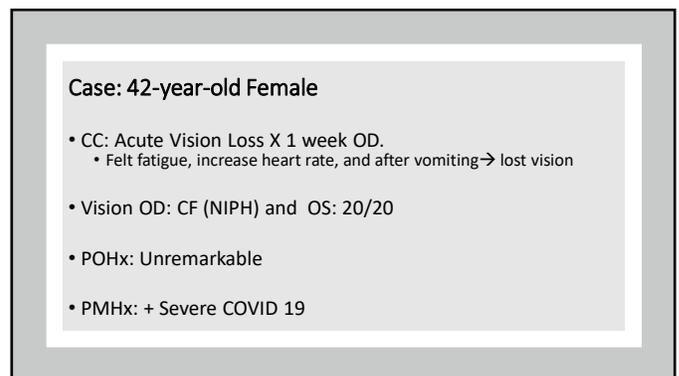
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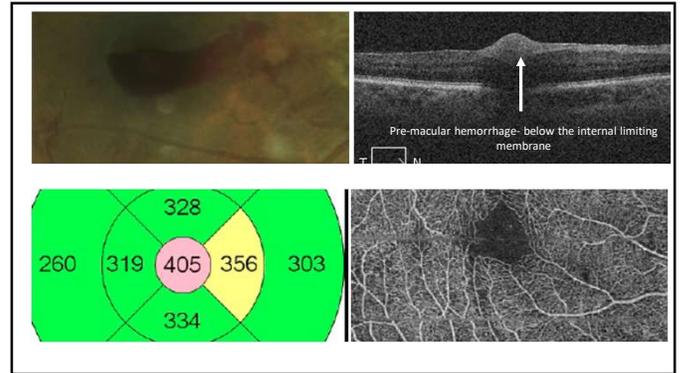
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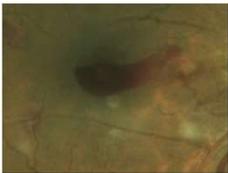
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8

Valsalva Maculopathy (VM)

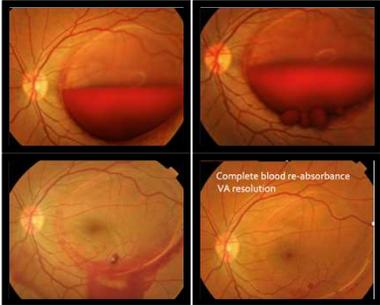
- **Valsalva Retinopathy**- increase in intrathoracic or intra-abdominal pressure → elevation in ocular venous pressure → spontaneous rupture of retinal capillaries → macular hemorrhage
 - Weightlifting, coughing, sneezing, vomiting/straining, **vomiting**, trauma (compression injuries)
 - Anticoagulant drugs, aspirin,
 - Medical work-up is necessary (RBC, WBC, platelets, and coagulation factors)
 - **Severe Anemia**
- Macular hemorrhage (pre-retinal or subhyaloid or intravitreal space)



9

Valsalva Maculopathy (VM) Management

- Most observed
- Yag membranectomy (posterior hyaloidotomy)
 - Laser induced hole into hyaloid → blood drains into vitreous



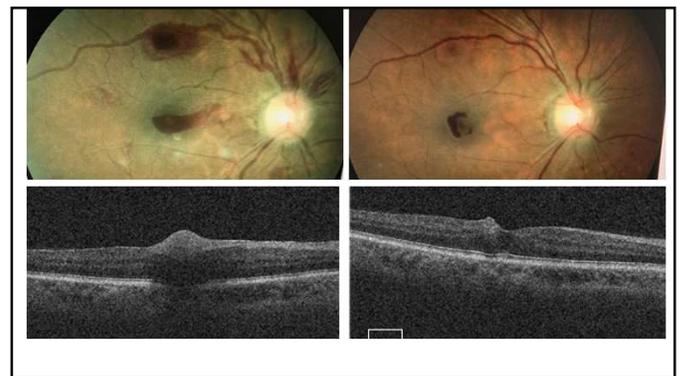
Acknowledgment: Mohamed A. Tawfik MD, FRCOphth
sourced from the Retina Image Bank

10

<table border="1" style="width: 100%; border-collapse: collapse;"> <tr><td style="width: 50%;">WBC</td><td style="width: 50%;">1</td></tr> <tr><td>Hb</td><td>8</td></tr> <tr><td>Hb (Absolute)</td><td>0.1</td></tr> <tr><td>Hematocrit</td><td>14.8</td></tr> <tr><td>Monocytes/Absolut</td><td>0.2</td></tr> <tr><td>WBC</td><td>4.0</td></tr> <tr><td>Hemoglobin</td><td>3.4</td></tr> <tr><td>MCV</td><td>74</td></tr> <tr><td>MDA</td><td>19.4</td></tr> <tr><td>RBC</td><td>144</td></tr> <tr><td>Platelets</td><td>119</td></tr> </table>	WBC	1	Hb	8	Hb (Absolute)	0.1	Hematocrit	14.8	Monocytes/Absolut	0.2	WBC	4.0	Hemoglobin	3.4	MCV	74	MDA	19.4	RBC	144	Platelets	119	<table border="1" style="width: 100%; border-collapse: collapse;"> <tr><td style="width: 50%;">Hemoglobin (Hgb) Solubility</td><td style="width: 50%;">Negative</td></tr> <tr><td>Iron and TIBC</td><td></td></tr> <tr><td>COLLECTION DATE</td><td>10/11/2021</td></tr> <tr><td>Order Date</td><td></td></tr> <tr><td>Result Date</td><td></td></tr> <tr><td>Ordering Physician</td><td></td></tr> <tr><td>Iron Saturation</td><td>2</td></tr> <tr><td>Iron</td><td>45</td></tr> <tr><td>Iron Status</td><td>9</td></tr> <tr><td>Iron (Total Cap (TIBC)</td><td>474</td></tr> <tr><td>Iron (Total Cap (TIBC)</td><td>250-480 ug/dL</td></tr> <tr><td>TIBC</td><td>485</td></tr> <tr><td>Tartrate, Serum</td><td>L</td></tr> <tr><td></td><td>(15-150 ng/mL)</td></tr> </table>	Hemoglobin (Hgb) Solubility	Negative	Iron and TIBC		COLLECTION DATE	10/11/2021	Order Date		Result Date		Ordering Physician		Iron Saturation	2	Iron	45	Iron Status	9	Iron (Total Cap (TIBC)	474	Iron (Total Cap (TIBC)	250-480 ug/dL	TIBC	485	Tartrate, Serum	L		(15-150 ng/mL)
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Tartrate, Serum	L																																																		
	(15-150 ng/mL)																																																		

Diagnosed with: Severe Iron deficiency anemia and thrombocytopenia
 ***Anemia is defined as hemoglobin less than 13 g/dL for men and less than 12 g/dL for women

11



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Anemias

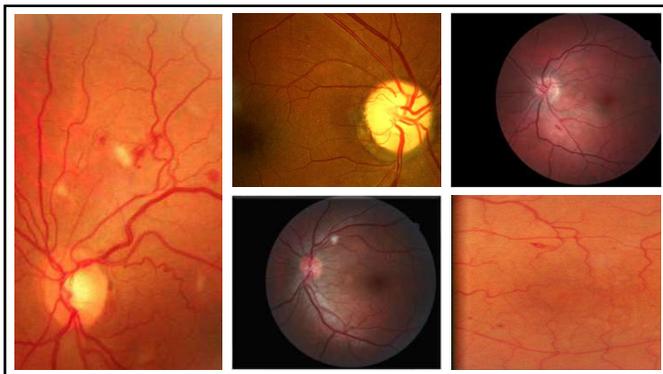
- **Iron-deficiency- most common**
 - Major component of transporting Hemoglobin oxygen
- **Vitamin Deficiency**
 - *necessary vitamins for RBC's production*
 - **Pernicious anemia** (Vitamin B12 anemia)
 - Lack intrinsic factor (IF) in gastric mucosa to absorb vitamin B12
 - Folate Deficiency Anemia
- **Anemia of chronic diseases- second most common**
 - Cancers, Autoimmune disease, HTN, diabetes, infections, and other conditions.

13

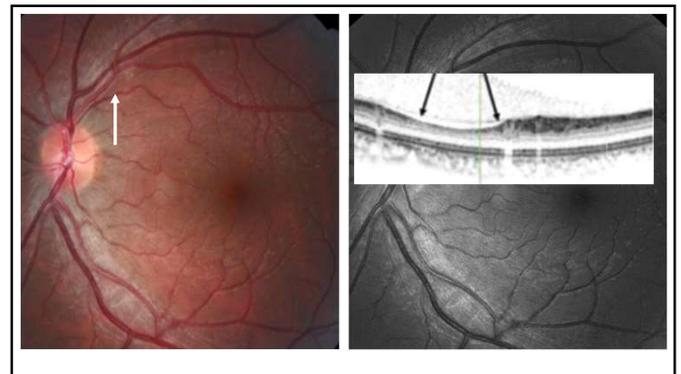
Anemia affects roughly **1 in 4** people *worldwide*, recent U.S. data shows that nearly **1 in 4 (about 22–30%)** American adults may have inadequate iron intake or deficiency

Tawfik YMK, Billingsley H, Bhatt AS, et al. Absolute and Functional Iron Deficiency in the US, 2017. 2020. JAMA Netw Open. 2024;7(5):e243326

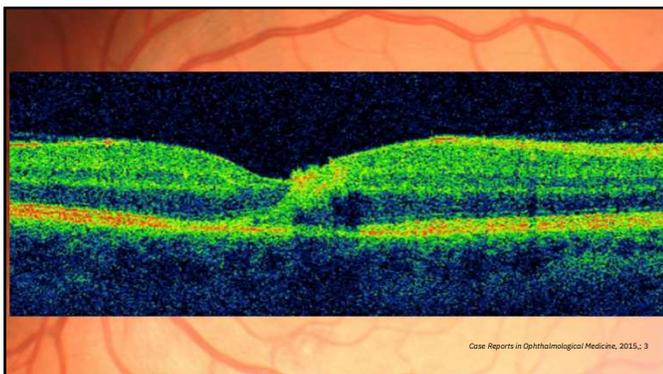
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15



16



17

Roth Spot Hemorrhages

- Extravasation of red blood cells follows vessel rupture with subsequent activation of the clotting cascade and formation of a platelet-fibrin plug at the site of the damaged endothelium.
- Histological studies revealed the white lesions are composed primarily of platelet-fibrin thrombi
- Endocarditis (Septic Emboli)
 - Bacterial
 - Fungal

18



19

ANEMIA KEY TAKEAWAY

- Prevalence of retinopathy among anemic patients ~ 30%
 - Anemic retinopathy is more likely to occur in patients with **severe** anemia or acute anemia
- Anemia and thrombocytopenia
 - Retinopathy more common (40%- 70%)
 - Particularly when the hemoglobin (Hb) level is below 6 gm/dL
- Retinopathy resolves with treatment of the underlying condition

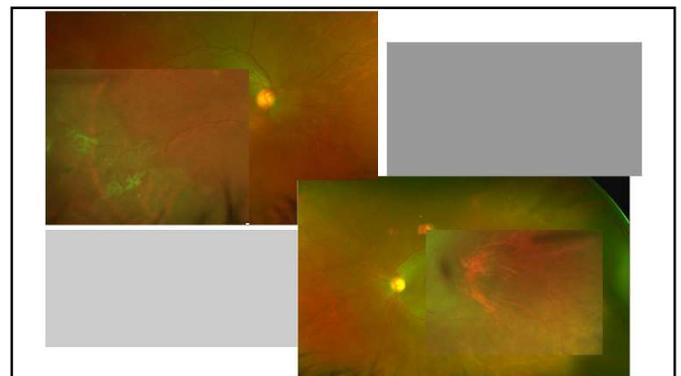
Prevalence of retinopathy in patients with anemia or thrombocytopenia. Eur J Haematol. 2001;67(4):238-244.

20

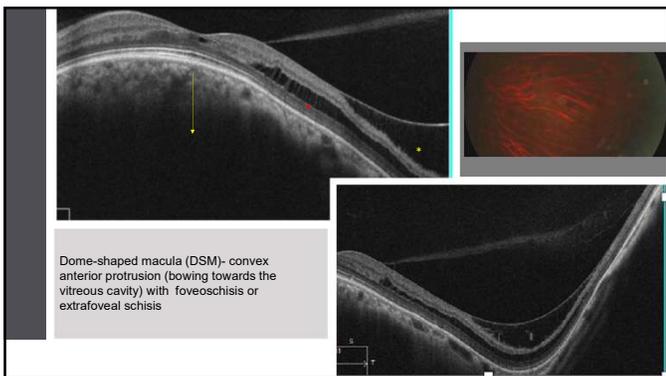
Case: 50-year-old Female

- CC: Acute Vision Loss X 1 week OD
- Vision OD: CF (NIPH) and OS: 20/20
- POHx: Stable SCR OU
- PMHx: Sickle-cell C disease (SC)

21

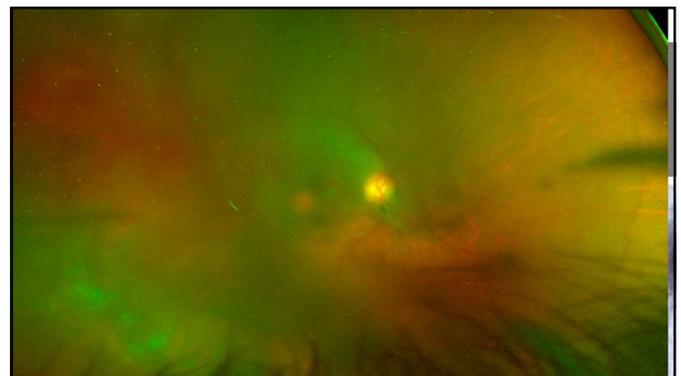


22



Dome-shaped macula (DSM)- convex anterior protrusion (bowing towards the vitreous cavity) with foveoschisis or extrafoveal schisis

23



24

Sickle Cell Disease (SCD) Update

- SCD has increased globally by 41.4 % (5.46 million to 7.74 million)
 - 100,000- 130,000 in the US have SCD
- The most prevalent genetic disorders in US
 - Autosomal recessive
 - 10-14% of African-Americans (1 in 365 Black or African American births)
 - Mediterranean ancestry
- Variants
 - HbSS, HbSC, HbS/ β -thalassemia, HbSA

Global, regional, and national prevalence and mortality burden of sickle cell disease, 2000-2021: a systematic analysis from the Global Burden of Disease Study 2021. Lancet Haematol. 2023; 1(8)

25

50-year-old Black Female - h/o stroke X 1month
 Sickle cell trait/ Hypertension/ Hypercholesterolemia/ aneurismal dilation of the ascending and thoracic aorta
 BCVA: 20/20 OD, 20/20 OS

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Risk factors for Sickling

Sickle Cell Today
 USA Comprehensive Sickle Cell Center

Volume 7 Issue 1 September 2016 National Sickle Cell Awareness Month September 2016

Sickle Cell Trait in Athletes

- Altitude**
 - Low oxygen tension
- Heat stress**
 - Dehydration
 - Acidosis
- Extreme physical exercise**
- Sustained maximal exertion**
- Concomitant diseases**
 - DM, HTN, CVD
- Pregnancy**

27

- The presence of Hyphema in Black patients under age 40 should raise the suspicion of underlying sickle cell disease
- Avoid carbonic anhydrase inhibitors (CAI's) -promote acidosis (sickling/outflow obstruction)

28

Non- Proliferative SCR	Proliferative SCR
<ul style="list-style-type: none"> • Venous tortuosity <ul style="list-style-type: none"> • peripheral vessels • Salmon Patch Hemorrhages • Black Sunbursts • Refractile deposit- • Angioid streaks • Dark Without Pressure 	<ul style="list-style-type: none"> • Stage 1. Peripheral Arteriolar Occlusion • Stage 2. Peripheral Arteriovenous Anastomoses • Stage 3. Neovascular and Fibrous Proliferations (Sea Fan formation) <ul style="list-style-type: none"> • Spontaneously regression (20-60%) due to auto-infarction • Stage 4. Vitreous Hemorrhage • Stage 5. Retinal Detachment

29

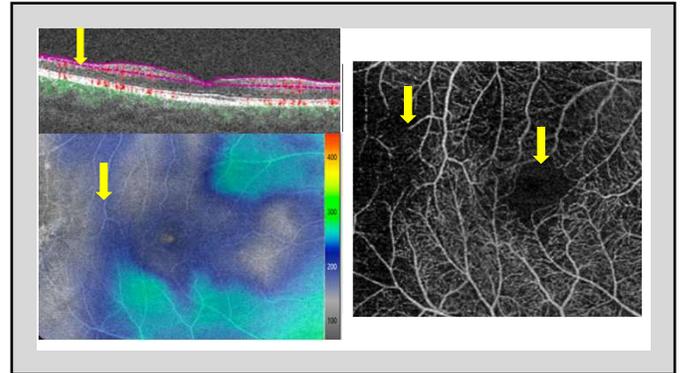


30

Name	Genetics	Systemic manifestations	PSR prevalence (Fekrat and Goldberg [7])	PSR prevalence (Dembélé et al. [8])	PSR prevalence (Bonanomi et al. [9])	PSR prevalence (Levezuel et al. [10])	PSR prevalence (Downes et al. [11])	
HbSS	Sickle cell anemia	Sickle cell homozygote	Most severe systemically	3%	5.2%	14.64%	18.1%	14%
HbSC	Sickle cell "C" trait	Sickle cell heterozygote, with another abnormal	Mild systemically	33%	12.4%	54.54%	54.6%	43%
<p>Sickle cell retinopathy (SCR) has been reported to develop in about 40% of individuals with SCD during the second decade of life.</p>								
HbSD, HbSE, HbSO	Sickle cell heterozygote, with another abnormal Hb allele	Varies systemically	-	-	-	-	-	-
HbAS	Sickle cell trait	Sickle cell heterozygote, with 1 normal Hb allele	Mild systemically	-	-	-	-	-

Graefes's Archive for Clinical and Experimental Ophthalmology volume 257, pages 1353-1364 (2019)
Rev Bras Hematol Hemoter. 2014; 36(5):319-21.

31



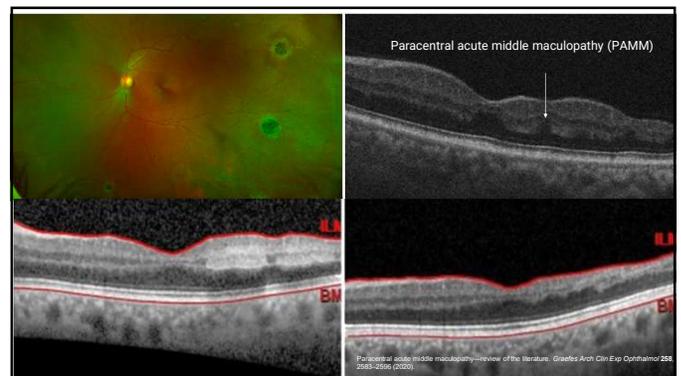
32

Sickle Cell Maculopathy (SCM)

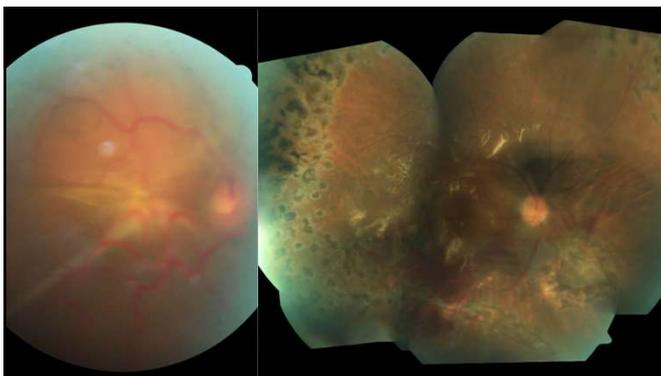
- Macular and peri-foveal temporal macular area
- Ischemia of the Superficial Capillary plexus (SCP) and Deep capillary plexus (DCP) and
- Symptoms
 - Asymptomatic
 - Paracentral scotoma
 - Metamorphopsia
 - Reduced contrast sensitivity
 - Color vision abnormalities

The image includes a schematic cross-section of the retina with labels for NFL (Nerve Fiber Layer), GCL (Ganglion Cell Layer), and IPL (Inner Plexiform Layer). Below it are two sets of capillary plexus images: 'Superficial Retina' and 'Deep Retina', showing the distribution of capillaries in different layers.

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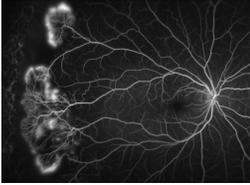
SCR/ SCM Treatment

- NPSCR
 - Follow-up every 3- 6 months
 - Salmon patch hemorrhage: 3 months
- PSCR
 - Stages 1-2: 3-6 months F/U
 - Stage 3-5: Laser Photocoagulation or retinal surgery. Peripheral circumferential retinal scattered photocoagulation (PCRP), PDT
 - Prophylactic Tx= not effective in preventing neovascularization
- SCM
 - Early complication of SCD
 - Studies have shown that SCM has been correlated to peripheral disease and may be a surrogate marker for peripheral retinopathy

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Leukemic Retinopathy

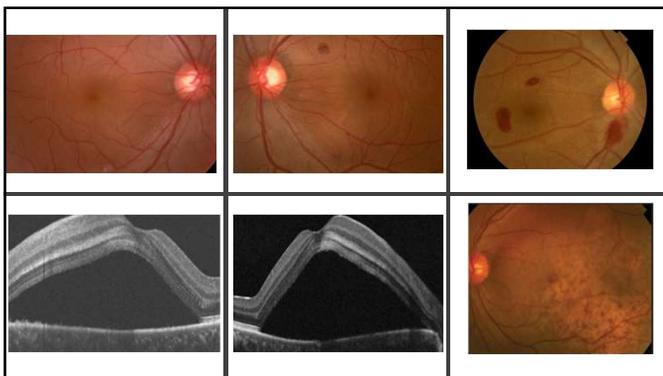
- Hemorrhages, exudates, CWS
- Retinal vein tortuosity and dilation
- Retinal leukemia infiltrates
 - gray-white sheathing
- Peripheral retinal microaneurysms and retinal neovascularization
 - Sea fans
- Direct infiltration of the ONH, Papilledema



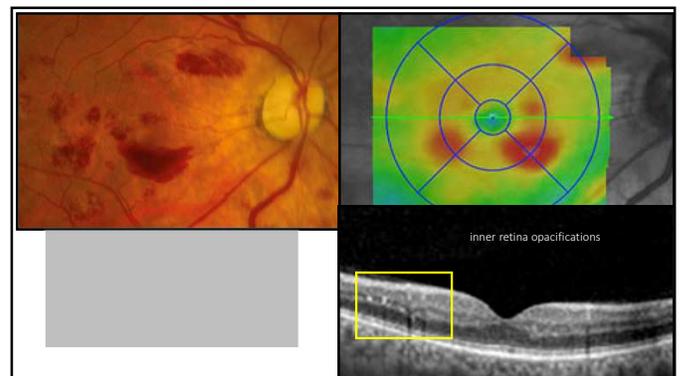
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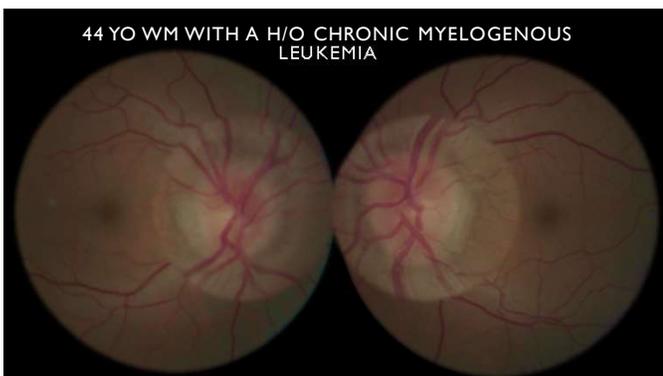
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Leukemic Retinopathy

- Leukemic Retinopathy
 - More common in Acute Leukemias
- Is an Ocular Emergency until proven otherwise
- Treatment
 - Systemic chemotherapy or direct radiation
 - Injection of subconjunctival chemotherapeutic agents
 - Bone marrow transplant

49

62-year-old Female

- CC: A growth on her eye for the past six-months
- Vision OD: 20/30 and OS: 20/30
- POHx: Cataracts OU
- PMHx: Hypertension and Hypercholesterolemia

50




Biopsy confirmed lymphoma

51

Lymphomas

- Classification:
 - Hodgkin Lymphoma
 - Multinucleated, giant cells called Reed-Sternberg cells
 - Non-Hodgkin Lymphoma(B-cells or T-cells)
 - More common
 - Most prevalent hematological malignancy
 - Approximately 3% to 4% of all cancer diagnosed annually

Shetty MK, Choudhary S, Loeffler KU. High-grade malignant B-cell lymphoma of the retina in a patient with concurrent gastric MALT lymphoma. *Graefes Arch Clin Exp Ophthalmol*. 2017 Mar; 245(3):448-50.

52

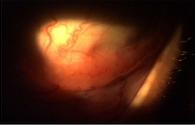
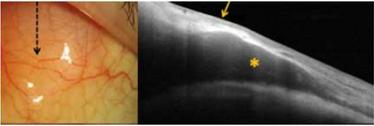
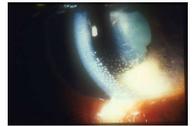
Ocular Lymphomas

- Hodgkin Lymphoma
 - Rarely causes ocular disease
- Non-Hodgkin Lymphoma (NHL)
 - The most common malignant orbital tumor
 - 55% of cases in adults
- Uveal form
 - Characterized by a non-resolving uveitis
 - Diffuse choroidal infiltration or exudative retinal detachment
- Vitreoretinal lymphoma (VRL)
 - Associated with primary central nervous system (PCNSL)

Westcott GC, Saini SS, Miller SE, Sloan MS, Pattenner A. Imaging of orbital lymphoproliferative disorders. *Radio Clin North Am*. Jan 1989;31(1):149-93.

53

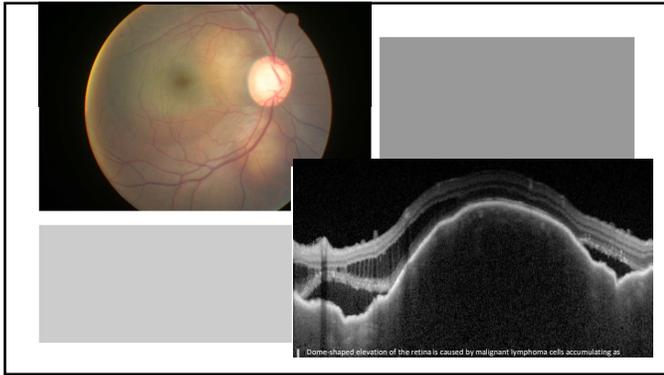


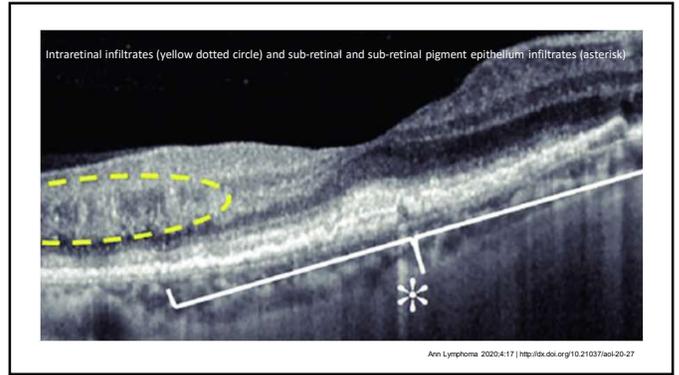
Eye and Vision volume 4, Article number: 22 (2019)

Clinical Pearl: Suspect lymphoma in Elderly patients with non-resolving uveitis.

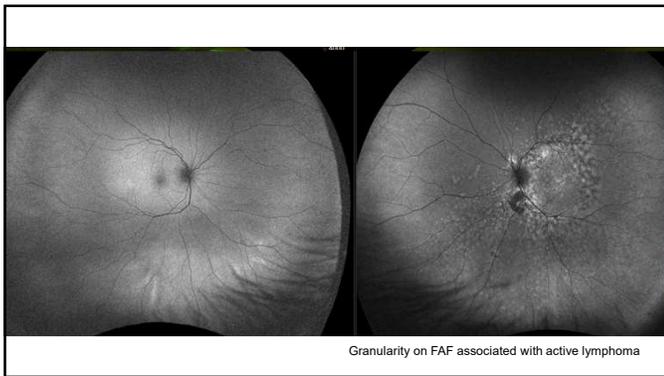
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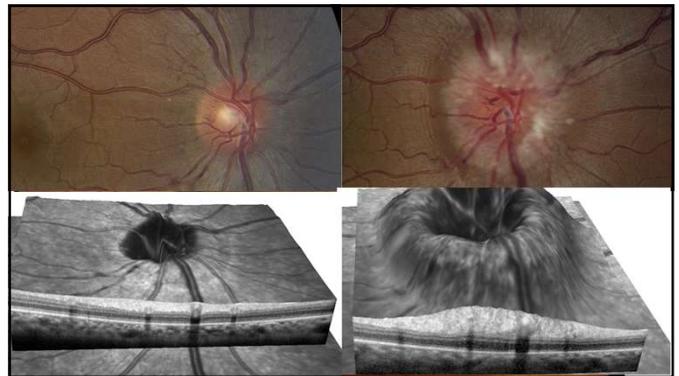
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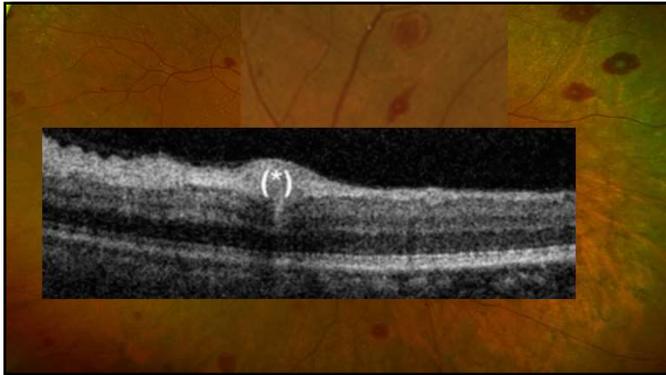
59

Table 1. Features of Various Types of Intraocular Lymphoma						
Lymphoma	Epidemiology	Laterality	Symptoms	Clinical Features	Subtype	Morphology
Primary Vitreoretinal Lymphoma	50-70 years	Frequently bilateral	Decreased vision Floaters	Vitreous cells Retinal/choroidal infiltrates CNS involvement	DLBCL	Large cells Minimal cytoplasm Prominent nucleoli
Primary Uveal Lymphoma	M-F 50-70 years	Usually unilateral	Decreased vision Metamorphopsia	Clear vitreous Diffuse choroidal thickening Exudative retinal detachment	EMZL	Small centrocyte-like cells with variable plasmacellular differentiation
Secondary Intraocular Lymphoma	Variable	Unilateral or bilateral	Decreased vision	Variable: Choroidal thickening Iris infiltrates Pseudohypopyon Vitreous cells	Dependent on systemic NHL	Similar to systemic NHL

M: males, F: females, DLBCL: diffuse large B-cell lymphoma, EMZL: extranodal marginal zone lymphoma, NHL: non-Hodgkin's lymphoma

Primary lymphoma of the central nervous system. Ophthalmol Clin North Am. 2005;18:199-207.

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61

Multiple Myeloma

- Plasma cells (WBC) are affected
 - usually of the IgG (60%) or IgA (25%) with a decrease in normal immunoglobulin synthesis
- The myelomic cells erode into the hard bone predisposing patient to pathologic fractures and hypercalcemia due to bone dissolution
 - Bone pain, anemia, and increased ESR strongly suggest myeloma...must be ruled out elderly patients
 - Predisposes patient to anemia and recurrent infections
- Increases with age, more common in males and Blacks

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Myeloma Ocular Complication

- Hyperviscosity retinopathy
- Retinal Microaneurysms
- Retinal Detachment
- Central Retinal Vein Occlusion
- Proptosis
- Conjunctival Crystalline Deposits

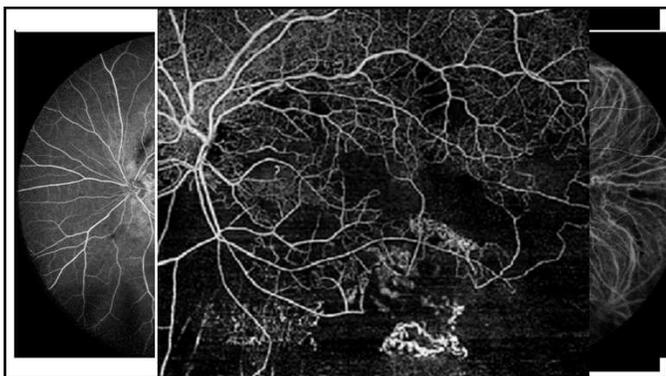
Retina Image Bank

63

Hematological Malignancies Key Takeaway

- Up to 50% of patients with leukemia may have retinal abnormalities, which is associated with poor prognosis.
- Detection of vitreoretinal lymphoma, characterized by diffuse choroidal infiltration or exudative retinal detachment, is critical in non-Hodgkin, diffuse large B cell lymphoma, as it indicates central nervous system involvement.
- The presence of retinopathy in patients with multiple myeloma has not been found to be associated with worse prognosis and can improve with systemic treatment.

64



65

... 21-32 sec

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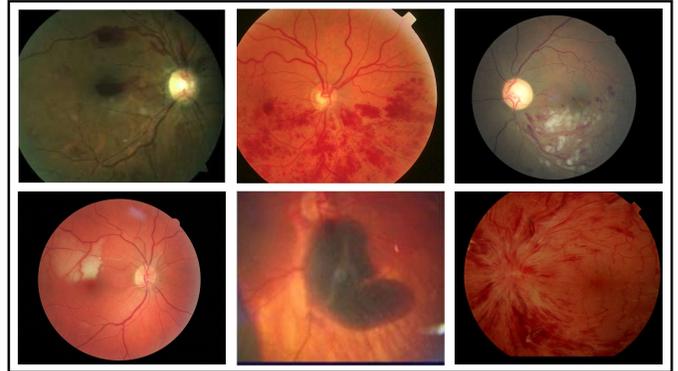
... 21-32 sec

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Platelets

- **Thrombocytopenia**
 - Low platelet count (<100,000)
 - Excessive bleeding
 - Normal platelet survival is 8-10 days, while only 1-3 days with thrombocytopenia
- **Thrombosis**
 - High platelet count/ abnormal function
 - Blood clots or emboli
 - Stroke (cerebral vascular accident), transient ischemic attack, pulmonary embolus and/or a heart attack

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Coagulation Monitoring

- Portable or POC in-office INR self-testing



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Conclusion

- Hematological disorders are a major threat to both systemic and visual health.
- Early detection, management and treatment is vital in preventing vision threatening and potentially saving lives.
- Optometry plays an important role

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Any Questions



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THANK YOU

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sherrolrey@gmail.com

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