Disseminated T Cell Lymphoma

Presenting With

Autoimmune Retinopathy, Unilateral Chorio-retinal Infiltration And Positive Anti-retinal Antibodies

Lidia Alvarez
Sarah Coupland
Neil Parry
Alec Ansons
Aim

To report an unusual case where intraocular lymphoma (IOL) and autoimmune retinopathy (AIR) coincided in the same patient to cause bilateral visual loss.

Discuss the diagnostic challenges of these two entities, in the immuno-compromised with strong autoimmune background.
Woman, 62 years old
Autoimmune hepatitis and cirrhosis
**Immunosuppressed** for years on Azathioprine

Attended the emergency services with
*left retroocular pain and general malaise;*
normal examination and VA 6/7.5 BE

FBC found **leucopenia**
-> Colony growth stimulating factors
Azathioprine switched to steroids
-> symptoms improved
3 months later ...

From initial visual acuity (VA)
6/7.5 both eyes

Re-attended with acute visual loss in the right eye to counting fingers (CF)

Vascular attenuation
Optic pallor
Central Retinal Artery Occlusion?
5 days later...

Re-attended with drop of VA in the left eye to 6/24

Macular oedema after Branch retinal artery occlusion?
Vasculitic/ embolic artery occlusions?

**Admitted for IV high dose steroids**

- Bloods inflammatory + vasculitis markers
  - CRP, ESR, ANCA, ANA
  - Thrombophilia
- Infectious screening (HIV, syphilis, CMV viral load, HSV, quantiferon)
- Temporal artery biopsy
- MRI scan brain and orbits
- Eco-doppler carotid vessels
- Chest X-ray
5 days after admission...

LE VA drops to counting fingers

Fundoscopy
No macular oedema, no optic disc swelling

Sluggish pupils

Anterior chamber:
Mild very fine keratic precipitates and odd cell in both eyes.
Non-specific findings....

- Cardiolipin raised
- c-ANCA positive
- Left pansinusitis biopsied -> inflammatory infiltrate
- Non-specific small lung inflammatory infiltrate
Electrodiagnostics

Wide-spread retinopathy

R  L

Light-adapted

3.0 30Hz

Dark-adapted

3.0

10

Photoreceptors affected

Paraneoplastic retinopathy???

Whole body CT scan

Only same lung “inflammatory” infiltrate: non-specific
Systemic deterioration
Cough, Skin rash
Chest X ray

Increased infiltrate
Sputum culture + *strep pneumonia*

Wide spectrum antibiotics
and *as steroids decreased further*...

Retinal biopsy confirmed *T cell lymphoma*
Cause of loss of vision in the RE? Cause of rod dysfunction in BE?

ERG

Antirretinal antibodies

*Positive (Enolase)*

Autopsy

*Disseminated T cell lymphoma (both lungs)*
# Intraocular lymphomas

<table>
<thead>
<tr>
<th>Intraocular lymphomas</th>
<th>Most common clinical features</th>
<th>Origin</th>
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<tbody>
<tr>
<td><strong>Primary intraocular high grade B-cell lymphoma</strong></td>
<td><strong>Masquerade syndrome posterior uveitis:</strong> Vitreitis, Floaters</td>
<td>Arise in neural tissue: Retina, brain and spine.</td>
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<td></td>
<td>Bilateral 60-90%</td>
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<tr>
<td><strong>Choroidal B-cell lymphoma</strong></td>
<td><strong>Diffuse choroidal mass posterior pole/ optic nerve</strong> Metamorfopsia/ loss of VA</td>
<td>Metastasis to the uveal tissue of B lymphomas from solid viscus</td>
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<tr>
<td>Primary (low grade) or metastatic</td>
<td>Unilateral &gt;Bilateral</td>
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<tr>
<td><strong>Intraocular T-cell lymphoma</strong></td>
<td><strong>Masquerade syndrome:</strong> Vitreitis, floaters, retinal infiltrates, anterior uveitis</td>
<td>Systemic disseminated T-cell lymphoma (mycosis fungoides)</td>
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<tr>
<td>Metastatic</td>
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Autoimmune retinopathy

1. Cancer-associated retinopathy (CAR)
2. Melanoma-associated retinopathy (MAR)
3. Presumed non-paraneoplastic autoimmune retinopathy (npAIR)

Break of retinal immune privilege?

Very rare
Strong history of autoimmune disease

Rapidly progressive, bilateral, painless visual deterioration asymmetric/sequential.

Often no obvious signs at presentation
No evidence-based treatment strategy: Immunosuppression.

Small-cell lung carcinoma, gynaecological tumors
Non-small cell lung cancer, lymphoma, prostate, colon

Antiretinal antibodies

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**Paraneoplastic retinopathy**

**CAR**
- Ab targeting outer retina: photoreceptors → **absent a and b waves**
- Loss of central and peripheral vision

**MAR**
- Ab targeting inner retina: bipolar cells → **absent b wave** (electronegative ERG)
- Photopsias and loss of peripheral vision
RE improvement with steroids....

First ERG
RE VA of CF

Second ERG
VA improved RE 6/60
Conclusions

- Unique case: heavy immunosuppression and autoimmune background → confusing factors in the diagnosis.
  - Open possibilities; vasculitis/ infection?
  - Suppression of lymphoma-related inflammation → late diagnosis.

- Extremely rare concurrence of simultaneous conditions, however:
  - Autoimmune background is predisposing for AIR and immunosuppression is for lymphoma (tumors).
  - When immunosuppression was decreased, immune recovery may have precipitated AIR, and lymphoma-related inflammation manifested with > tumor size.

- Intraocular lymphoma = bad prognosis.
  Histology important:
  - B lymphomas are mostly primary, life prognosis depends on concurrent CNS involvement.
  - T-lymphomas usually occur in systemic disease.

- High index of suspicion in bilateral or sequential loss of vision in a paucity of signs for AIR. Electrodiagnostics!
Thank you!