The dilemma of Multiple sclerosis

By
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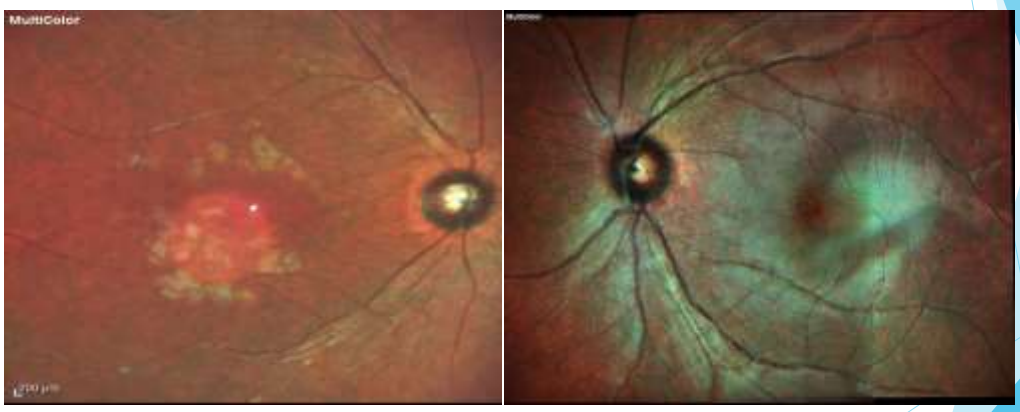
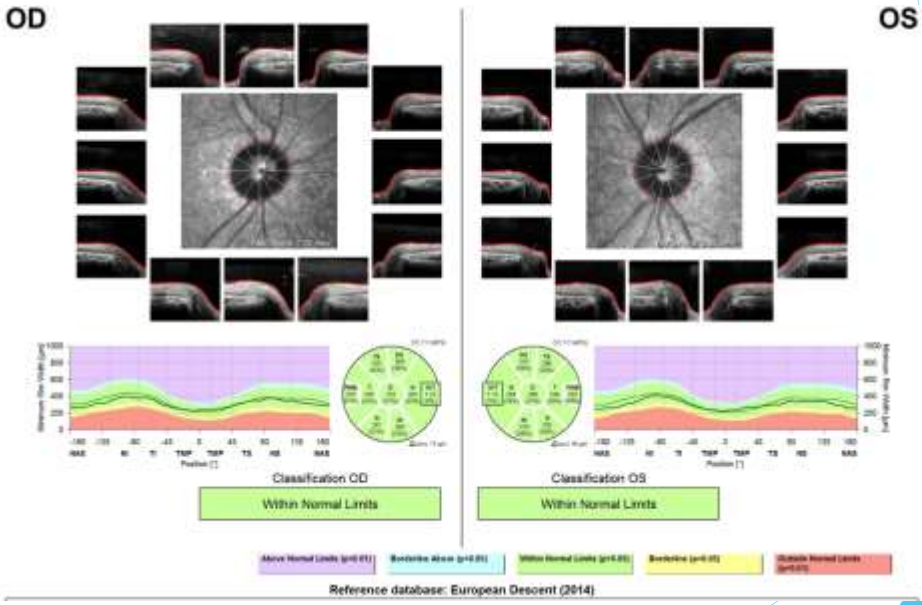
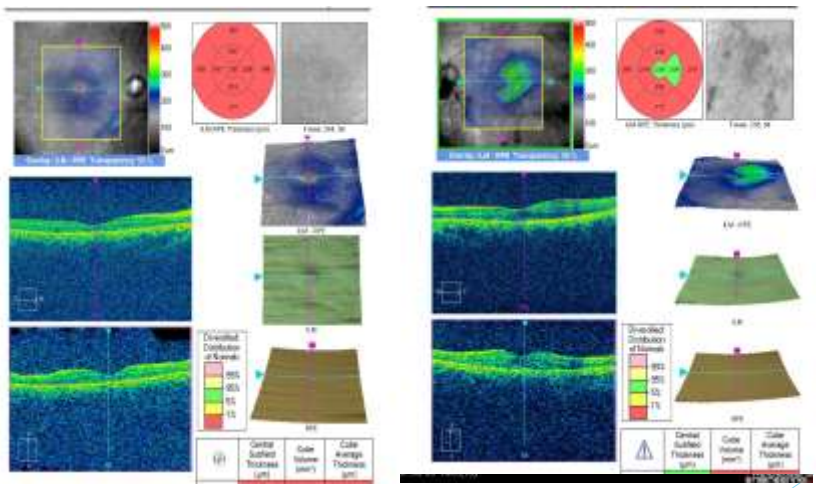
- A 20 -year old female patient gave a history that started 6 months ago of Rt diminution of vision.
- She seaked the advise of an Ophthalmologist who after thorough examination suspected M.S. and referred her to a neurologist who did her MRI brain and CSF tap.
- MRI showed no demyelination but the CSF showed positive oligoclonal bands
- she was given pulsed steroid therapy followed by a maintenance dose.

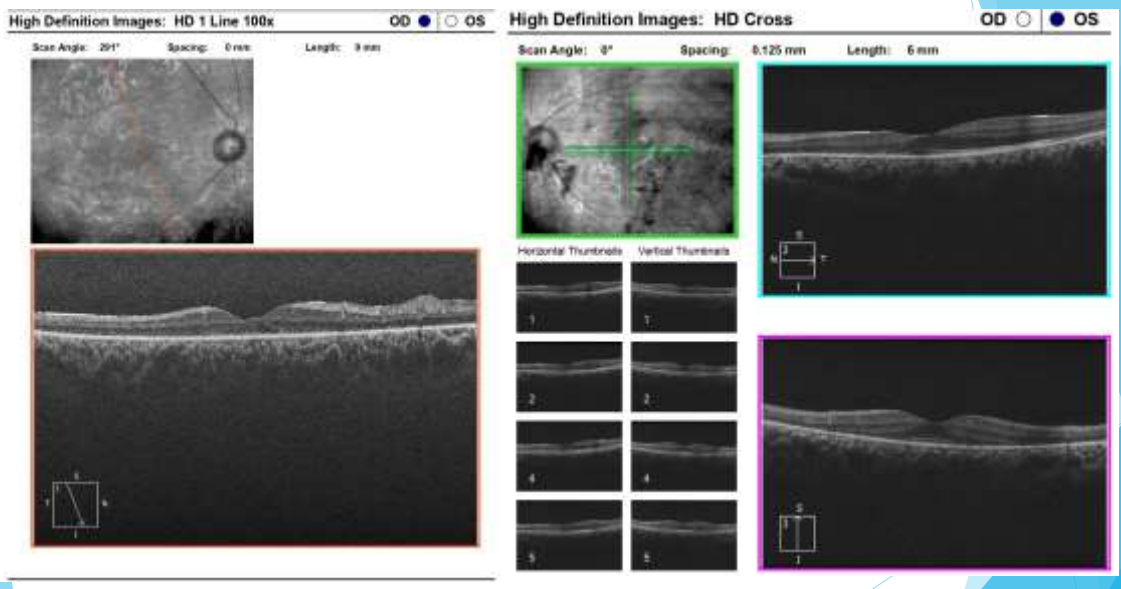
- She was referred by a second neurologist to our clinic for confirmation of M.S. diagnosis.
- Full history taking including neurological, bone, skin and chest were negative.
- on examination:

The visual acuity was 0.1 OD and 1.0 OS with no other anterior segment positive signs.




OD
OS
Classification OD
Within Normal Limits
Classification OS
Within Normal Limits
Reference database: European Descent (2014)



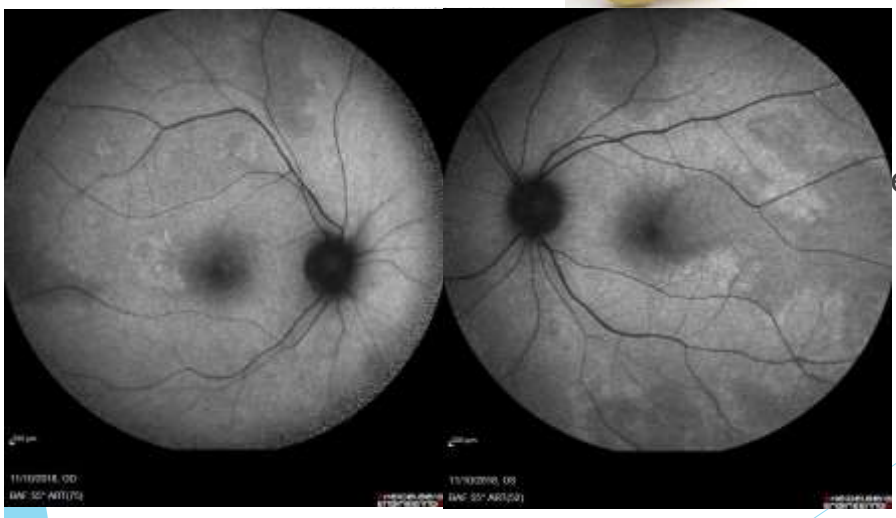


- since the main brunt of the disease was affecting the outer retina, we decided that she is a case of bilateral non-infectious chorioretinopathy more advanced on the right side.
- We knew that the picture of the disease was modified by the high dose of steroids she was receiving (60 mg /day)
- More detailed history was taken with no any systemic positive signs .

- To minimize the dose of steroids, Imuran(azathioprine) was prescribed with tapering of the dose of steroids to 40 mg prednisolone /day

- We took a second opinion and she was diagnosed as VKH and was referred to an immunologist who asked for investigations which were negative for rheumatoid, VDRL, B51 and B27 with a positive pathergy test.
- The immunologist accepted the VKH diagnosis and gave her HUMIRA (adalimumab) injection every 2 weeks
- He also stopped the Imuran(azathioprine) and tapered the steroid dose to 20 mg/D.

still not convinced



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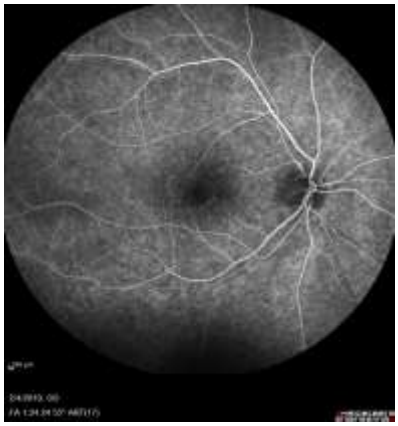
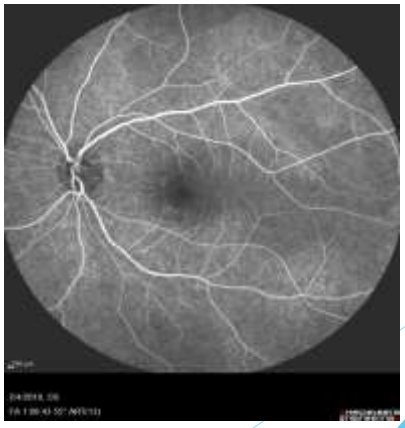
Still not convinced

- The patient reported that she had a genital nodule but no ulcers ???

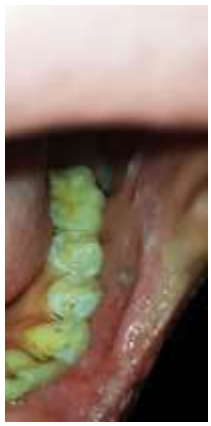
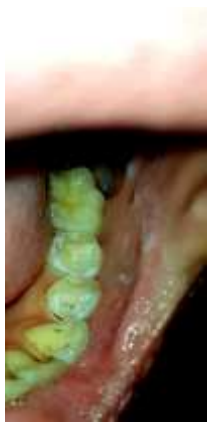
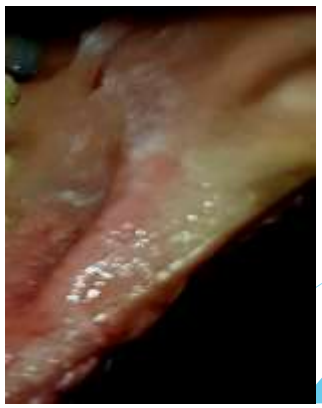




Still confused

Still confused till last week VA 0.4 eccentric fix and 1

- Recurrent mouth ulcer

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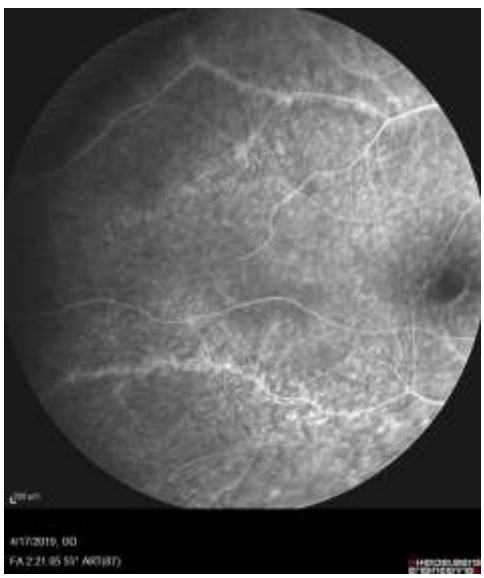



Select Your Final Diagnosis:

- MS
- VKH
- AZOOR
- Behcet's

Why Behcet's?

- Vasculitis on FA
- Recurrent ulcers
- Positive pathergy test (not pathognomonic)
- Steroids may have masked the skin lesions
- Steroids may have changed the whole picture

Why not behcet

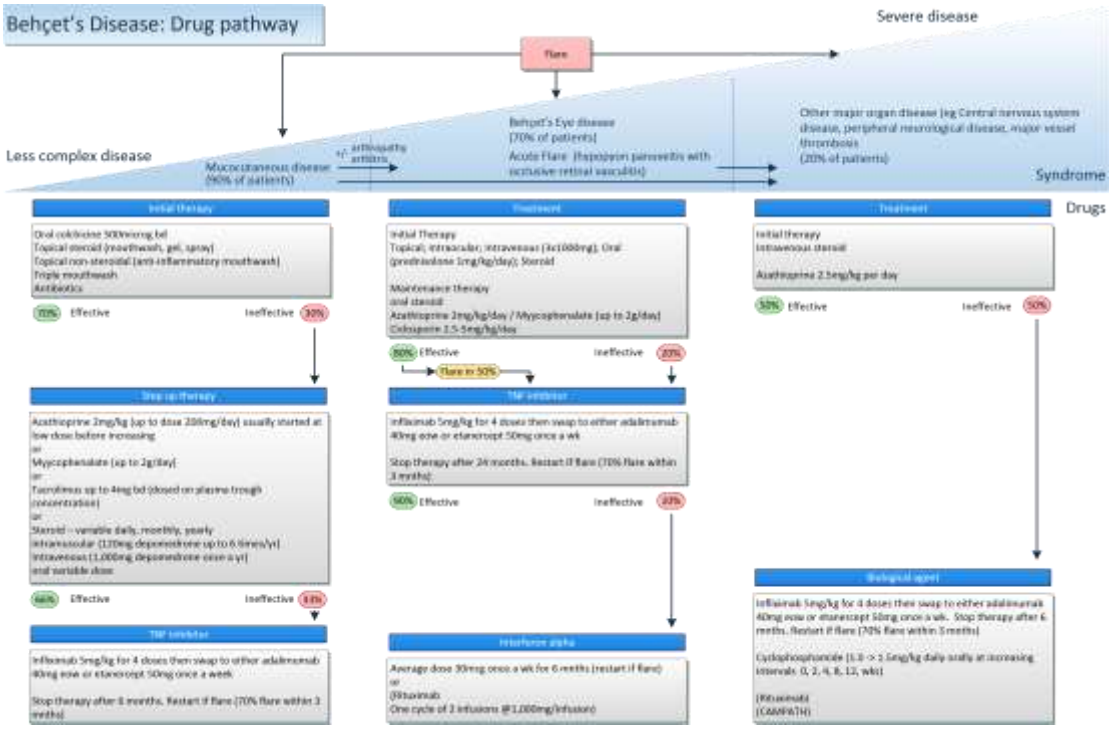
- Oligoclonal bands on CSF which is not a characteristic of Behcet's
- FAF picture.
- Early FA pictures which didn't fit with frank Behcet's.
- OCT Affection of the outer retinal layers.

Why relapse now ??

- Steroids at the start may have altered the picture and then after the immunologist stopped the Imuran (azathioprine) and tapered the steroids, reactivation occurred and vasculitis appeared

Take Home Messages

- Behcet's disease could be masqueraded in many forms.
- A team work, which may include a neurologist and an immunologist is needed.
- MS should be included in the DD but not always the diagnosis in this age group.
- Repeated FA, as well as FAF, may have a role in proper diagnosis.
- Do not rush for steroid therapy if pathology is uncertain.
- Continued FUP may be required to look for relapses.


Behçet's Disease: Drug pathway
Severe disease
Flare
Less complex disease
+/- arthropathy arthritis
Mucocutaneous disease (90% of patients)
Behçet's Eye disease (70% of patients)
Acute Flare (Papillitis with occlusive retinal vasculitis)
Other major organ disease (eg Central nervous system disease, peripheral neurological disease, major vessel thrombosis (20% of patients)
Syndrome
Drugs
Initial therapy
Oral colchicine 500microg bd
Topical steroid (mouthwash, gel, spray)
Topical non-steroidal (anti-inflammatory mouthwash)
Triple mouthwash
Antibiotics
70% Effective
Ineffective 30%
Step up therapy
Azathioprine 2mg/kg (up to dose 200mg/day) usually started at low dose before increasing
or
Mycophenolate (up to 2g/day)
or
Tacrolimus up to 4mg bd (dosed on plasma trough concentration)
or
Steroid – variable daily, monthly, yearly
intravitreous (120mg dexamethrone up to 6 times/yr)
intravenous (1,000mg dexamethrone once a yr)
oral variable dose
66% Effective
Ineffective 33%
TNF inhibitor
Infliximab 5mg/kg for 4 doses then swap to either adalimumab 40mg eow or etanercept 50mg once a week
Stop therapy after 6 months. Restart if flare (70% flare within 3 months)
Treatment
Initial Therapy
Topical, intraocular, intravenous (4x1000mg), Oral (prednisolone 1mg/kg/day); Steroid
Maintenance therapy
oral steroid
Azathioprine 2mg/kg/day / Mycophenolate (up to 2g/day)
Ciclosporin 2.5-5mg/kg/day
80% Effective
Ineffective 20%
Flare in 50%
TNF inhibitor
Infliximab 5mg/kg for 4 doses then swap to either adalimumab 40mg eow or etanercept 50mg once a wk
Stop therapy after 24 months. Restart if flare (70% flare within 3 months)
50% Effective
Ineffective 50%
Interferon alpha
Average dose 30mcg once a wk for 6 months (restart if flare)
or
(Rituximab
One cycle of 2 infusions @1,000mg/infusion)
Treatment
Initial therapy
Intravenous steroid
Azathioprine 2.5mg/kg per day
50% Effective
Ineffective 50%
Biological agent
Infliximab 5mg/kg for 4 doses then swap to either adalimumab 40mg eow or etanercept 50mg once a wk. Stop therapy after 6 months. Restart if flare (70% flare within 3 months)
Cyclophosphomide (1.0 -> 1.5mg/kg daily orally at increasing intervals. 0, 2, 4, 8, 12, wks)
(Rituximab
(CAMPATH)