A rare clinical presentation of Hyperviscosity syndrome

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BACKGROUND

Hyperviscosity syndrome (HVS) is a rare oncologic emergency occurring due to elevated serum viscosity. It can present with a triad of visual loss, neurological deficits and mucosal bleeding. Visual changes are due to microvascular changes due to thrombosis or haemorrhage.

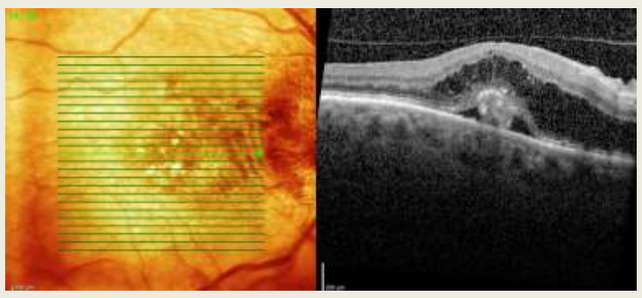
We present a rare ocular manifestation of HVS in an 85-year-old lady

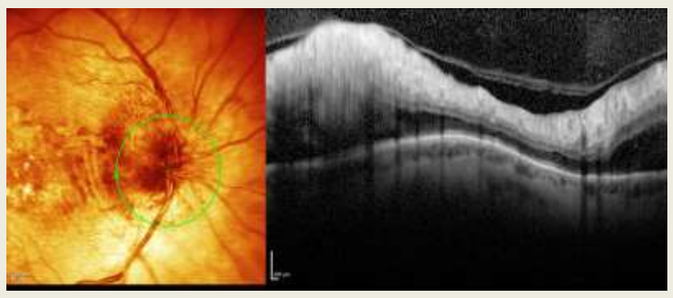
CASE REPORT

An 85-year-old lady presented to the eye emergency with right eye blurring of vision for the past 3 weeks. She had been seen by the optician a few days ago who noted right eye optic disc swelling.

O/E: Visual acuity OD LogMAR 1.1 and OS 0.1.

- *OD swollen optic nerve head with peri-papillary haemorrhages and macular hemorrhage, subretinal fluid extending to macula*
- Recent weight gain few months
- Occasional headaches
- Nil jaw claudication or polymyalgia rheumatica





POHx: RT Amblyopia since childhood (Age 11)

PMHx:

1. Under monitoring by Haematology for smouldering myeloma and CLL 2-3 years. (***Not required treatment but had been constantly under observation and seen around 6 weekly***)
2. Hypertension
3. Treated for right breast carcinoma with primary endocrine therapy 5 years ago (stable for over three years).
4. TIA May 2021

Management:

- Inflammatory markers: CRP WNL, ESR 128
- MRI brain with orbit
- Letter to Haematology and r/w 1/52

One week review

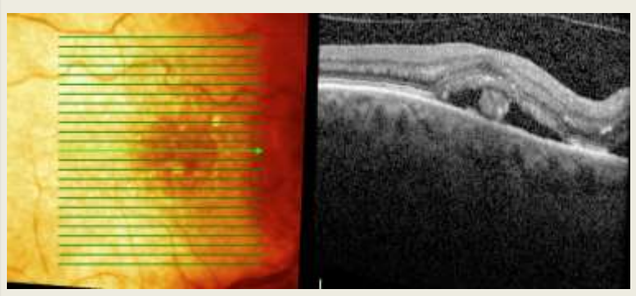
- Visual acuity stable
- Early cataracts, Nil NVI, Nil RAPD
- Fundus: *OD swollen optic disc with haemorrhages surrounding the optic disc and diffuse cystoid macular oedema present along with a small neurosensory detachment at macula*. CRT 424 microns. Nil NVE/NVD
- MRI awaited

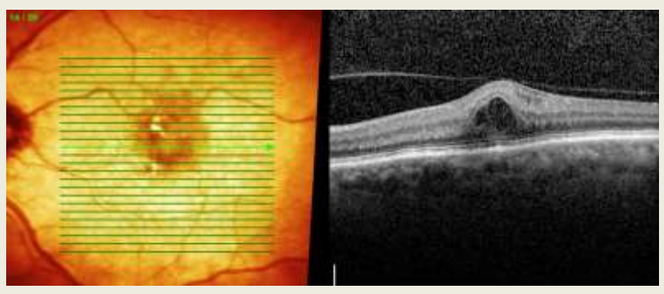
D/d:

1. CRVO with cystoid macular oedema.
2. ? Wet AMD
3. ? Hyper viscosity syndrome
4. ? AION

Management: Urgent referral to Medical Retina clinic

10-day review from presentation (Medical retina clinic):





- MRI and Haematology review awaited; Referral done again with suspicion of "infiltration/deposition of myeloma in the right eye
- OD suspicion of wet AMD, listed for right anti-VEGF
- OS suspicion of Vitelliform maculopathy and superior to the lesion

One week review:

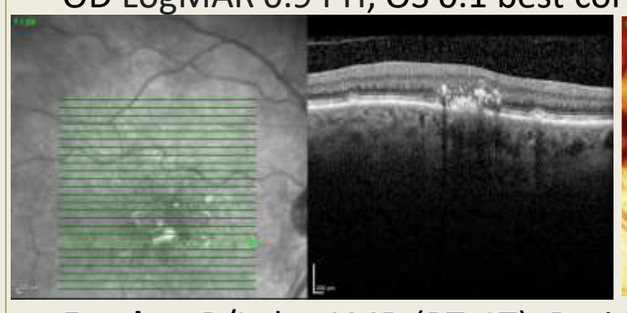
- Patient reported improved vision in both eyes
- OD LogMAR 1.0, OS 0.1 best corrected
- OD CRT 347 microns, *significant decrease in subretinal fluid*
- OS 297 microns, *no evidence of intraretinal fluid*
- **FFA:** *No evidence of subretinal neovascular membrane.*
- *Both fundi show significant improvement in vascular profusion.* Nil NVE
- *Removed from W/L for RT Lucentis.*
- Patient seeHaematology consult, CT, blood tests, awaiting further review.
- **MRI:** Moderate small vessel disease. No other cause of visual problem.

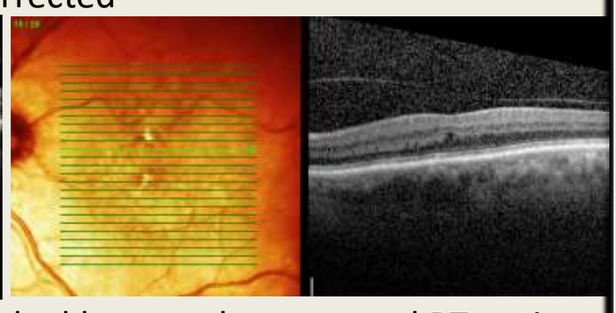
2 months review from presentation:

- OD LogMAR 0.9 PH, OS 0.1 best corrected
- *B/L improved vascular perfusion, near disappearance of vascular tortuosity. Few superficial retinal haemorrhages in the para-papillary area RT eye and absorbed superficial retinal haemorrhages LT eye*
- **OCT**: Nil intraretinal fluid both eyes.
- **Diagnosis:** Hyper-viscosity syndrome involving both eyes
- Treated for myeloma: medication, plasmapheresis

4 months review from presentation:

- Further vision improvement noted
- On Rx for HTN and Apixaban and statins
- OD LogMAR 0.9 PH, OS 0.1 best corrected





- **Fundus**: B/L dry AMD (RT>LT). Residual haemorrhage around RT optic disc.
- Further improvement in RT EYE unlikely from amblyopia and dry AMD.

DISCUSSION

HVS: Oncologic emergency that classically presents with the triad of:

Elevated blood viscosity due to either RBC shape deformity or pathological increase in serum proteins, RBC, WBC or platelets.

Cellular:		**Acellular (proteins):**
1. Polycythemia vera	CAUSES	*1. Myeloma (2nd MC)*
2. Leukemia		*2. Waldenstrom macroglobulinemia (M.C)*
3. Thrombocytosis		3. Cryoglobulinemia
4. Sickle cell disease		4. Seropositive rheumatoid arthritis
5. Spherocytosis		5. SLE
		6. Sjogren syndrome
**Myelomas are the second leading cause of HVS.*		7. Castleman disease
		8. HIV

- *Multiple myeloma (MM)* is a rare cancer with excessive production & improper function of plasma cells found in bone marrow.
- *Smouldering myeloma* is an early form of myeloma with abnormally high levels of atypical plasma cells in bone marrow without symptomatic disease. Half of those develop MM within 5 years.

DIAGNOSTIC CRITERIA:

Serum M protein (IgG or IgA) ≥3 g/dL or urinary M protein ≥500 mg/24 h and/or clonal BMPCs 10%-60% (Bone marrow Plasma Cell), and Absence of MDEs or amyloidosis

- Visual changes: Due to microvascular changes (thrombosis or haemorrhage). "***Sausage link" or "boxcar" engorgement of retinal veins, papilledema, flame-shaped hemorrhages, exudates***

Treatment:

Mainstays: Supportive therapy, judicious fluid administration , Plasmapheresis Phlebotomy

Definitive : Chemotherapy for underlying hematologic condition.

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