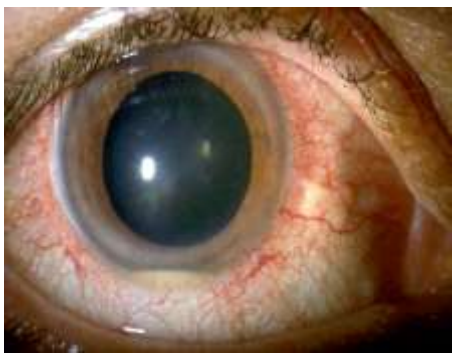
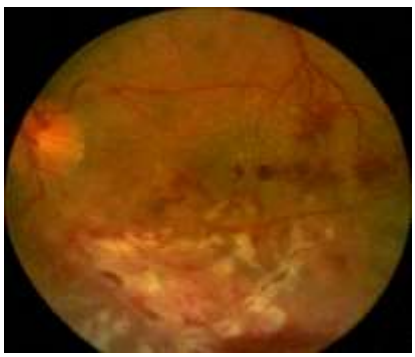
NEURO BEHCET

Mohamed Abdel Azim FRCOphth

Visual impairment in Behcet's disease is not always caused by uveitis





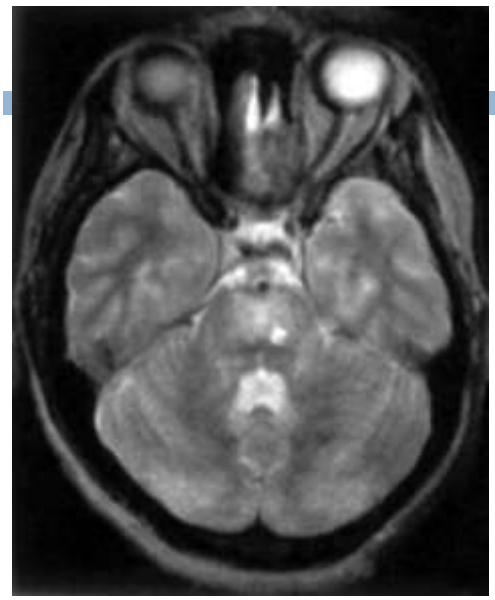
Neurologic manifestations of Behcet's disease

Central nervous system :

(1) parenchymal involvement, which includes brainstem involvement, hemispheric manifestations, spinal cord lesions, and meningoencephalitic presentations

(2) nonparenchymal involvement, including dural sinus thrombosis, arterial occlusion, and/or aneurysms.

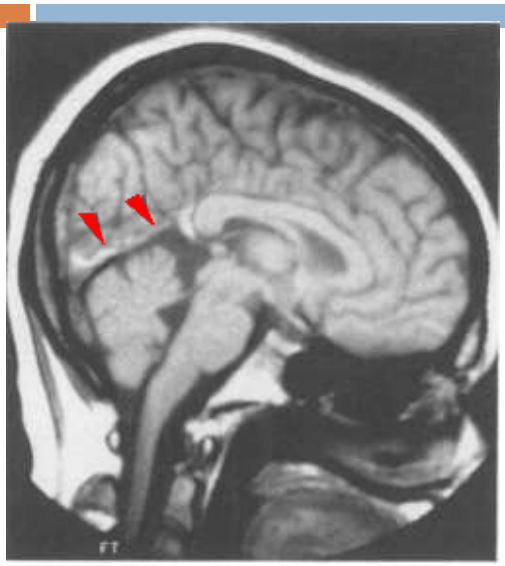
Peripheral neuropathy and myopathy are relatively rare.



Cerebral vasculitis

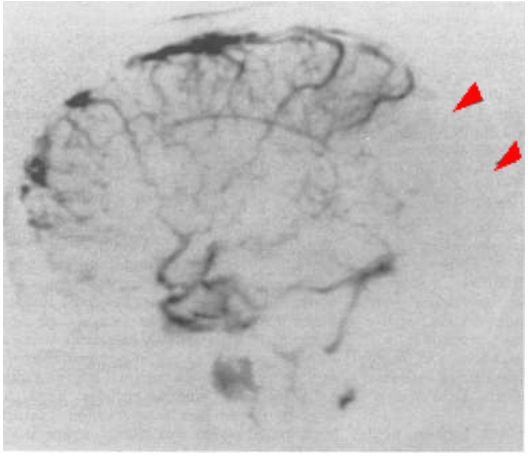
Example: Foci of hyperintensity in the pons and midbrain bilaterally

Venous sinus thrombosis

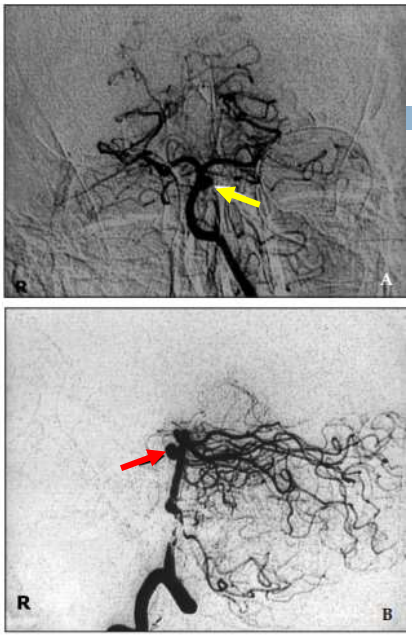


Example: irregular filling defects in the straight sinus

Venous sinus thrombosis



Example: Cerebral angiography confirms non-filling of the posterior half of the sagittal sinus



Arterial complications

Stenosis, occlusions and aneurysms (most serious).

(very rare-exceptional)

Cerebral vasculitis affecting the visual centers / visual pathway.

History

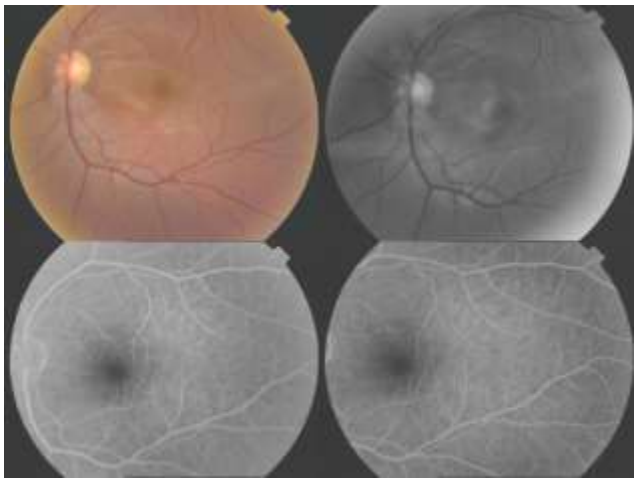
- **PC:** 25 years old engineer presented to my clinic with a 2 years history of poor vision more in the right eye.

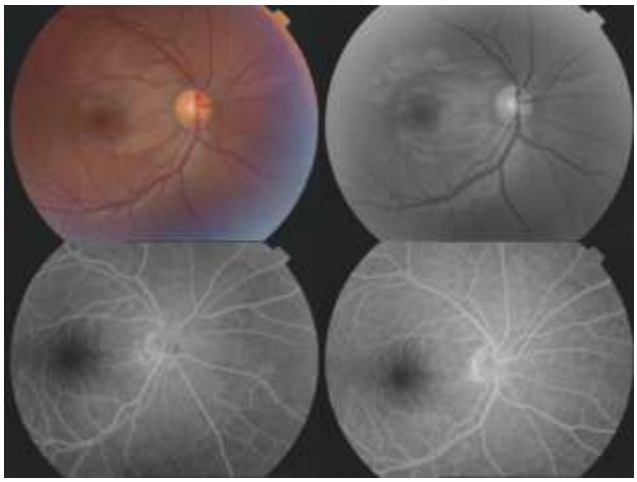
History

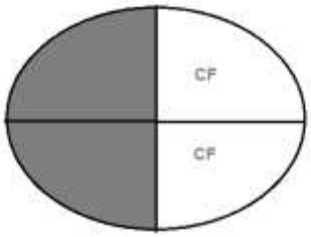
- He also complained of constant headache for which he had several brain MRIs revealing brain vasculitis.
- Erroneous diagnosis of uveitis as the cause of his poor vision.
- He received several doses of methyl prednisolone pulse therapy and several doses of peri-ocular steroids with limited improvement.
- Methyl prednisolone pulse therapy X 3 was repeated in addition to oral cyclophosphamide

Ophthalmic examination

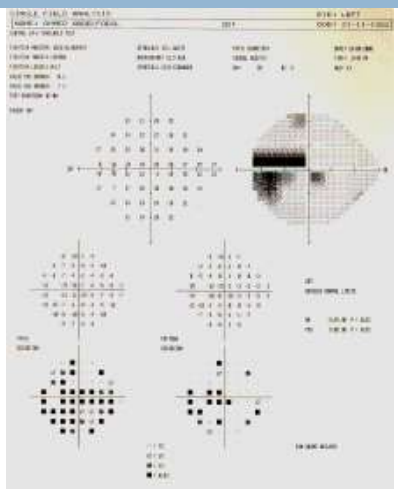
RVA: CF 1 meter with central scotoma		LVA: 6/24-
Q&D	AC	Q&D
RRR	pupils	RRR
NAD pale disc?	Fundus	NAD

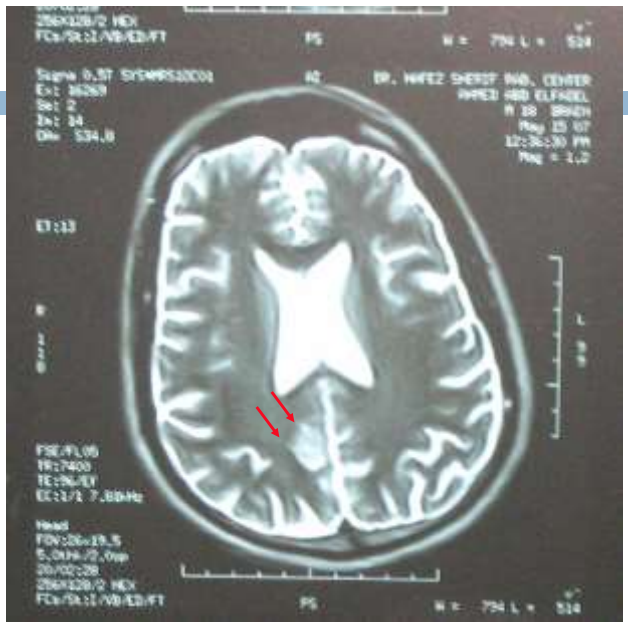


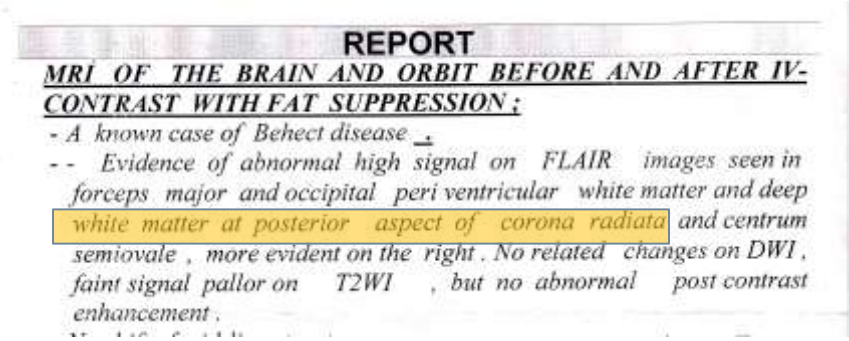




RVA too poor to allow VF testing however it could be obtained by confrotation







REPORT

MRI OF THE BRAIN AND ORBIT BEFORE AND AFTER IV-CONTRAST WITH FAT SUPPRESSION :

- A known case of Behect disease .

- - Evidence of abnormal high signal on FLAIR images seen in forceps major and occipital peri ventricular white matter and deep white matter at posterior aspect of corona radiata and centrum semiovale , more evident on the right . No related changes on DWI , faint signal pallor on T2WI , but no abnormal post contrast enhancement .

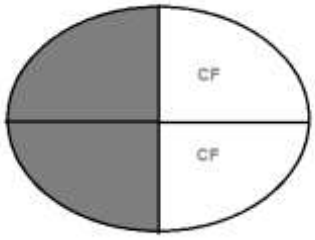
Treatment with Ramicade (Infleximab)

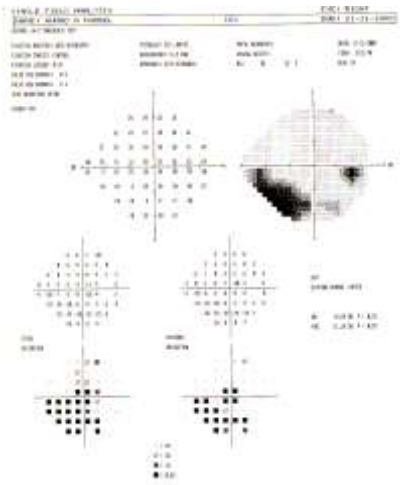
- Since there hasn't been any appreciable improvement on maximum immunosuppressive therapy (steroids + cyclophosphamide).

Post-treatment

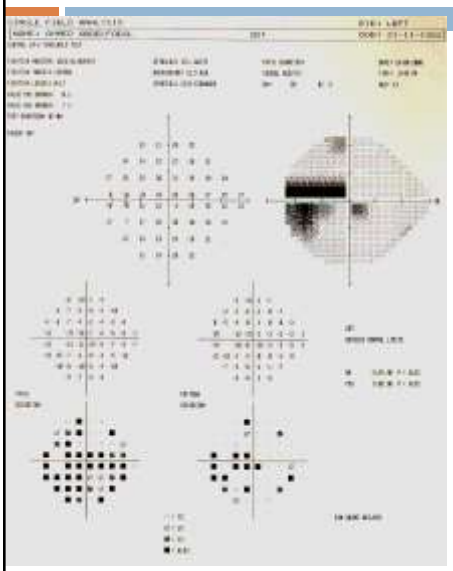
- Almost immediately the patient noticed marked improvement of his symptoms.
- I examined him one month following his initiation doses and found his RVA 6/24 and LVA of 6/18+.
- FA was normal (unchanged).
- His VF revealed great improvement.

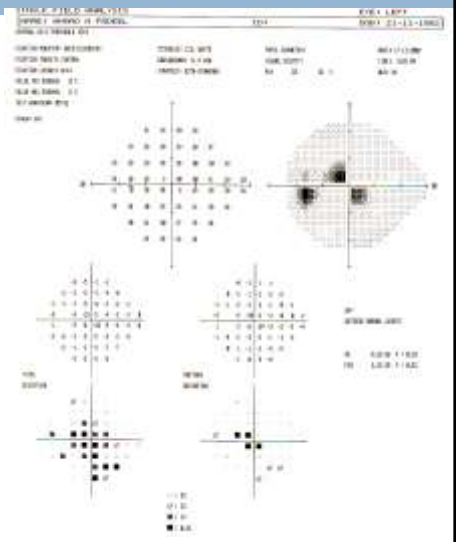
Post treatment R & L VF





Pre- vs. Post- treatment LVF





- Completely rehabilitated and back to his job as an engineer.
- He is maintained on a low dose methotrexate (10 mg / week) to reduce the possibility of the production of anti-(anti-TNF) antibodies.
- This patient is currently controlled on 2-3 monthly Ramicade and low dose prednisolone since more than 9 years.

Present

Venous sinus thrombosis causing intracranial hypertension

History

Date: *(12/1/09)* **22 year old male student**

PC:

- **2 months history of headache and diplopia of acute onset.**

POH:

- **Bilateral Hypermetropia with mild amblyopia.**
- **Bilateral Papilledema for which he had several LP (++ opening pressure but of normal cytology)**

PMH:

- **Behcet's disease (recurrent mouth and genital ulcers, superficial thromophlebitis).**

DH:

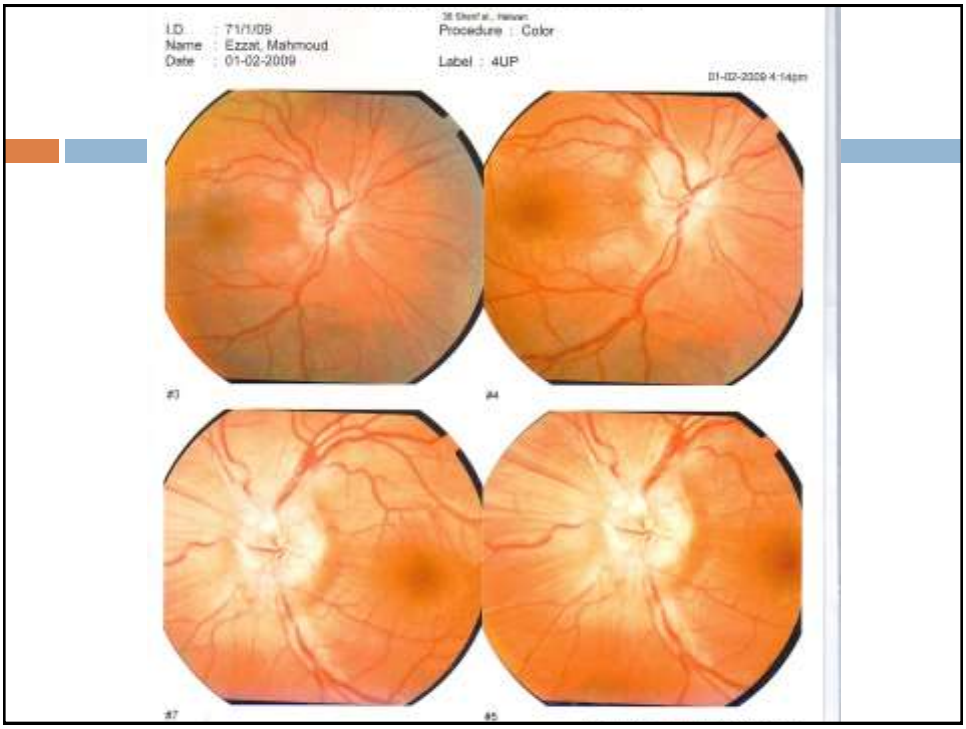
- **Several courses of systemic steroids.**

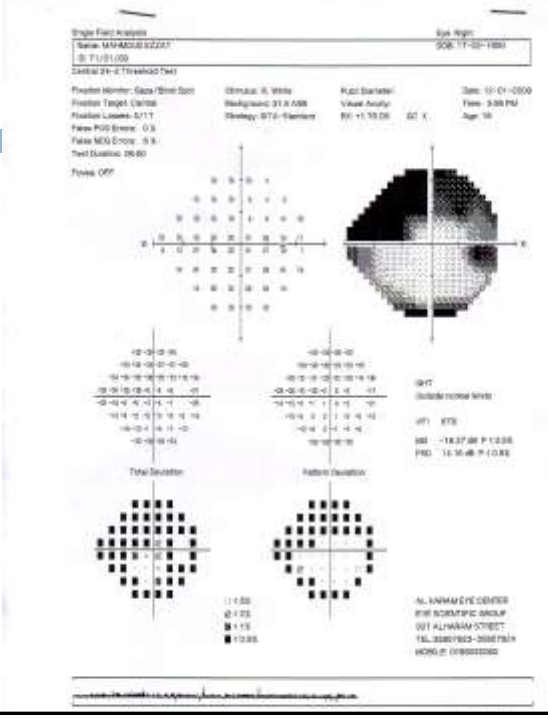
Examination

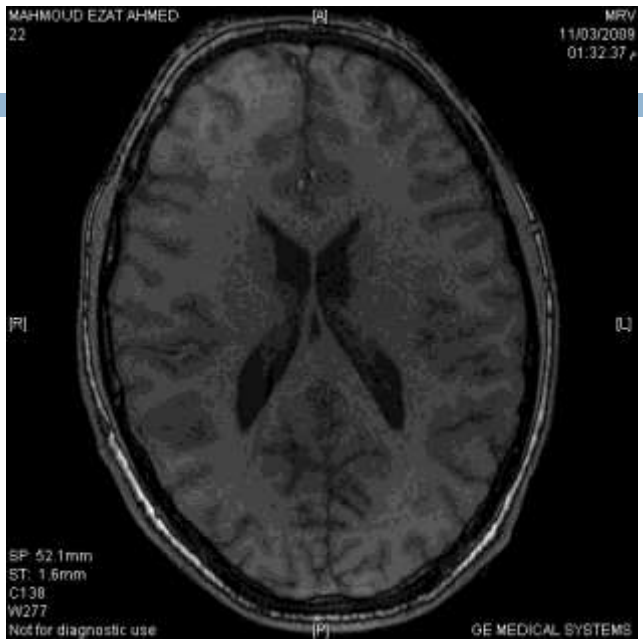
- Appearance:

Examination

- RVA: 6/18 LVA 6/12
- Normal Anterior segment bilaterally
- Bilateral established papilledema.
- R incomittent esotropia due to R 6 th N palsy.





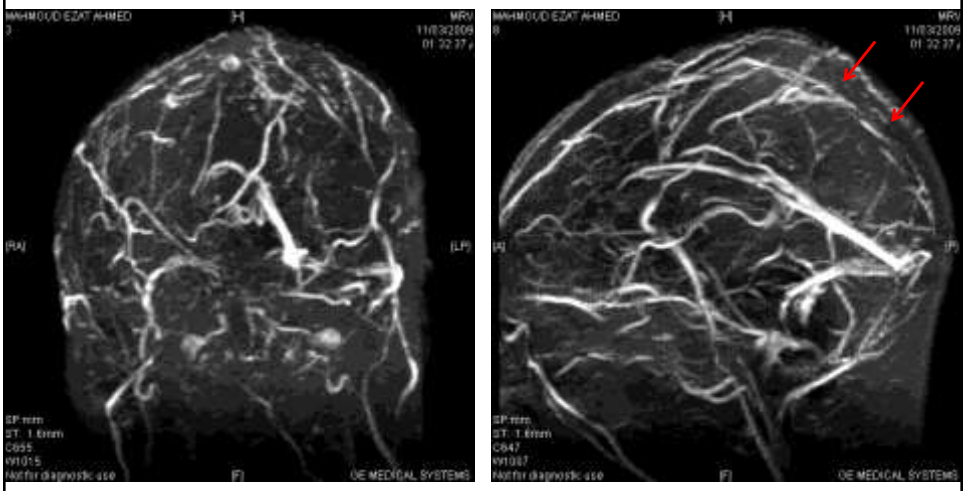

MAHMOUD EZAT AHMED
22
[A]
MRV
11/03/2009
01:32:37
[R]
[L]
SP: 52.1mm
ST: 1.6mm
C138
W277
Not for diagnostic use
[P]
GE MEDICAL SYSTEMS

Management

- Relation between papilledema and Behcet's disease, Coincidental ?
- Treated with diuretics, repeated LP and steroids however with little response.
- Bilateral ON sheath decompression was scheduled to relief the unresponsive ICP.

Managment

- MRV in patients diagnosed with IIH revealed more 10% were have cerebral venous sinus thrombosis (CVST)in presence of normal MRI. [*Occurrence of Cerebral Venous Sinus Thrombosis in Patients with Presumed Idiopathic Intracranial Hypertension Ophthalmology (December 2006)*]
- Finding venous sinus thrombosis ;a potentially fatal condition; means the patient needs anti-coagulants , systemic immunosuppression and closer monitoring.

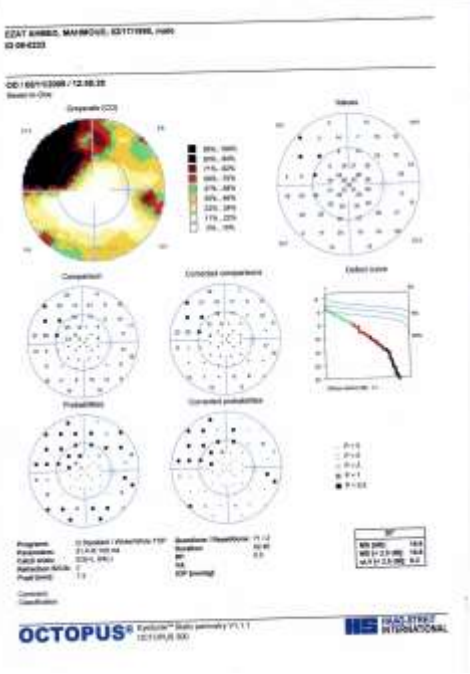


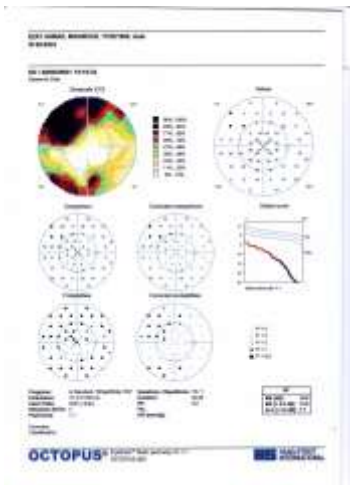
Management

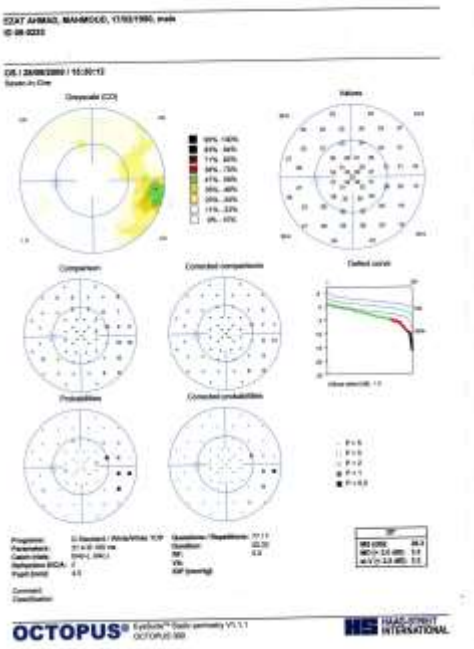
- The patient was started on anti-coagulants aiming at an INR >2<3.
- Started on Infliximab (Remacade), Azathioprin (Immurane) and prednisolone since the venous occlusion is probably secondary to Behcet's related vasculitis.
- In addition; he continued on Lasix to control the raised ICP.

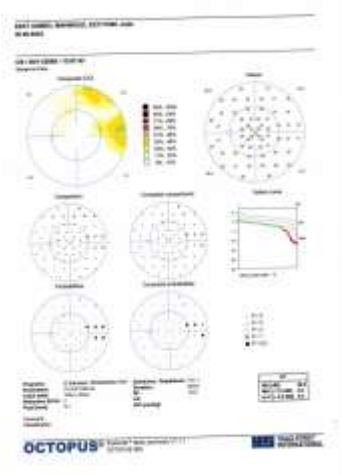
Managment

- Only few weeks later his headache started to resolve.
- Fundus exam revealed complete resolution of the papilledema.
- VF testing revealed no further deterioration over the next 4 months.
- MRV was repeated reporting resolution of the venous thrombosis.









Present

- There has been a remarkable improvement of his visual acuity to 6/9+ bilaterally.
- The reason for this could probably be explained by the resolution of an associated cerebral edema. *[**Cerebral venous thrombosis presenting as acute visual loss** BJO (October 26, 2009)].*
- There was a complete resolution of the 6 th nerve palsy.
- Up-today he is maintaining convalescence and is completely rehabilitated only on 3 monthly Ramicade and Immurane 50mg/d.

History

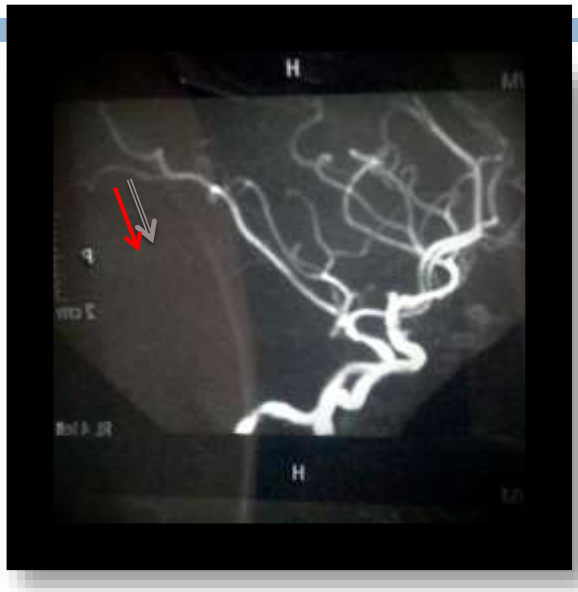
- 29 years old male Behcet's disease
- Presented 9/2014 blurred vision L>R severe headache
- LP done +++ opening pressure no cells or growth
- MRI and MRA –free
- MRV—occluded extra-cranial dural sinus
- Tuberculin, HCV, HIV –ve

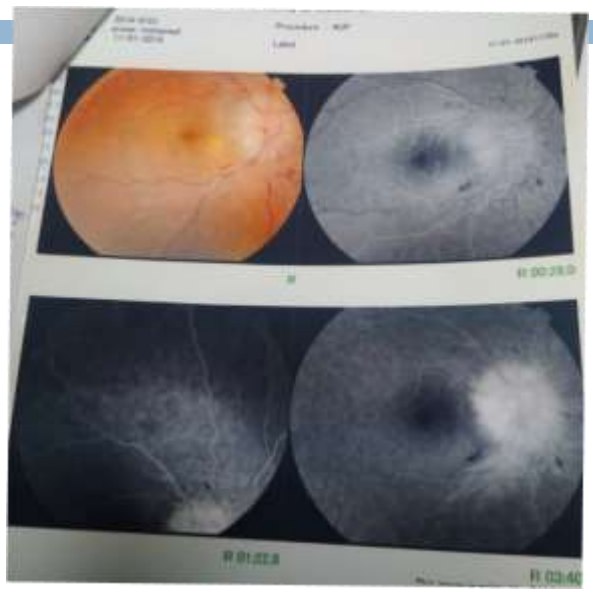
Examination

- RVA 6/9 LVA NPL
- Fundus examination :

-Bilateral markedly elevated discs with hges.

-Left optic disc was "choked" and extremely elevated with totally empty retinal vessels, retinal hemorrhage and associated vitritis





Managment

- FA 11/2014 Right disc edema with splinter hges delayed venous filling
- VEP markedly delayed in L eye
- TT: Clexan 60mg X2, pred 60mg, Marevan 6mg

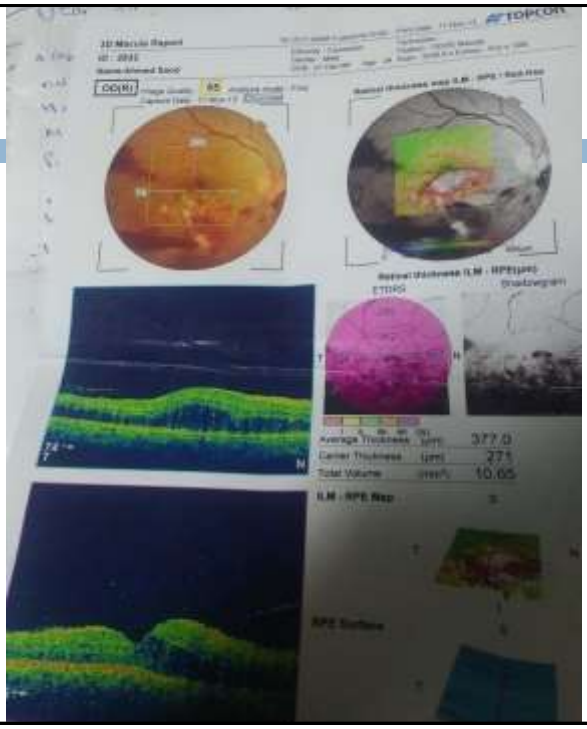
Endoxane (cyclophosphamide) 850mg and Cidamex 250mg X4

- Hemorrhages partially resolved leaving an established papilledema.
- We had to repeat the lumbar puncture twice to reduce the IC pressure for fear of optic atrophy.
- He was then started on cyclopsporin A 200 mg instead of endoxan (which I think is not very effective) gradually stopping the anticoagulants.

Present

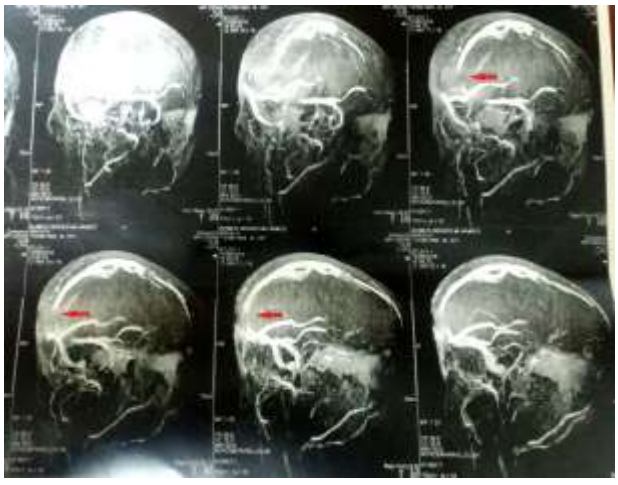
- Last visit 8/2017 papilledema has completely resolved maintaining 6/6 vision.
- curently he is on CSA 200mg and pred 10mg

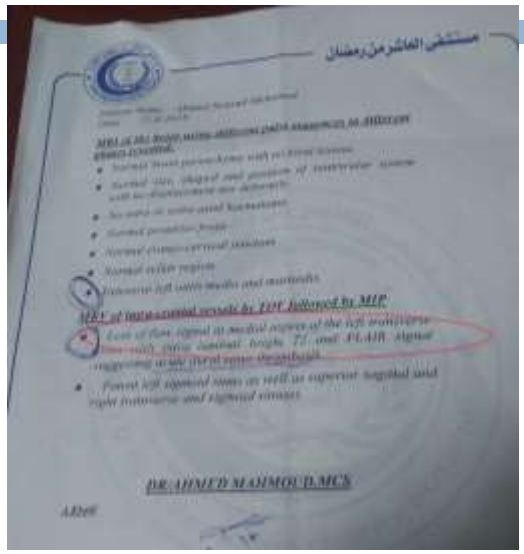
- 27 years old Behcet patient.
- Presented with severe headache, ocular redness and ?diplopia.
- Past history of retinal phlebitis 3 years ago.



examination

- RVA =LVA = 6/6
- AC: quiet and deep
- Fundus exam: no evidence of active uveitis however there was bilateral established disc edema.
- ? R 6th nerve palsy.
- MRV revealed venous sinus thrombosis.





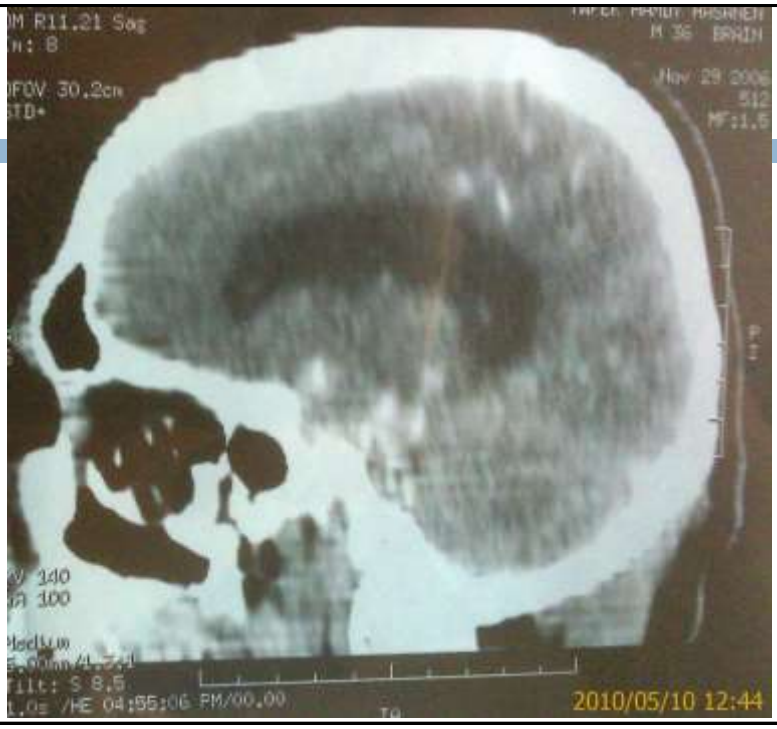
- Patient was treated methyl prednisolone 1g X3 followed by oral 30mg/d
- Monthly cyclophosphamide (shifted later to CSA 300mg) as well as clexan.

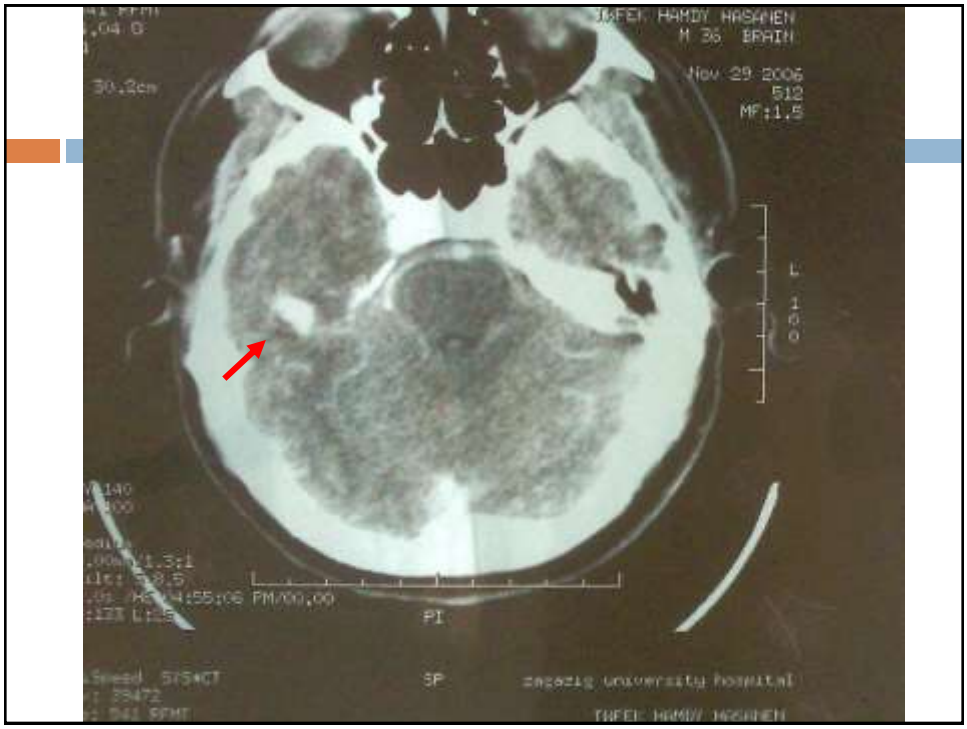
Arterial complications including aneurysms

History

- Young fit male.
- One morning went into coma! (2006)
- Brain CT scan revealed intracerebral hemorrhage– ruptured aneurysm.
- Visiting interventional radiology expert– titanium clip.
- Recovered with no residues.
- Routine ophthalmic exam—6/6 vision

slightly swollen discs ?/ within normal.

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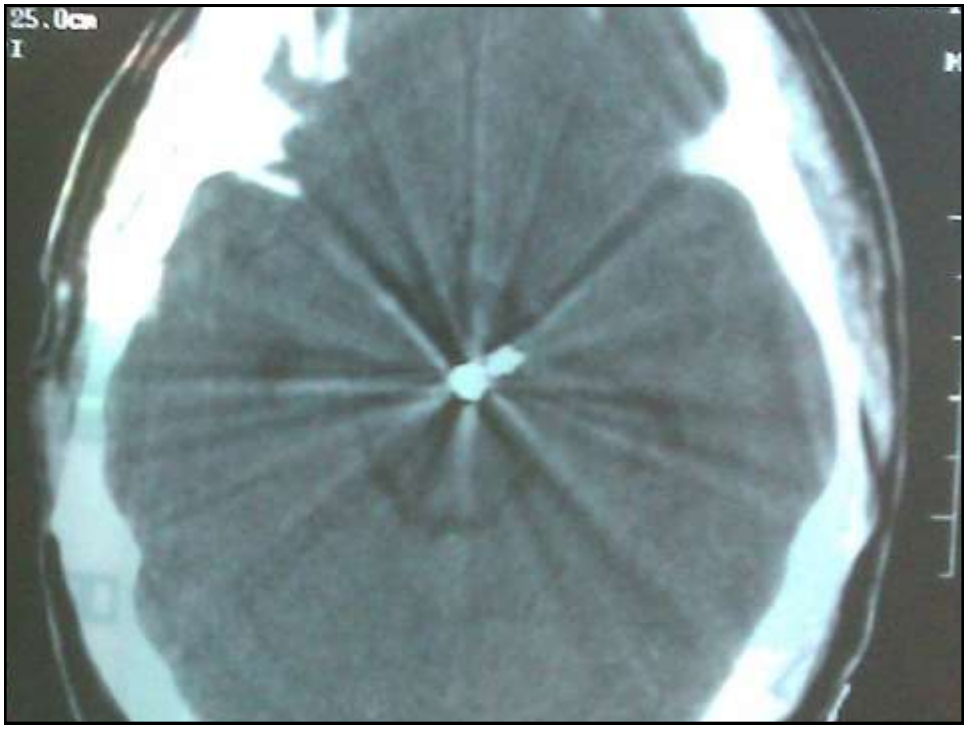
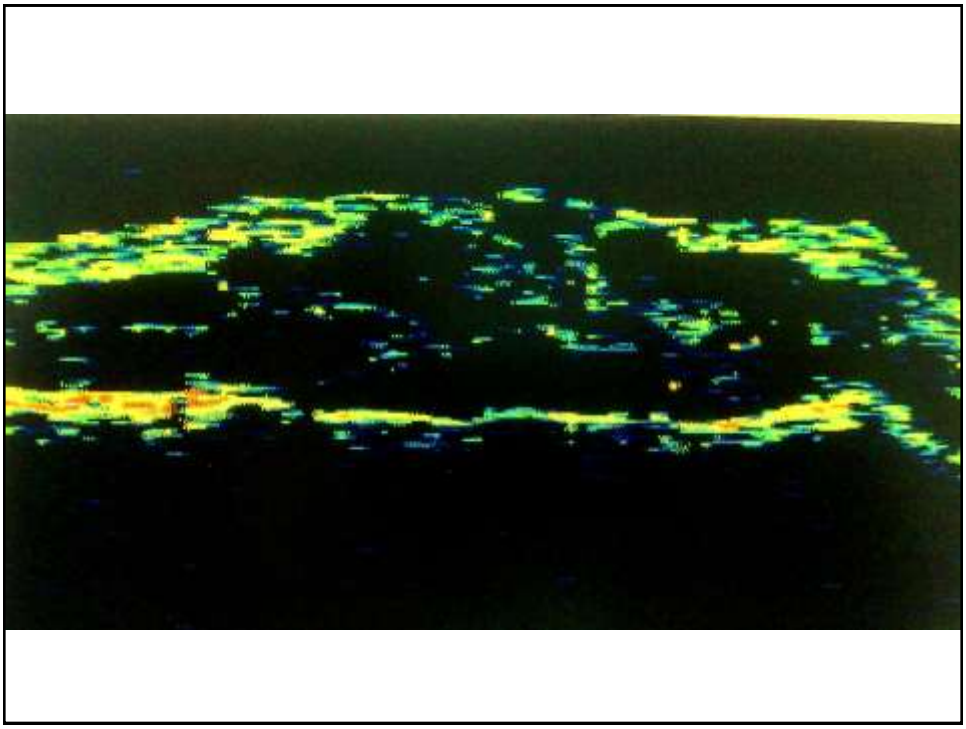
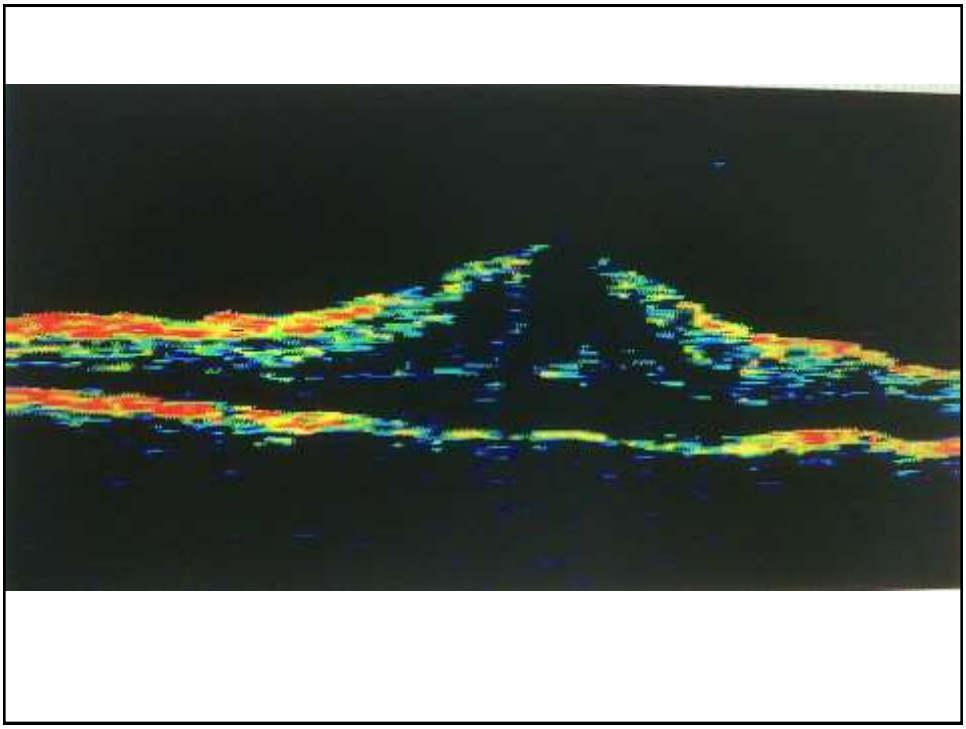
M 36 BRAIN
Nov 29 2006
512
MF:1.5
30.2cm
PI
SP
JAFEI HAMDY HASANEN

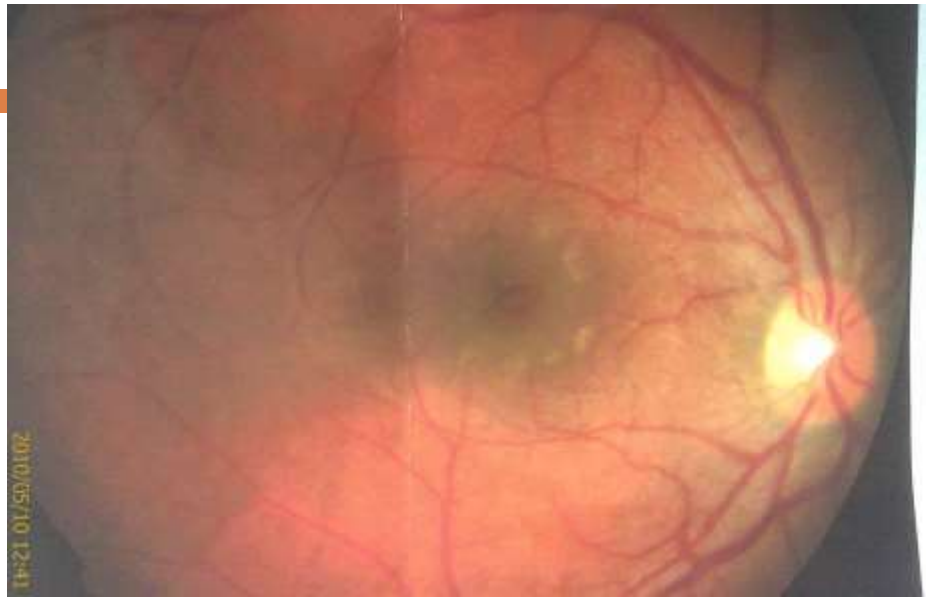

25.0cm

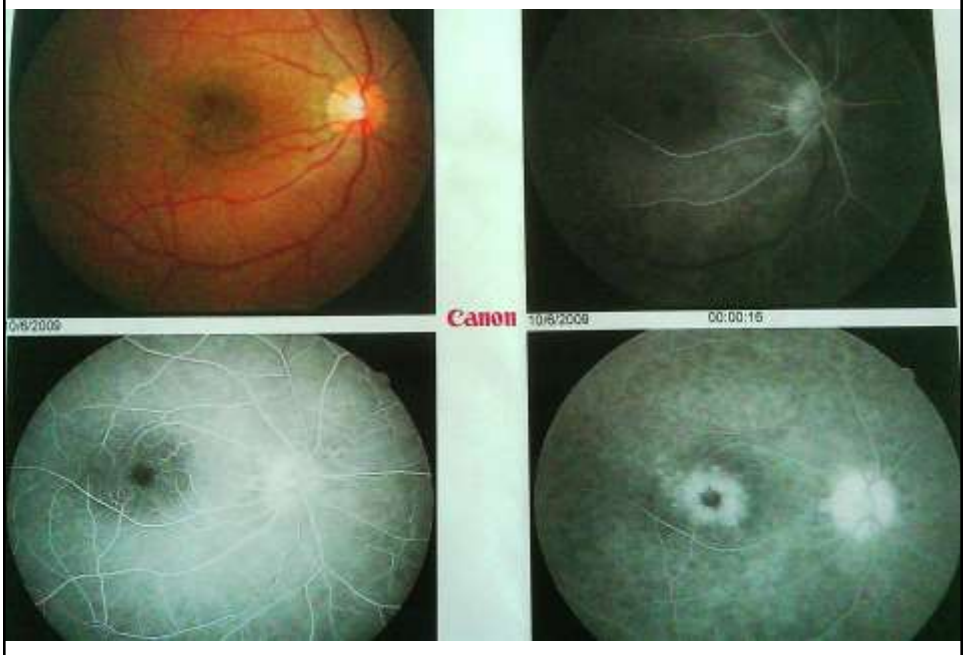
- More than 3 years later (Late 2009) he re-presented with reduced vision (very gradually progressive)
- O/E : VA: 6/12 bilaterally
  AC: Q&D
  Fundus: bil macular edema and slight disc swelling

Cause???





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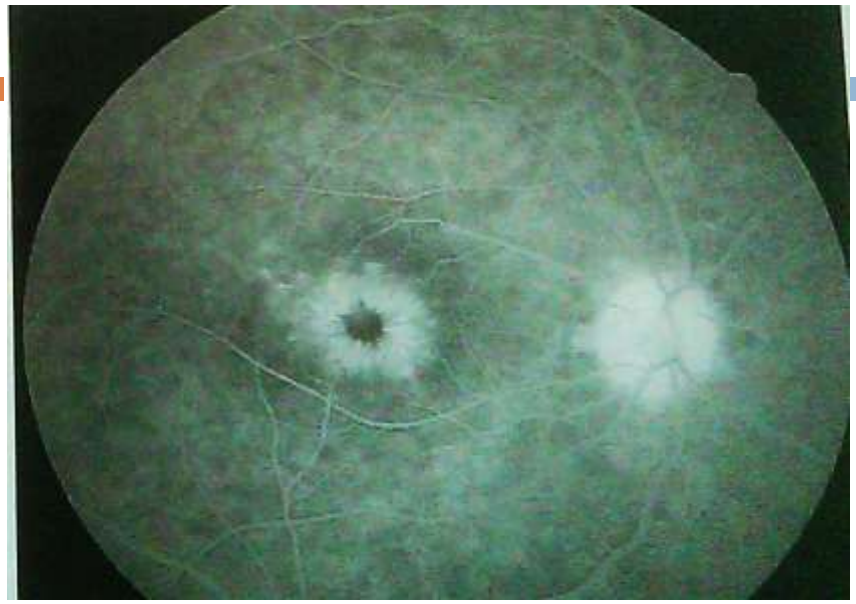
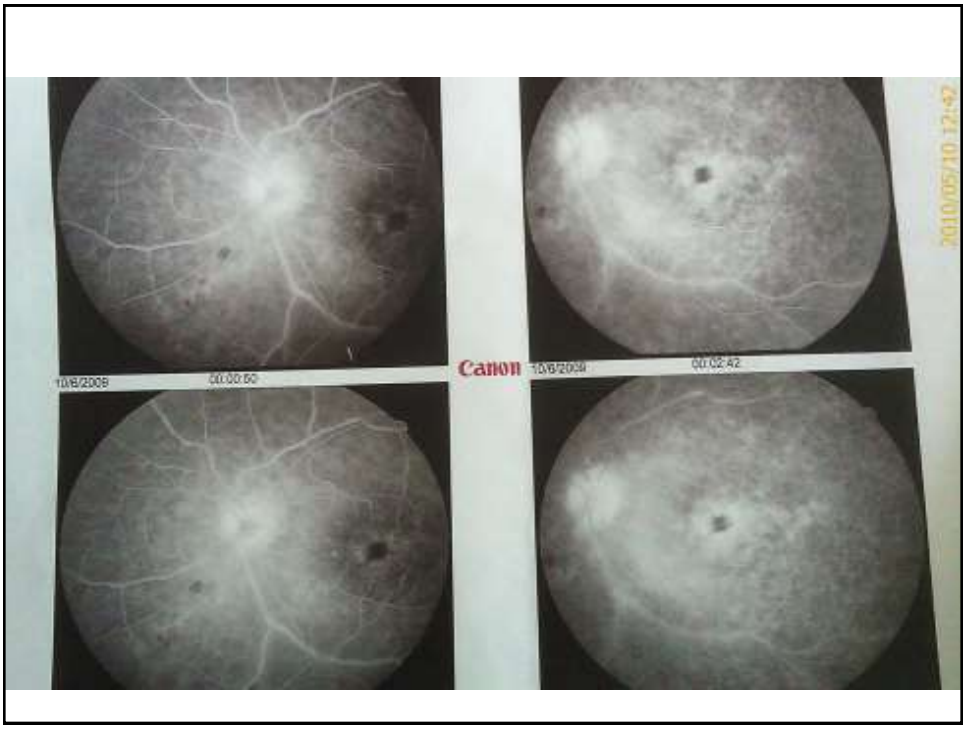


- What is going on?!
- What is the relation between his ruptured aneurysm and the macular edema?!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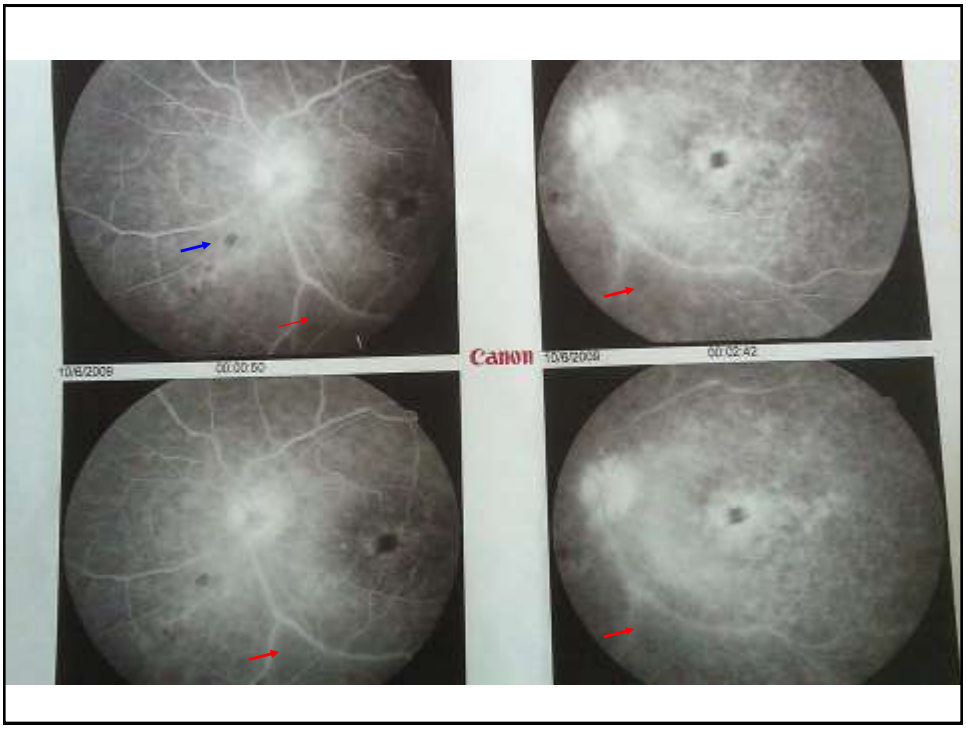
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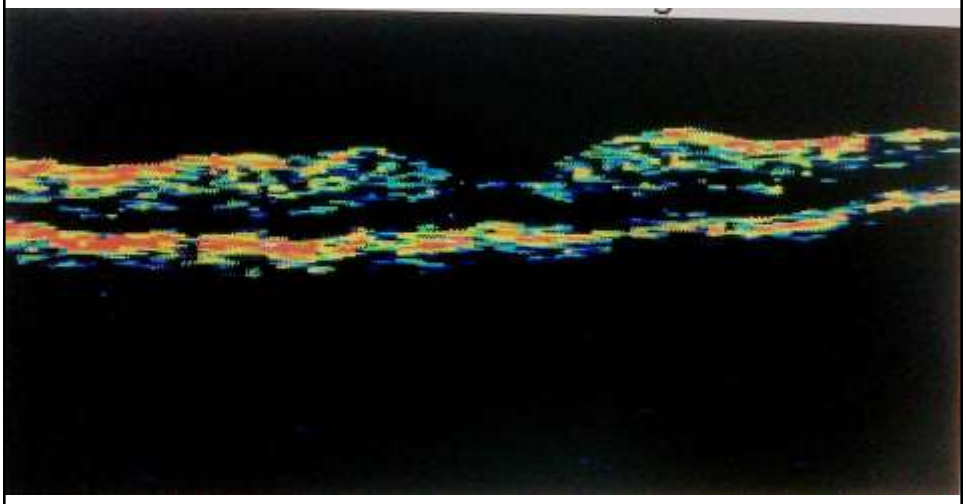
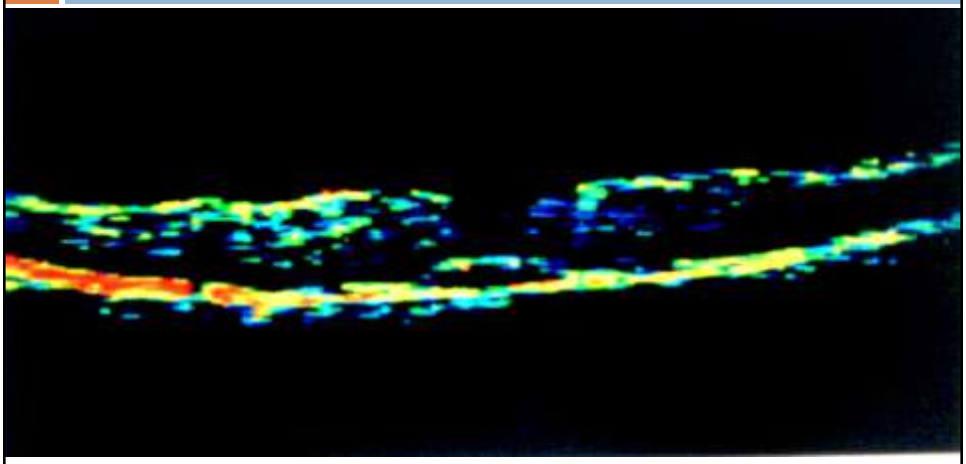
- Diagnosis was revised as retinal vasculitis including cerebral vasculitis (cause of ruptured aneurysm).
- The diagnosis of—Behcet's disease was made?
- Systemic immunosuppression started : prednisolone 40mg + CSA 200mg.
- One month latter he reported great improvement in quality of vision.
- RVA 6/9, LVA 6/6.





- His condition is controlled on prednisolone 10mg/d and Cyclosporin A 100mg twice /day for almost 3years (was later lost from follow up).
- His systemic and ocular conditions are kept under regular observation.

Conclusion

- I wanted to high-light the entity “neuro-Behcet” as a cause of ophthalmic complaint.
- Proper management of these conditions can reverse the pathology allowing rehabilitation of the patients.
- Importance of MRV in all cases of intra-cranial hypertension for the presence of venous thrombosis.

Thank you