

Cyprus - Crete Meeting

10-12-November-2018

Chania Crete

Serious CNS complication of a systemic rheumatic disease

Victor Hajiroussos

19 year old soldier

January 2018

Arthritis fingers and wrists for 2 years on & off

Mouth ulcers and genital ulcers for 2 years

O/E

Arthritis right wrist, oral ulcers, scrotal ulcers

Lab tests

Rh factor neg, ANA neg, ESR 26, CRP 39

Diagnosis: Behcet`s disease Rx colchicine 1.5mg day

End March 2018

Unwell, weakness, drowsy, difficult to walk, change in personality, lough or cry for no reason, jumpy legs.

Admitted

O/E- Loss of balance, tendency to fall, weakness in the legs, brisk reflexes, extensor plantar reflex.

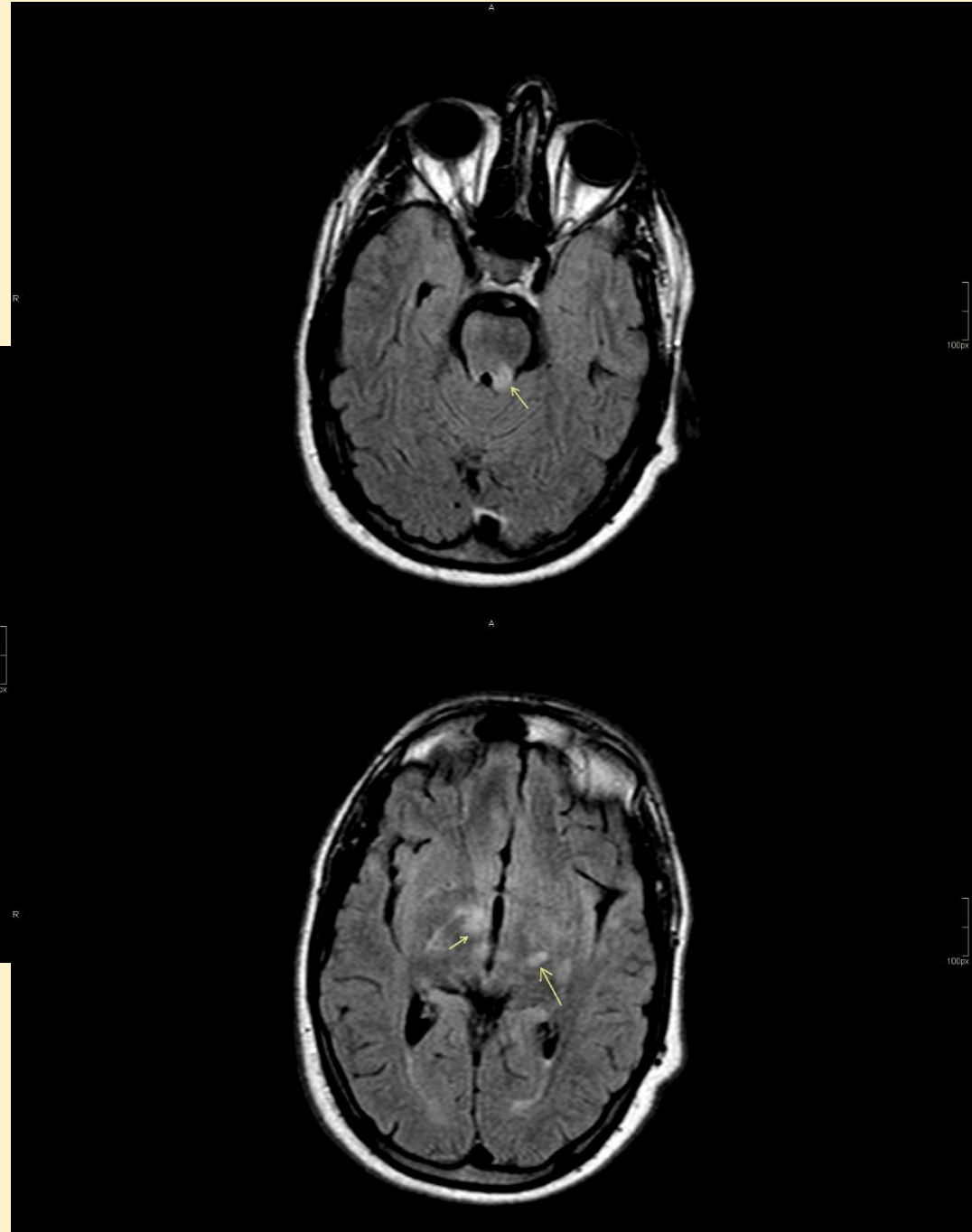
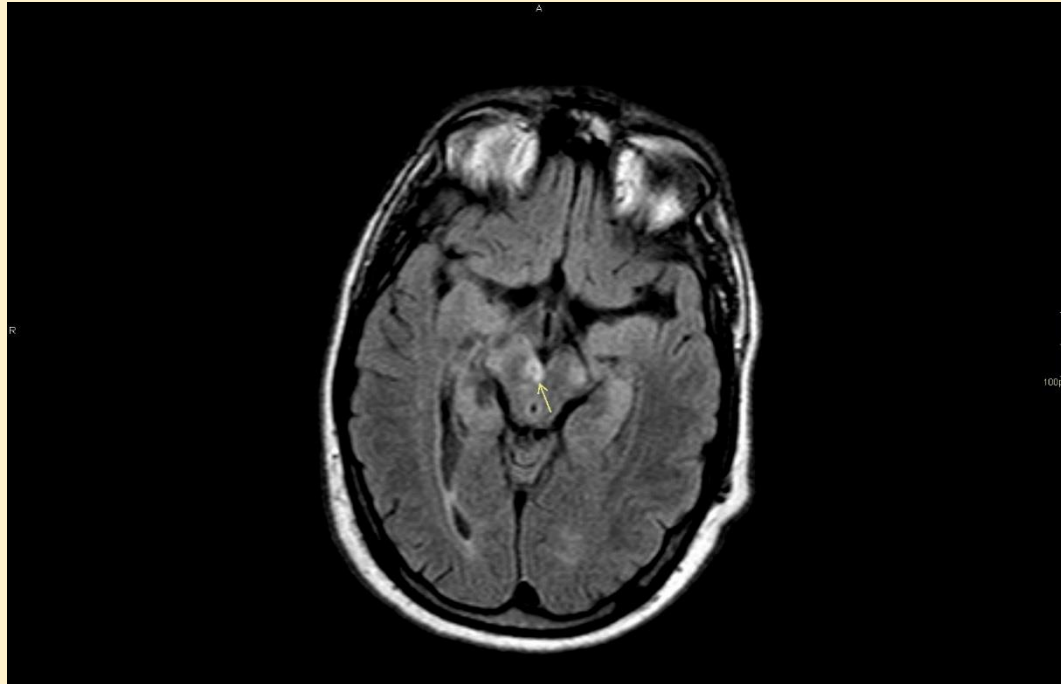
Lab- ESR 20, CRP 5, ANA, ANCA, ACA, LA, ACE,
CSF- glucose 45, protein 46, WBC 18, RBC 200, oligoclonal bands
in CSF and serum, culture neg, PCR viruses neg

MRI Brain - 10.4.2018

Ανάδειξη ενδοπαρεγχυματικών αλλοιώσεων υψηλής έντασης σήματος στις T2 και FLAIR εικόνες εντοπιζόμενες στην δεξιάν πλάγια μοίρα του προμήκη, τα εγκεφαλικά σκέλη, τις έσω κάψες, ιδία αριστερά, την δεξιά υπερμεσολόβιο έλικα και την υποφλοιώδη λευκή ουσία του αριστερού ινιακού πόλου

Μετά την ενδοφλέβια χορήγηση παραμαγνητικής ουσίας παρουσιάζουν οζώδη τμηματική ενίσχυση στο εμβαδόν τους

MRI Brain - 10.4.2018



Rx Solumedrol 1G x5

Prednisolone 60mg reducing

Azathioprine 50mg.....150mg

Good recovery within 3-4 days except for his behavior

Discharged on the 6th day

End of May

Readmission

Difficulty to walk, weakness, diplopia, vertigo, abnormal behavior

Just reduced prednisolone to 30mg

O/E

Unequal pupils, nystagmus, ptosis left eye, slurred speech,

weakness left leg, brisk reflexes, clonus both ankles, up going plantars

Lab

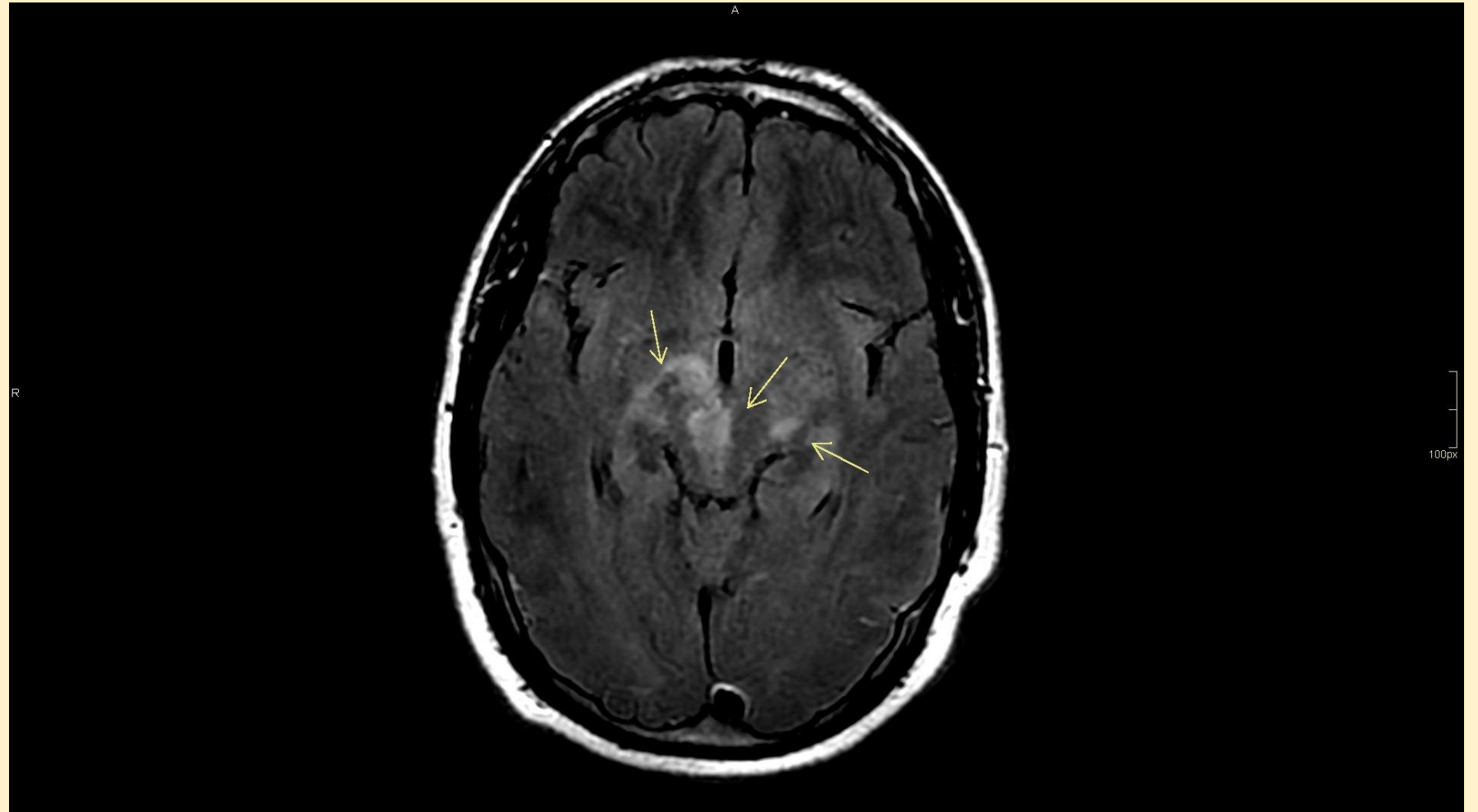
Hb 15.5, WBC 17500, ESR 8, CRP <5

MRI Brain - 25.5.2018

There are areas of increased signal intensity on T2 and FLAIR sequences in the basal ganglia area bilaterally, more prominent on the left side, in the thalamic-hypothalamic areas, in the mesodiencephalic junctions bilaterally, in the splenium of the corpus callosum, in the subcortical white matter of the right parietal lobe, in the left superior cerebellar peduncle and in the right aspect of the medulla oblongata.

There is a new large area lesion in the right aspect of the mesencephalon that was not evident in the previous study.

MRI Brain - 25.5.2018



Rx Solumedrol 1g x7

Prednisolone 60mg

Azathioprine stopped

Infliximab 500mg on the 3rd day

No appreciable improvement on the 5th day

Cyclophosphamide 1g

Gradual improvement, walking with help, still abnormal behavior

Discharged on the 12th day [early June]

Therapy after discharge early June:

Cyclophosphamide 1.3g every 2-3 weeks

Infliximab 500mg as per RA regime

Prednisolone 60mg reducing gradually

Readmitted 18.8.2018

Fever, shivering, off his food, not well for few days

Able to walk but low back pain

Pain in the left hemi thorax especially on deep breathing and laughing

Medication

Four pulses of cyclophosphamide so far

Three pulses of infliximab

Prednisolone 30mg daily

Lab

Hb 15.5, **WBC 15400**, **CRP<5**, **ESR 28**, **SGPT 116**, blood cult neg

MSU neg, CMV IgM neg, Hep B and C neg

CxR normal

Rx Tazocin and gentamicin, prednisolone 30mg continued

Afebrile after 24 hours

Second day of admission

X ray of lumbar spine for low back pain

Loss of height of D12 and L1 vertebrae. Prednisolone reduced 20mg

Next two days

Fever returned, severe headache, mouth ulcers

Repeat CRP 8 [N<10], pro calcitonin neg

Active Behcet`s rather than infection

Rx Solumedrol 1G x3 days and prednisolone 50mg

Next 2-3 days

Weakness in his legs especially the left. Clonus in both ankles and positive Babinski on the left

Severe low back pain unable to turn in bed, he could not walk

Few days later pain in both knees L>R

Tazocin and gentamicin stopped and levofloxacin started

MRI brain, thoracic and lumbar spine, pelvis, left knee

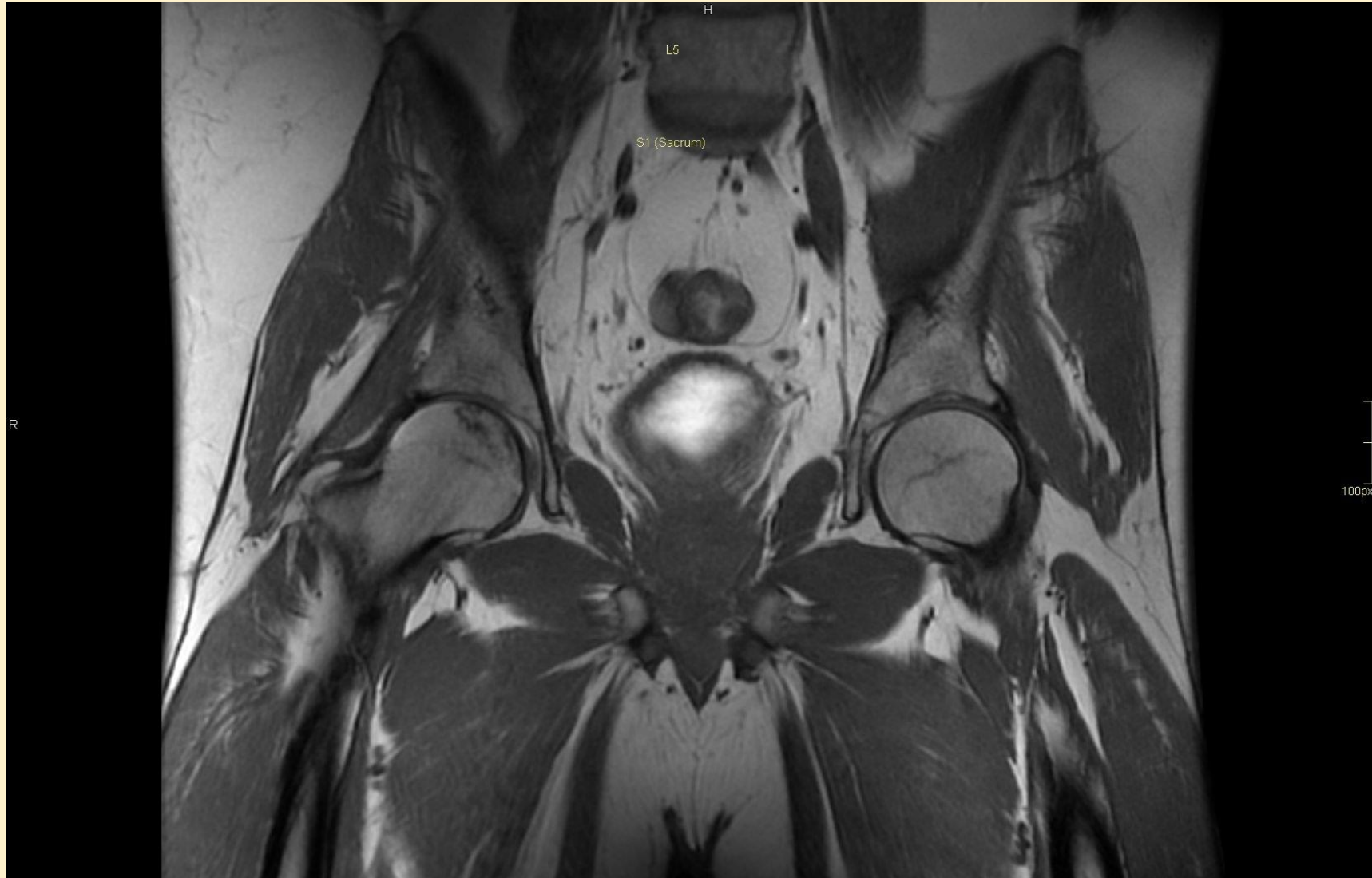
MRI brain 28.8.2018

Four new lesions in the brain stem compared with MRI in June 18
Further new lesions in corpus callosum and external capsule

MRI Thoracic & Lumbar Spine - 27.8.2018



MRI Hips - 22.8.2018



MRI Left Knee - 8.9.2018



Photo 1



Photo 2



Further progress during August admission

Fever subsided, headache improved, still severe low back pain, knee pain, leg weakness gradually improved, depressed, frequently crying and withdrawn. Relatively cooperative

Discharged on the 18th day

Rx Denosumab and teriparatide

Oxycodone [Targinact]

Cyclophosphamide pulses to be continued and prednisolone 50mg reducing, infliximab stopped

Early October 2018

Relatively stable

Lab tests: AP 66, gGT 465, ALT 686, AST 173, LDH 1315, CPK 20

? Cotrimoxazole liver toxicity, ? PPI

Stopped and Cotrimoxazole replaced by inhaled pentamidine

Liver enzymes improved

Late October 2018

Sixth pulse of cyclophosphamide 1.3g

Prednisolone reduced to 30mg

On 4th of November 2018 readmitted

Similar complaints: difficult to walk, weak left leg, weak left arm, slurred speech, drowsy, depression

O/E: Weak left leg, clonus and positive Babinski, weak left arm, some difficulty looking up, left ptosis

Lab: Hb 16.0, WBC 10.100, ESR 13, CRP 6, SGPT 289, SGOT 61, gGT 363

MRI brain

There is deterioration of the T2 hyperintense lesions in the mesodiencephalic junction-cerebral peduncle on the right that shows enhancement on post contrast images

Rx Solumedrol 1g daily x5

Prednisolone 50mg daily reducing

Seems to be neurologically stable on the 4th day

Discharged on the 6th day

Summary

19 year old boy with cerebral Behcet`s, with mainly brain stem, thalamic and basal ganglia involvement and severe behavioral disturbance

He is steroid dependent [$>30\text{mg}$ prednisolone daily] with severe toxicity from steroids

He failed to respond to three pulses of infliximab 500mg

He failed to respond to six pulses of cyclophosphamide

Further management ?? HELP!! **THANK you!**

