OCULAR MANIFESTATIONS OF SYSTEMIC DISEASES
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CASE #1

67 year old gentleman present with gradual decrease in vision in the right eye over 2-3 weeks. He denied GCA/PMR symptoms.

PMH
• DM Type 2 for 7 years
• HTN
EXAMINATION

- VISION: 20/50 OD, 20/20 OS
- IOP: 12 OD, 14 OS
- CVF: normal OU
- EOM: full OU
- Pupil: normal OU
- Lid/Adnexa: normal OU
- SLE: normal OU

OPTICAL COHERENCE TOMOGRAM RIGHT EYE
NO IV ACCESS FOR FLUORESCEIN ANGIOGRAM
NEXT STEP?
ADDITIONAL WORK-UP?

- Carotid ultrasound
- ESR/CRP

RESULT

- Normal ESR/CRP
- Carotid stenosis:
  - 99% right eye
  - 50-70% left side
OCULAR ISCHEMIC SYNDROME

• Common or internal carotid stenosis or occlusion → ocular hypoperfusion
• Seen in 5% with internal carotid stenosis/occlusion
  • > 90% stenosis → 50% reduction in central retinal artery perfusion pressure
  • 50% have complete occlusion
  • 20% bilateral
  • 70% present without a diagnosis of carotid stenosis
• Mean age: 65
• 2:1 Male: Female

CAUSES

• Atherosclerosis
• Giant cell arteritis
• Dissecting carotid artery aneurysm
• Fibrovascular dysplasia
• Takayasu arteritis
• Aortic arch syndrome
• Behcet’s disease
• Trauma or inflammation causing stenosis of the carotid arteries
• Intravitreal anti-VEGF
• Radiotherapy for nasopharyngeal carcinoma

CO-MORBIDITIES

• Hypertension 73%
• Diabetes mellitus in 56%
• Myocardial infarction 4%
• 40% Mortality rate within 5 years of onset secondary to:
  • Cardiovascular disease (66%)
  • Stroke
SYMPTOMS

- Visual loss (>90%)
  - 67% Gradual over a few weeks or months
  - 13% occurs over several days
  - 13% the loss is sudden over a period of minutes or seconds
  - 10% transient (minutes/seconds) vision loss
- Pain (40%)
  - Orbital pain
    - with normal IOP it may be caused by hypoxia of the eyeball and/or dura mater

ANTERIOR SEGMENT CHANGES

- Rubeosis iridis (66%)
- Neovascular glaucoma (50%)
- Spontaneous hyphema
- Uveitis (20%)
- Hypotony due to ciliary body ischemia
- Anterior and posterior synechia

POSTERIOR SEGMENT CHANGES

- Narrowed retinal arteries
  - Spontaneous retinal arteries pulsations
- Dilated retinal veins
- Midperipheral retinal hemorrhages (80%)
- Microaneurysms
- Retinal telangiectasia
- Cherry-red spot
- Cholesterol emboli

- Neovascularization (optic disc, retina)
  - Vitreous hemorrhage
  - Anterior and posterior ischemic optic neuropathy
  - Cotton-wool spots
  - Optic neuropathy
  - Glaucoma
    - MIG
    - NTS
DIFFERENTIAL DIAGNOSIS

- Giant Cell Arteritis
- Diabetic retinopathy
- Central retinal vein occlusion
- Hyperviscosity syndromes
- Autoimmune uveitis

WHEN SHOULD CAROTID DISEASE BE CONSIDERED?
WHEN SHOULD CAROTID DISEASE BE CONSIDERED?

- BRAO/CRAO
- Uveitis
- Rubeosis
- Diabetic retinopathy asymmetry

CASE #2:

32 year old gentleman presents to his optometrist for his yearly diabetic examination with no visual complaints.

PMH:
- DM Type 1 x 17 years, Hgb A1c 9.3
- No HTN

EXAMINATION

- VA: 20/20 OU
- IOP: 13 OD, 12 OS
- CVF: normal OU
- EDM: full OU
- Pupil: normal OU
- L/Alex: normal OU
- S/E: normal OU
ROTH SPOTS

- White-centered retinal haemorrhages
- Reflect a relatively acute systemic change, therefore rare in chronic conditions (DM, HTN)
- Acute systemic insults to homoeostasis → capillary rupture → platelet-fibrin thrombus formation
- White center could represent
  - platelet-fibrin thrombus
  - Inflammatory infiltrates
  - Infectious organisms
  - Accumulation of neoplastic cells
ROTH SPOTS SEEN IN THE FOLLOWING:

• Anemia
• Leukemia
• Subacute bacterial endocarditis
• Diabetic retinopathy
• Hypertensive retinopathy
• Anoxia
• Carbon monoxide poisoning

WORK UP?

CBC

• WBC: 328K
• Hgb 9
• Platelet 98
CHRONIC MYELOGENOUS LEUKEMIA (CML)

Blast cells replace the normal elements of the bone marrow

Complications:
• 2° Disease:
  • Bone marrow failure → anemia, thrombocytopenia and neutropenia
  • Infiltration of liver, spleen, lymph nodes or, less frequently, CNS
• 2° Treatment:
  • Chemotherapy +/- radiotherapy toxicity
  • Bone marrow transplantation → graft versus host disease
  • Immunosuppression → infections

OCULAR INVOLVEMENT

• With improvement of the survival ↑ the incidence of ocular manifestations (in up to 90%)
• Ocular involvement much more likely in acute than chronic leukemia
• Ocular involvement is associated with a higher frequency of bone marrow relapses and CNS compromise weeks or months later, which means a poor prognosis and a low survival rate.
• Leukemic relapses are often diagnosed after ocular presentation.

OCULAR INVOLVEMENT

• All the structures of the eye and its adnexal structures can be affected
• Ophthalmic involvement can be classified into two major categories:
  • (1) primary or direct leukemic infiltration
    • show three patterns
      • anterior segment uveal infiltration
      • orbital infiltration
      • neuro-ophthalmic signs of CNS leukemia that include optic nerve and cranial nerve involvement
  • (2) secondary or indirect involvement:
    • Due to anemia, thrombocytopenia, hyperviscosity, and immunosuppression
FUNDUS FINDINGS
- Venous dilatation and tortuosity
- Retinal hemorrhages
- Roth's spots
- Cotton wool spots
- Exudates
- Perivascular sheathing
- Neovascularization
- Vascular occlusion
- Microaneurysms

CHOROIDAL INVOLVEMENT
- The most frequently involved ocular tissue
- Often not clinically apparent
- There can be a diffuse or perivascular involvement

VITREOUS INVOLVEMENT
1. Internal limiting membrane generally acts as an effective barrier to leukemic cell infiltration
2. Leukemic cells occasionally invade the vitreous body, possibly emerging from the optic nerve head
OPTIC NERVE

- Optic nerve is usually involved in CNS leukemia (13 to 18% of leukemias)
- CNS involvement is becoming more frequent with improvement in survival
- Symptoms of CNS leukemia depend
  - ↑ ICP
  - Cranial nerve involvement
- Disc swelling is the most frequent sign of optic nerve involvement. Due to
  - Direct infiltration of the optic nerve
  - ↑ ICP
- Always means a poor prognosis, esp. if it occurs during treatment, rather than after it.
- Optic nerve is relatively unaffected by systemic chemotherapy and serves as a sanctuary of acute lymphoblastic leukemia. It usually includes intrathecal chemotherapy and radiotherapy.

MOST COMMON QUESTION IN MY PRACTICE?

“HOW DO YOU SAY YOUR NAME?”
Thuc Anh Thi Multerer
ThucAnh (“Taupe Un’)
Thi vs Ti
Multerer