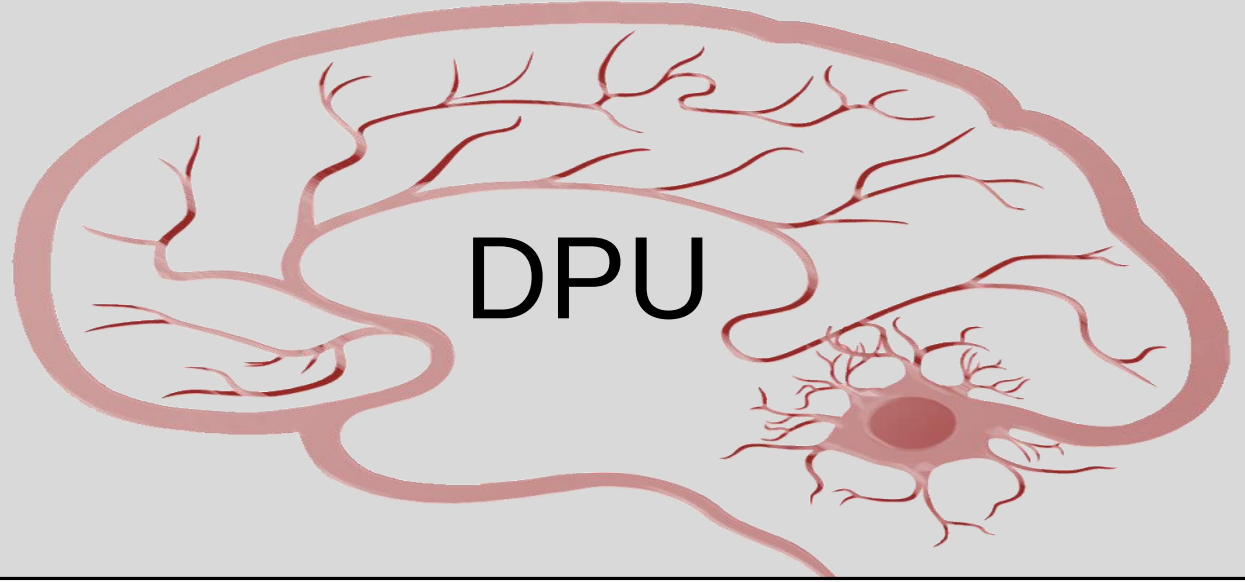


DEPARTMENT OF NEUROLOGY

x

A case of progressive quadriparesis

Case History:

- 49 year old male K/C/O T2DM for 10 years, stable CKD for 1 year presented with chief c/o:

  Tingling and burning in bilateral lower limb for 15 days.

  Weakness of bilateral lower limb for 5 days.

TIME LINE	EVENTS	CLINICAL ASSESSMENT	MANGEMENT
MAY 2021	COVID 19 INFECTION	No Neuro Deficit	O2 therapy, antiviral, steroids.
JUNE 2021	UREMIC SYMPTOMS INCREASED	No Neuro Deficit	Started on peritoneal dialysis
JULY 2021	FEVER OF UNKNOWN ORGIN	No Neuro Deficit	peritoneal fluid ,blood ,urine cultures, hrct thorax ,abdomen All negative
AUG 2021	PUO PERSIST	No deficits. Developed painful skin rashes which faded over 1 month.	PET CT-FDG AVID cervical, mediastinal, illiac lymphnodes, FNAC reactive lymphadenitis. ATT started empirically.
SEPT 2021	Asymptomatic		On 4 drug ATT.

TIME LINE	EVENT	CLINICAL ASSESSMENT	MANAGEMENT
OCT 2021	Tingling burning sensation followed by weakness in b/l lower limb	**UL 5/5 BOTH SIDE** **LL 4-/5PROXIMAL AND DISTAL** PIN PRICK 50% LOSS UPTO KNEE. JPS,VIBRATION IMPAIRED UPTO KNEE Reflex –knee +1 on right, absent on left . ANKLE B/L absent.	NCS:Axonal >>demyelinating neuropathy involving all four limbs.
Mid OCT 2021	Admitted . Weakness increased, now not able to walk or stand. Worsening of CKD Breathlessness, chest pain.(IHD)	UL SHOULDER 4-/5 ELBOW4+/5 LL **HIP 2/5.** **KNEE 2/5** **ANKLE 2/5.**	ANA IFA ,ANCA-NEGATIVE. CSF –PROTIEN -45,SUGAR-61(BSL-89)CELL-2. Started on PLEX and HAEMODIALYSIS. Nerve biopsy done.

SUMMARY

- 49 year old male well controlled T2DM,stable CKD controlled on drugs, presented with sensory motor quadriparesis without bladder bowel cranial nerve involvement with background of acute worsening of CKD,respiratory function and developing acute coronary syndrome.

DIFFERENTIAL DIAGNOSIS:

- DRUG INDUCED PERIPHERAL NEUROPATHY.
- DIABETIC SENSORY MOTOR POLYNEUROPATHY.
- CIDP
- VASCULITIC NEUROPATHY.

TIME LINE	EVENTS	CLINICAL ASSESSMENT	MANAGEMENT
NOV 2021	Further detoriation in general condition Quadriparesis. Breathlessness ,chest pain. Dxn M.I, Cardiac Arrest , Intubated , Resuscitated.	UL-1/5 LL-0/5	PLEX 7 CYCLES done. MMF started. Steroids started i/v/o COP improvement in respiratory symptoms.
	Even after recovery persistent breathlessness . Always sitting. HRCT Repeated- cryptogenic organising pneumonia.		
DEC 2021	ESRD/COP/IHD/QUADRIPERESIS	UL -2/5 LL-0/5	Thrice weekly dialysis. MYCOPHENOLATE. Oral steroids.

PRATHA CHAKRABARTY
Pune

PID:35005249

Age: 45.00 Years Sex: MALE

Reference:Dr.--
Sample Collected At:
--
Dr. D. Y. Patil Medical College,
Hospital & Research Centre,
Sant Tukaram Nagar, Pimpri Colony, Pune.

Collection Date:
23-10-2021 01:13 AM
Registration Date:
23-10-2021 01:13 am
Report Date:
27-10-2021 04:06 PM

HISTOPATHOLOGY REPORT

PE no. :	B/4010/21
linical details :	
lature of specimen	**HPE of right sural nerve.**
Gross Examination	Received single tissue piece measuring 0.5 cms in length. All for processing.
Microscopy :	The nerve biopsy shows elements of epimysium, perimysium and endomysium along with blood vessels and nerve elements. The blood vessels are essentially normal in appearance as are the nerve fibres (Cut in both transverse and longitudnal section)
Diagnosis :-	**Nerve biopsy - Within normal histological limits.** Remarks- Opinion only on H&E study of biopsy.

Note :-

If Specimen preserved, it will be retained in the laboratory for 12 weeks from the date of receipt.

National Institute of Mental Health and Neuro Sciences

Hosur Road, Bengaluru , 560029

Department of Neuropathology

M-2792

UHID:	EXT21015914	**Referring Hospital:**	Hospital, Pune, Maharashtra 411018
MRD No :		**Referring Dept:**	
Patient Name:	**Mr. PARTHA CHAKRABORTY**	**Sample Collection Date:**	26/10/2021 11:33 AM
Age :	50 years	**Lab Reference No:**	X-4279/2021
Gender:	Male	**Report Generated Date:**	05/11/2021 03:14 PM
Ward Name/Collection Centre:	Biopsy Room	**Lab Name:**	Neuropathology

Sample Details : H-2110260019 (Nerve)

NERVE BIOPSY

Nature Of Specimen:
Received 1 nerve segment 0.8cm. All processed-A1, A2
Grossed by Dr.Poonkodi 26/10/2021

Histopathology Report:
Nerve biopsy shows increased hyalinisation in the epineurium along with focal neovascularisation and perivascular sprinkling of lymphocytes, better seen in the section stained for collagen. The endoneurium shows focal acute axonal breakdown with myein ovoid formation. Kpal stain for myelin shows mild to moderate non uniform loss of myelinated fibres with many regenerating clusters. Pinch and stretch artefacts are present. Perl's stain for hemosiderin is negative. LCA immunostain highlights the sparse lymphocytic infiltrates in epineurium (Predominantly around blood vessels)

Final Impression:
Inflammatory neuropathy with axonopathy, consistent with 'probable' vascultis; left sural nerve biopsy

Drafted By:
Dr. Abhishek Chowdhury
DM Resident
05/11/21

Reported By:
Dr Yasha TC
Professor
05/11/21

Level 1 Entered by
(Dr.Yasha T C)

Level 2 Verified by
(Dr.Yasha T C)

Dr. Yasha TC
Professor & Head

FINAL DIAGNOSIS

SERONEGATIVE VASCULITIC NEUROPATHY

- As the patient was not responding to immunotherapy given.He was having multiple system involvement in a short span with very low general condition. It was thought to be a part of systemic vasculitis.(10% patient may be ANCA negative)
- Cyclophosphamide therapy was advised.
- Risk vs Benefits were explained.
- Cyclophosphamide in modified dose for 7 cycle was given at a monthly interval.
- Presently he is on rituximab therapy.

Present status:

Vasculitis neuropathy:

- Vasculitis affects the epineurial and endoneurial vessels supplying peripheral nerves, inflammation and subsequent critical ischemia result in nerve damage .

- Vasculitic neuropathy is usually only one feature of a systemic vasculitis that can involve the skin, lungs, kidneys, and other organs.

- Vasculitic neuropathy typically presents acutely as a focal, painful neuropathy that progresses over weeks to involve other regions of the body with objective sensory or sensorimotor deficits on serial examinations.

- Three primary patterns of vasculitic neuropathy are present, which include multiple mononeuropathy (also called mononeuritis multiplex), distal sensorimotor polyneuropathy, and plexopathy.

UNUSUAL CASE OF COMPLICATED HEREDITARY SPASTIC PARAPLEGIA

HISTORY:

33 year old male , farmer, born out of non-consanguineous marriage , presented with :

-Stiffness in lower limbs since 4 years

-Multiple episodes of seizures since 4 years

No history of tingling numbness in extremities

No history of DOV/hearing loss

No history of bladder /bowel involvement

No history of toxin exposure / high risk sexual behavior

- No Family history
- Normal birth history
- Significant delay in milestones
- Received levodopa for 2 months

Examination :

- Blood pressure – normal
- MMSE – 21/30 (moderate cognitive impairment)
- Bilateral lower limb grade-3 spasticity (modified ashworth scale)
- Bilateral lower limb power grade 4 +/5
- DTR: UL ++, LL +++
- Plantar- ↑/ ↑
- Gait – spastic
- Sensory exam - N

Differential diagnosis :

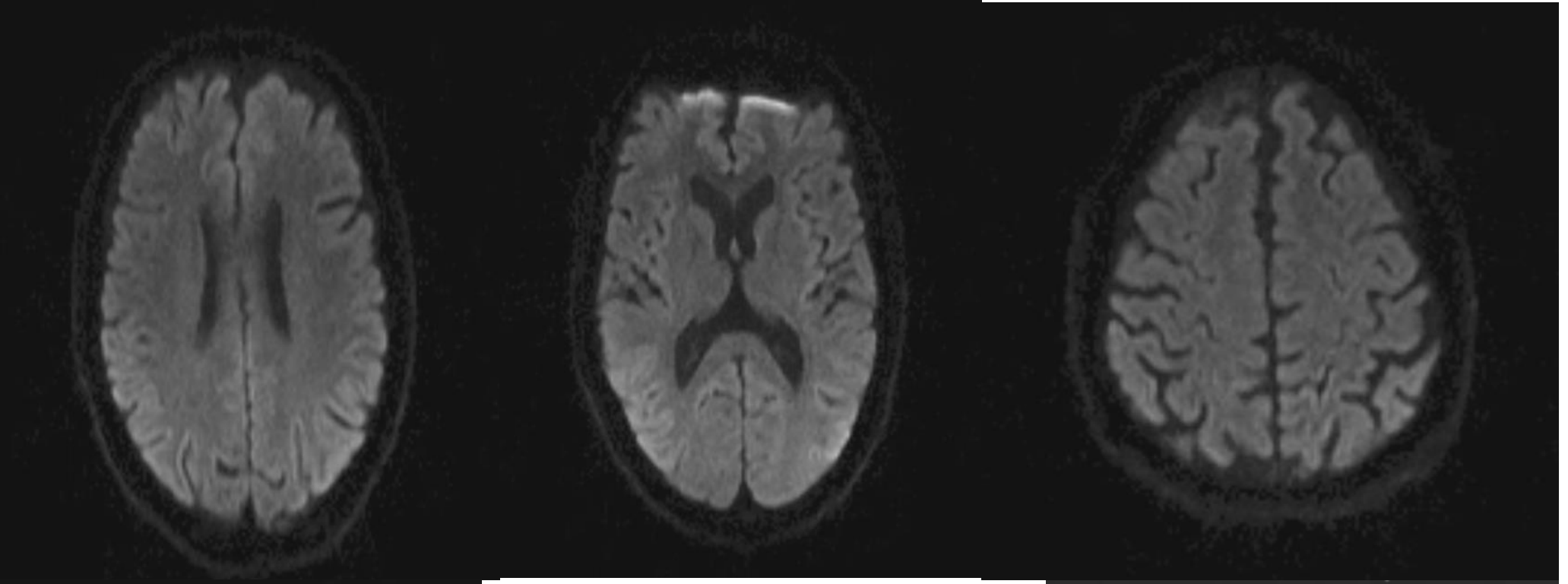
-Complicated HSP

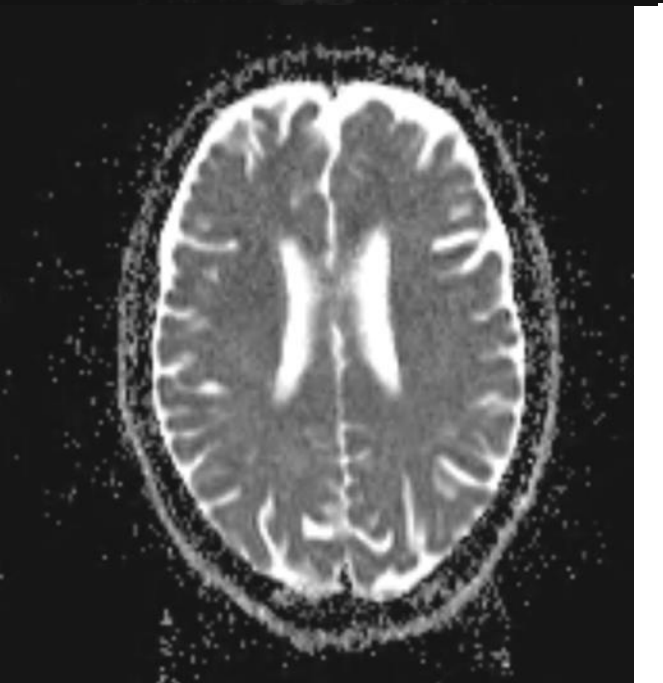
-Adrenomyeloneuropathy

-HIV myelopathy

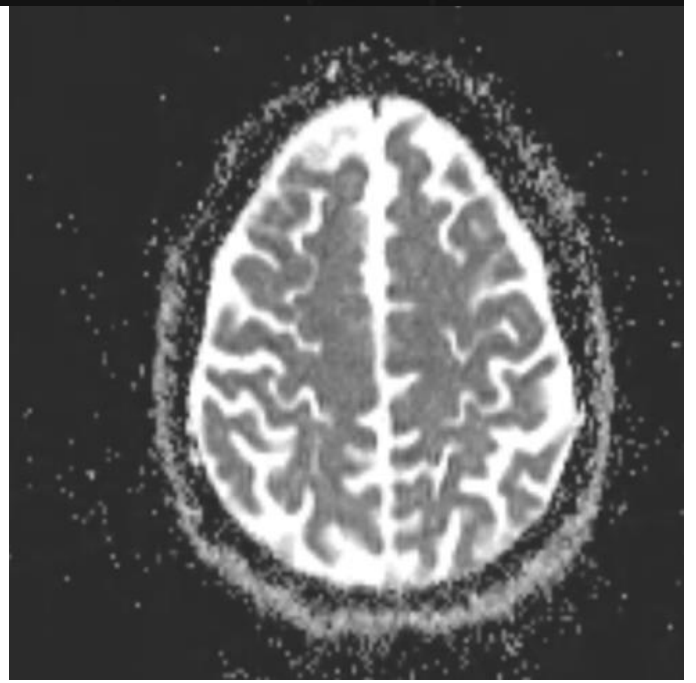
Investigations :

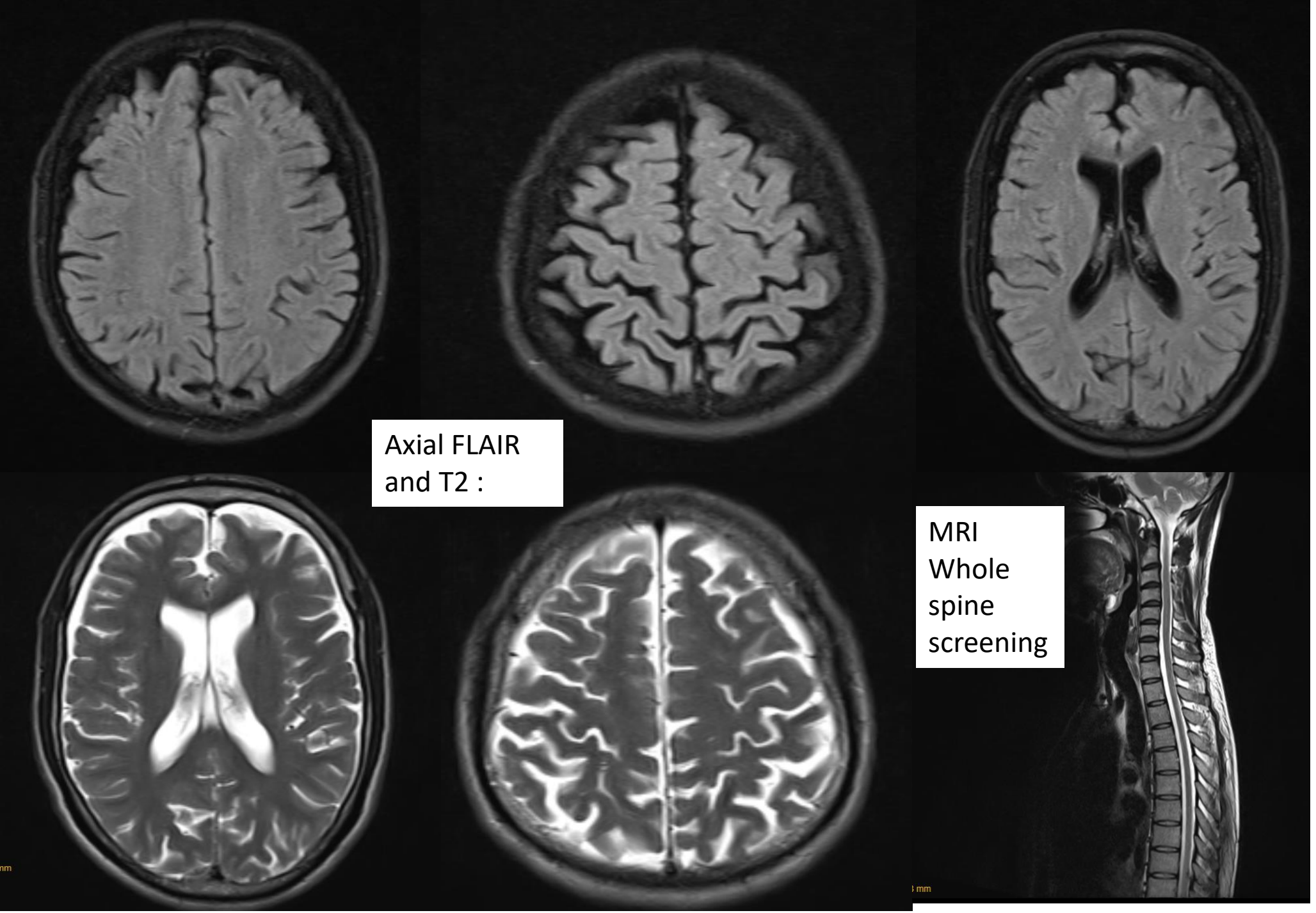
- MCV, serum-B12 – normal
- HIV- negative
- EEG - normal





Axial diffusion and
ADC :





Axial FLAIR and T2 :

MRI Whole spine screening

CSF examination :

- CSF: P– 38 mg/dl , G -**10** mg/dl (corresponding BSL-128) , 2 cells

- Progressive myelopathy
- Seizures

} 4 years

- Developmental delay, scholastic backwardness
- Normal Brain and Spine MRI

Low CSF sugar

? Non- infectious causes of low CSF sugar

Diagnosis ????

LifeCell Diagnostics

Lifecell International Private Limited
#26, Vandalur Kelambakkam Main Road, Keelakottaiyur, Chennai 600127
CALL 1800 266 5533
www.lifecell.in

Name:	[redacted]	Case ID:	**KT20900103425**
Age:	**32 years**	Sample Type:	**Peripheral Venous Blood**
Sex:	**Male**	Sample Collection Date:	**07/09/2022**
Referring Clinician:	[redacted]	Sample Receipt Date:	**10/09/2022**
Test Required:	**Whole Exome Sequencing**	Reporting Date:	**23/09/2022**

RESULT SUMMARY

Pathogenic variant causative of the reported phenotype was identified
*Correlation with clinical profile and family history is required.

FINDINGS RELATED TO PHENOTYPE

Gene & Transcript	Variant	Location	Zygosity	Disorder (OMIM)	Inheritance	Classification
SLC2A1 NM_006516.4	c.988C>T (p.Arg330Ter)	Exon 8	Heterozygous	GLUT1 deficiency syndrome 2, childhood onset (612126)	Autosomal Dominant	**Pathogenic**

GLUT-1 deficiency syndrome with AD inheritance pattern

Currently being treated with ketogenic diet and levetiracetam

Family members could not be screened because of financial constraints

Discussion :

- Impaired glucose transport through blood brain barrier.
- Inherited in an autosomal dominant trait, which results from mutations of SLC2A1 gene encoding GLUT1

- Clinical spectrum has broadened to include non-classic phenotypes, such as paroxysmal exercise-induced dyskinesia and epilepsy, paroxysmal choreoathetosis with spasticity, atypical childhood absence epilepsy, and myoclonic astatic epilepsy.

- Laboratory hallmark for GLUT1-DS is low glucose of the CSF (hypoglycorrhachia) with normoglycemic .
- Less reliable biomarker is the glucose ratio of CSF to blood below 0.45
- NGS is definitive.
- **Ketogenic diet is the gold standard** treatment for GLUT1-DS

Infectious Etiologies commonly associated with hypoglycorrhychia :	Infectious Etiologies uncommonly associated with hypoglycorrhychia :
• Bacterial meningitis (including atypical bacteria like nocardia and brucella) • Fungal meningitis • Mycobacterial (tuberculous meningitis) • Amebic meningoencephalitis • CMV-associated progressive polyradiculopathy or meningoencephalitis	• Syphilitic meningitis • Lyme meningitis • Viral meningitis • Neurocysticercosis • CNS toxoplasmosis

Non-infectious causes :	
• Carcinomatous meningitis • Leukemia/lymphoma with CNS involvement • Subarachnoid hemorrhage	• Cholesterol-induced leptomeningitis secondary to Currarino syndrome • Neurosarcoidosis • Rheumatoid meningitis • Systemic lupus erythematosus with CNS involvement • Neuro-Behcet’s Disease • Dermoid cyst • Granulomatous angiitis of the central nervous system • Malignant atrophic papulosis • GLUT 1-deficiency syndrome

References :

- 1. **Glucose Transporter Type 1 Deficiency Syndrome Synonyms: De Vivo Disease, Glut1 Deficiency Syndrome, Glut1 DS** Dong Wang, MD, Juan M Pascual, MD, PhD, and Darryl De Vivo, MD

UNUSUAL COMPLICATION OF POISONOUS SNAKE BITE

- 17 /F was bitten by a snake following which she developed
- Breathlessness
- Chest pain
- Difficulty in swallowing
- Drooping of both eyelids

- She was admitted to a private hospital
- Was intubated and was on ventilatory support for 6 days
- NO documented hypotension/ hypertension
- NO history of swelling/ bleeding at site of snake bite
- 40 vials of polyvalent antisnake venom was administered

- She was extubated on 7th day after which she developed diminution vision in both eyes which improved over 2 days
- involuntary movements of all four limbs and tongue on 9th day
- Referred to our hospital for further management

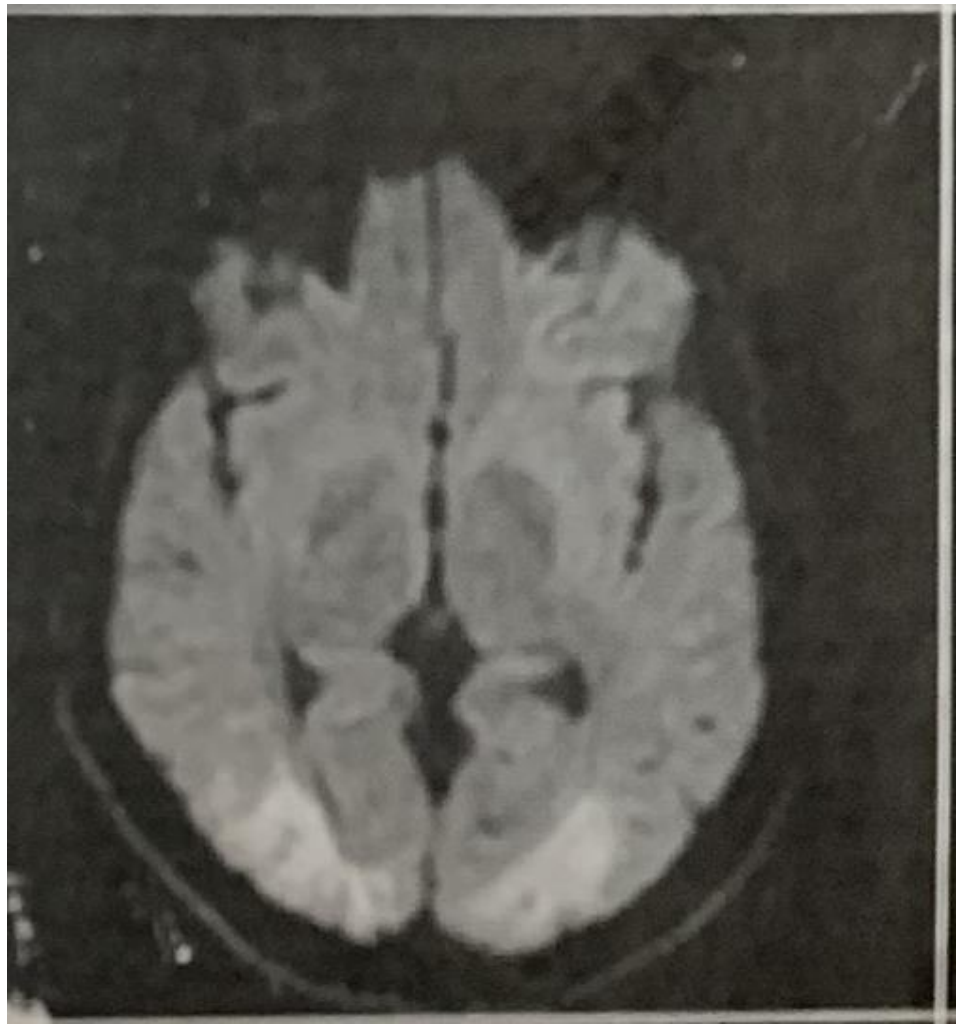
- ON EXAMINATION
- P-80bpm
- BP 110/70 mmhg
- She was conscious oriented and obeying commands
- Heart sounds were normal
- She had generalised chorea in all four limbs and tongue tremors .

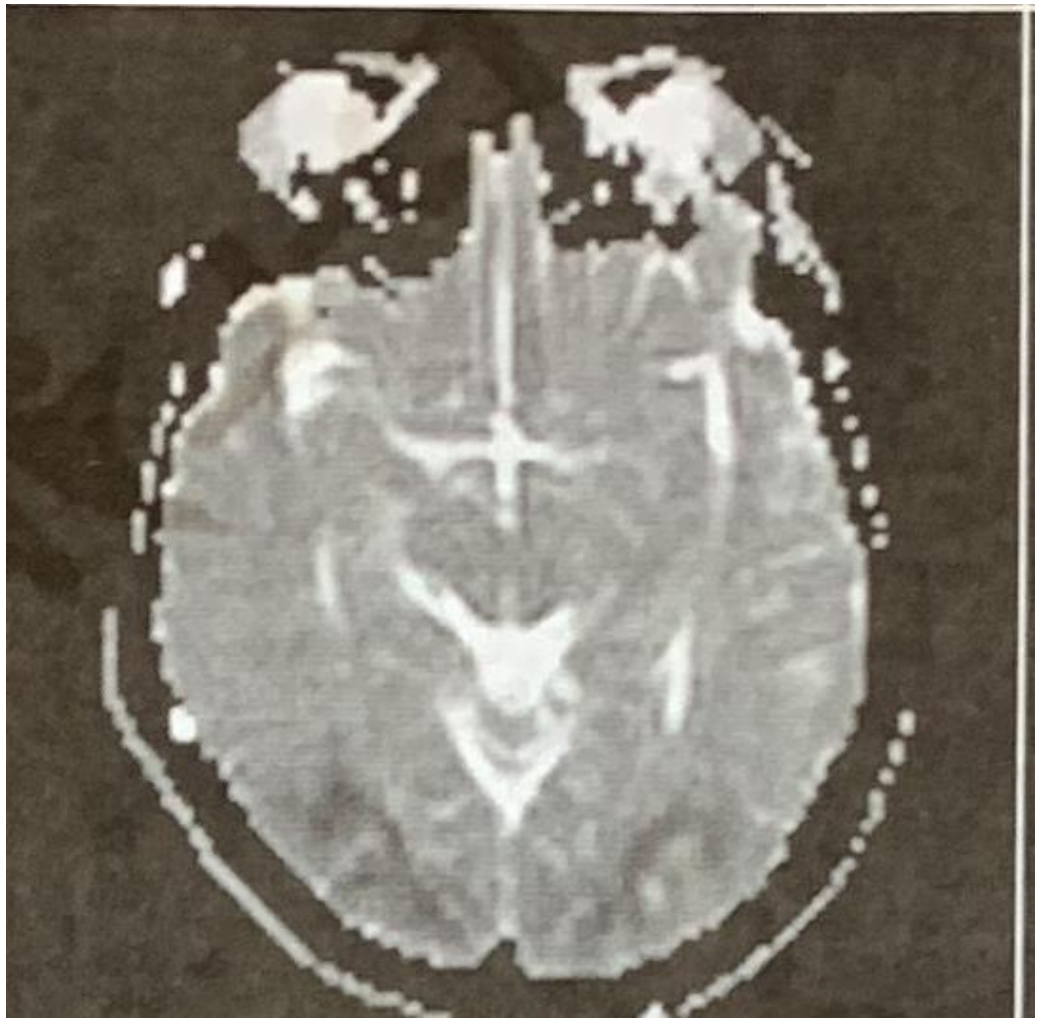
- Fundus examination was normal.
- Visual acuity was normal in both eyes.
- Pupils were bilaterally equal reactive to light .
- Slit lamp examination did not reveal any Kayser Fleischer rings.
- Other neurological examination was normal.

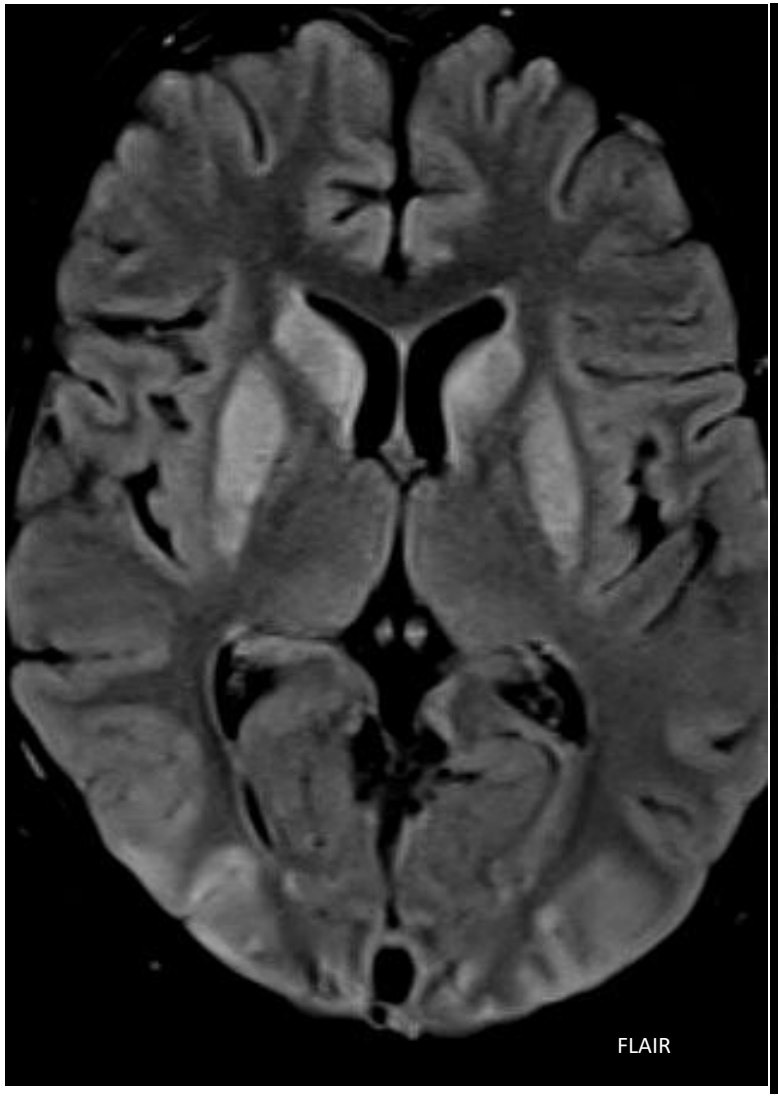
- GENERALISED CHOREA WITH TONGUE TREMORS
- ?DRUG INDUCED
- ?IMMUNE MEDIATED (? AUTOIMMUNE ENCEPHALITIS)
- ? SYDENHAM’S CHOREA
- ?? RELATED TO SNAKE BITE

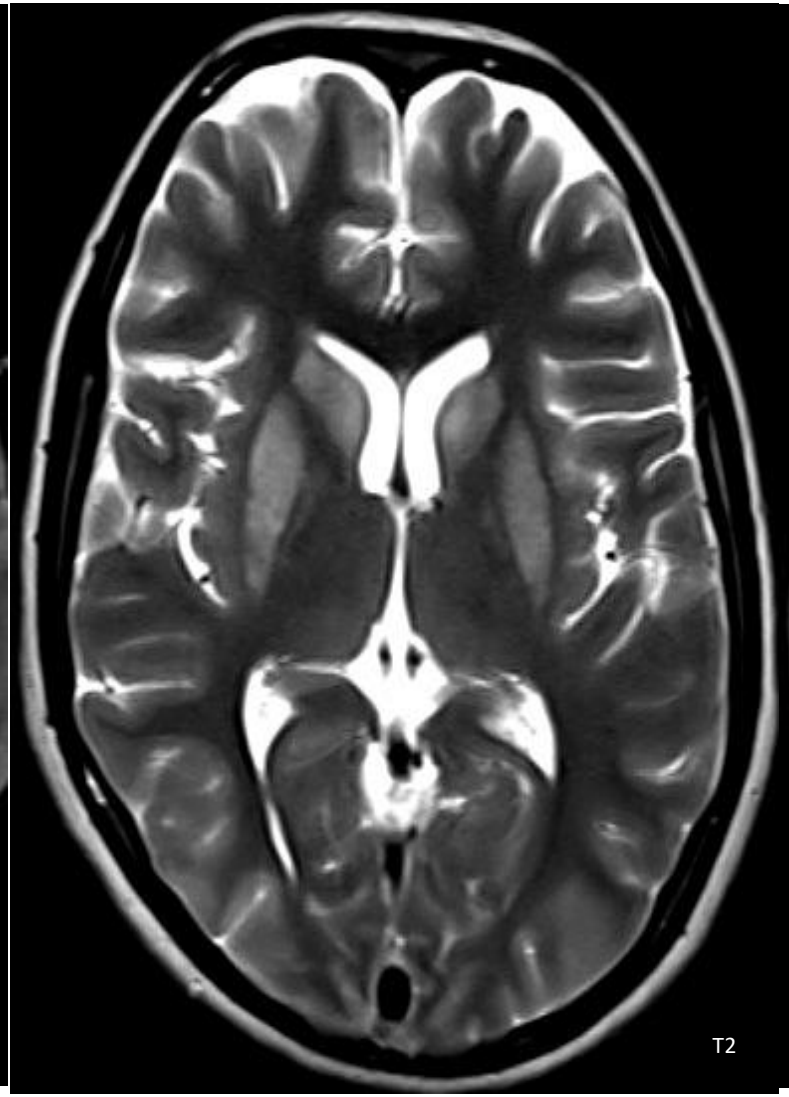
- ON INVESTIGATION
- Routine investigations - complete blood count (Hb - 12.7), electrolytes, liver function test, coagulation profile, renal function tests, blood sugar levels ,ASO Titre, CRP and 24 hrs urinary copper, thyroid profile, were normal. ANA was negative .
- 2DECHO was normal
- CSF was normal.

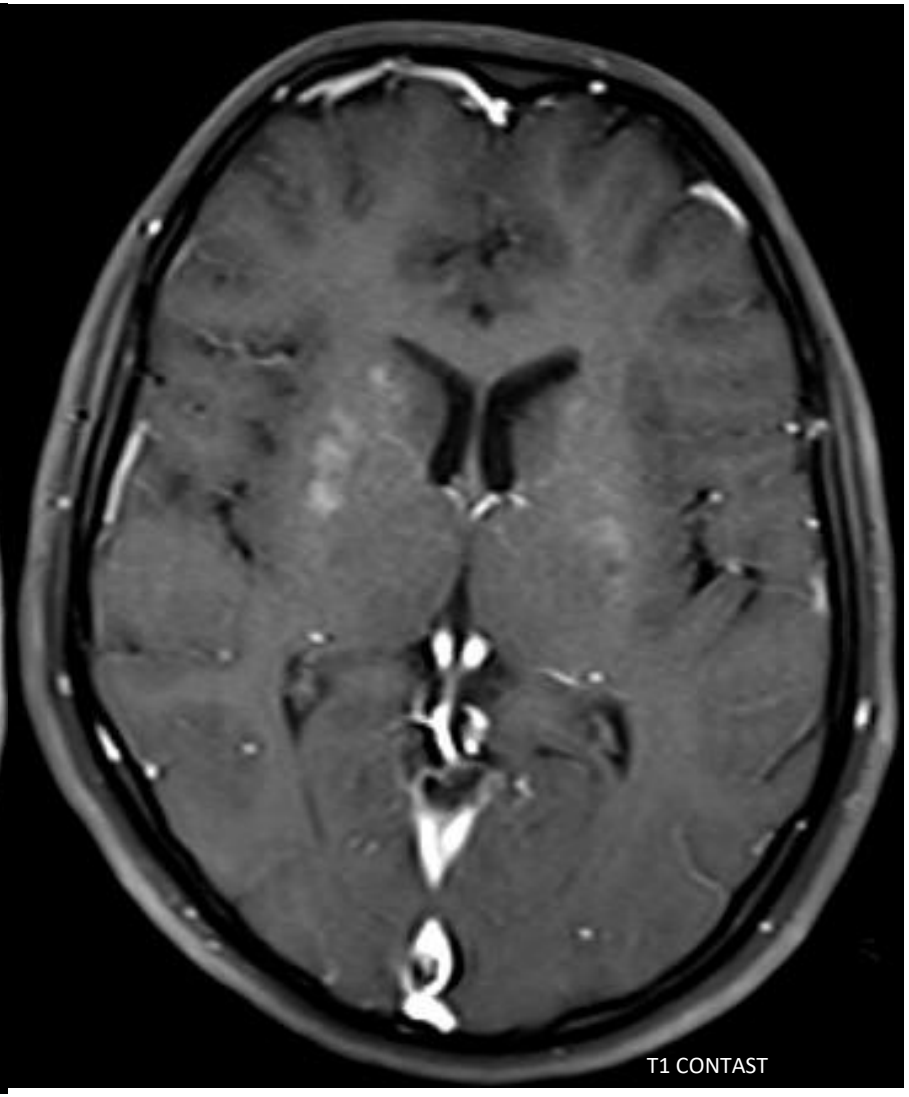
MRI DONE AT OUTSIDE HOSPITAL



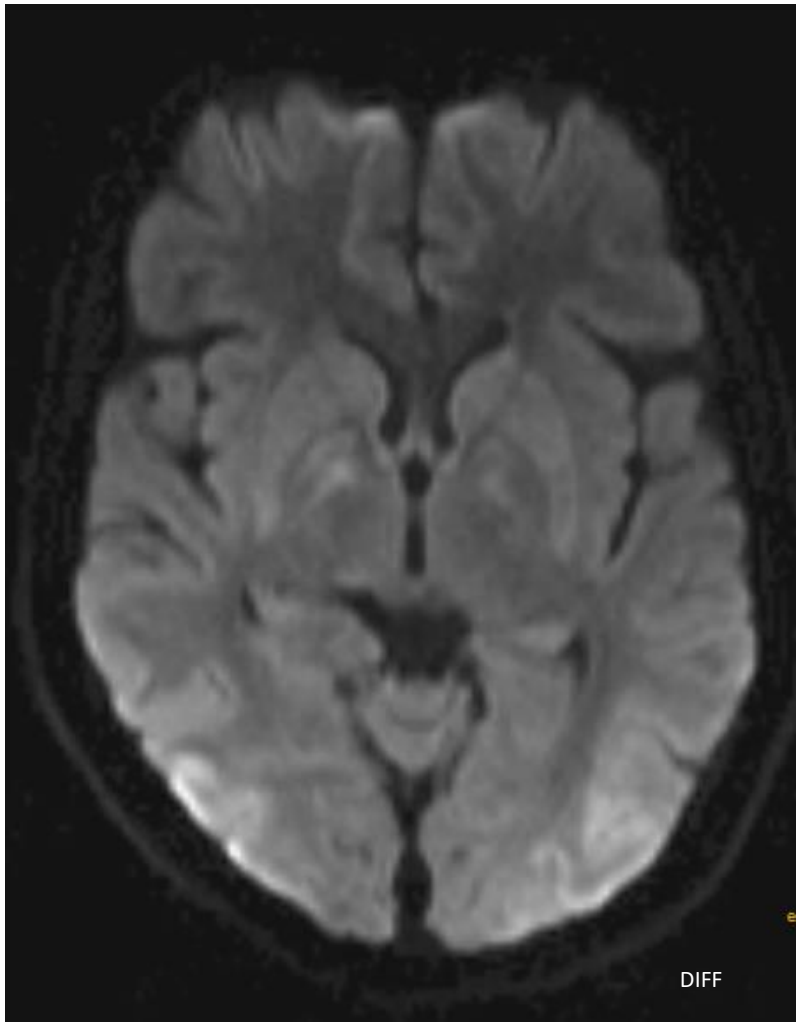








MRI brain revealed bilateral T2 hyperintensities in both lentiform nuclei and caudate, bilateral parieto-occipital region involving both grey and white matter with diffusion restriction .MRI of spinal cord was normal.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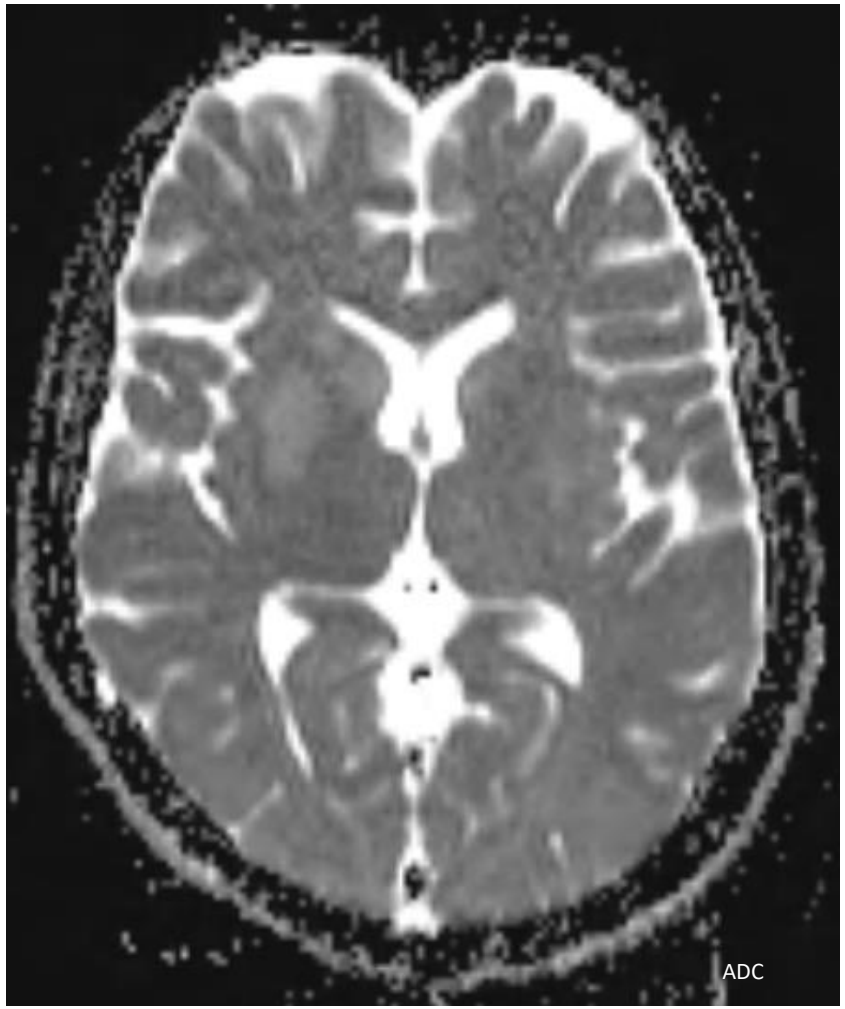
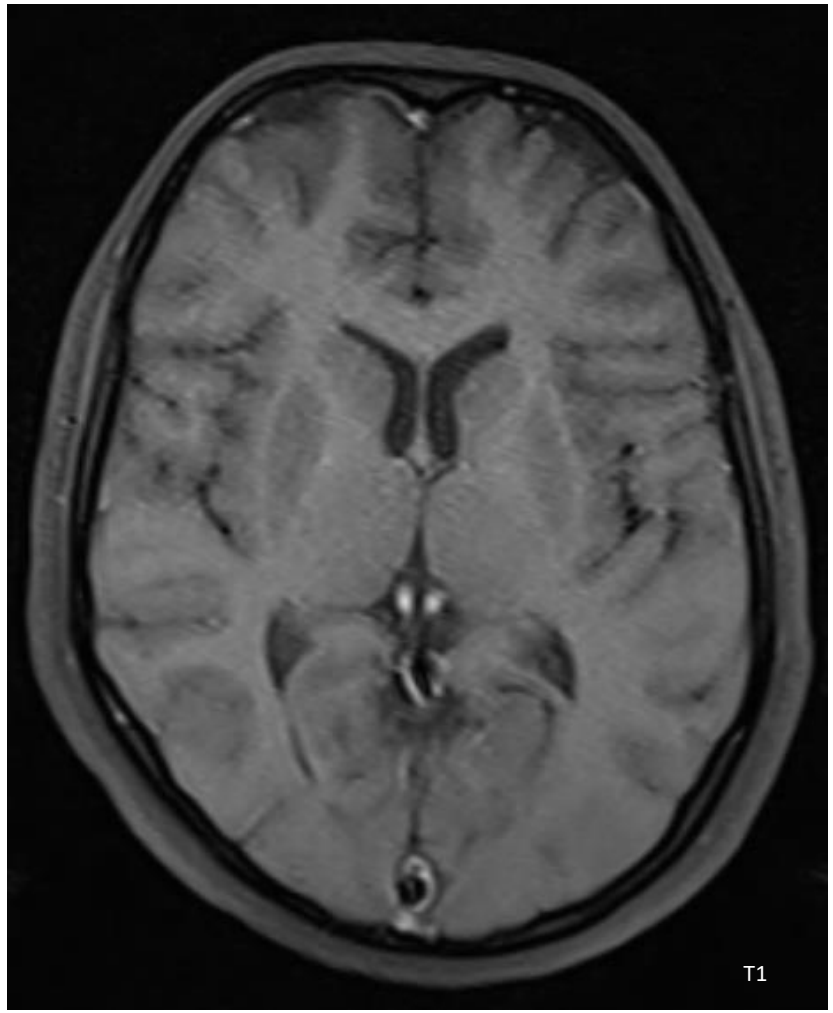

ADC


T1

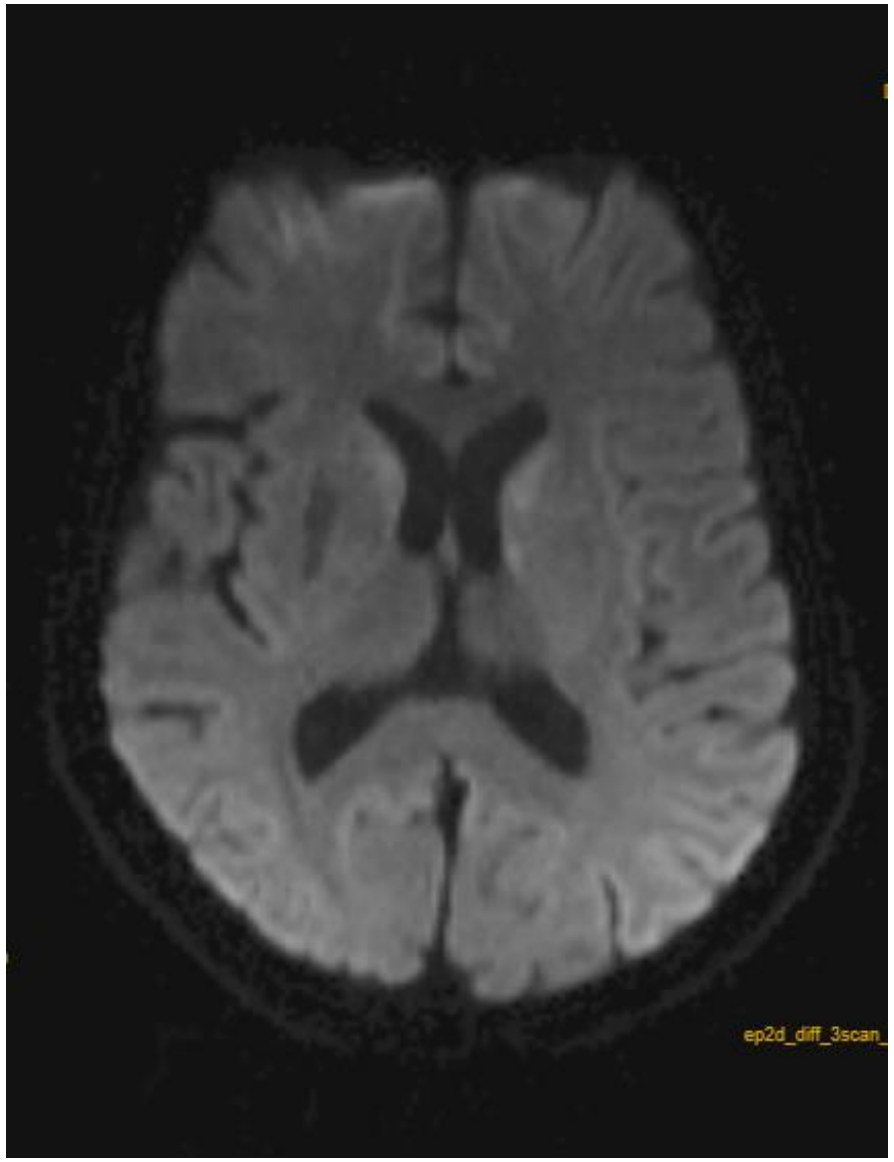
DIAGNOSIS ??

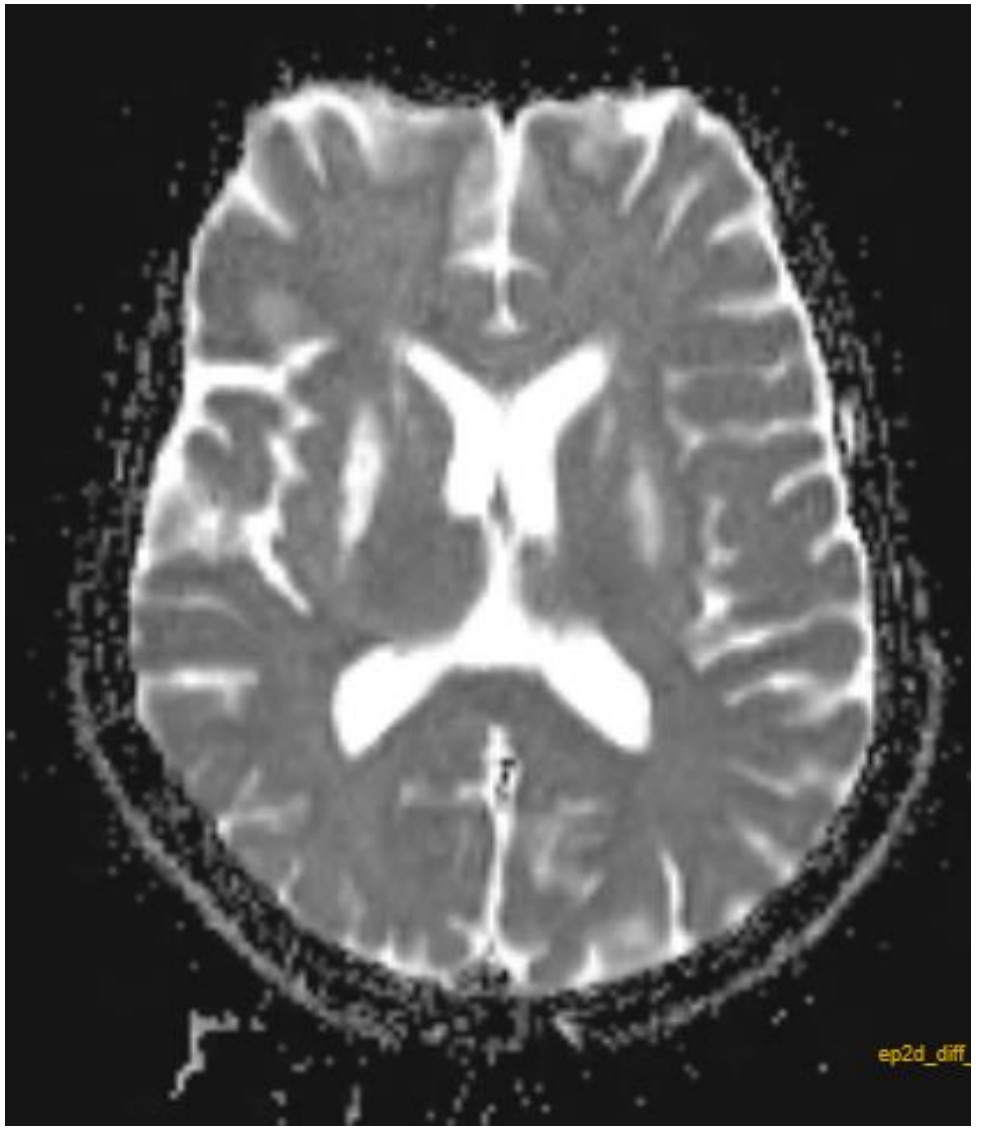
? POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME following ASV / snake bite

? ADEM - (acute disseminated encephalomyelitis) following snake bite / ASV administration

She was treated with intravenous methyl prednisolone 30mg/kg for 5 days along with tetrabenzine.

Chorea and tongue tremors gradually improved .





MRI AT FOLLOW UP

DISCUSSION

This case is unique as PRES in patients after snake bite is rare . Only few such cases have been reported.

Presentation of PRES as a movement disorder post snake bite has not yet been reported.

	Type of snake	Onset	Antivenin Received	Manifestations								Reversibility
				Coagulopathy	Renal impairment	Respiratory failure	Hypertension	Neurological symptoms				
								Seizures	Visual disturbance	DLC	Motor disorders	
Delgado and Del Brutto	*Bothropsasper*	2 days	Yes, before symptoms	Yes	No	Yes	No	Yes	No	Yes	No	Yes
Chaudhary et al.	Not identified	30 min	Yes, after symptoms	Yes	No	No	No	No	No	Yes	Yes	Partially
Varalaxmi et al.	Pit viper	2 days	No	No	Yes	No	No	No	Yes	Yes	No	Yes
Kaushik et al.	*Bungaruscaeruleus*	hours	Yes, after symptoms	Not clear	Not clear	Yes	Yes	Yes	Yes	Yes	Yes	Yes

The proposed mechanisms for PRES are 1)systemic inflammatory response leading to hypoperfusion 2) hypertension leading to cerebral autoregulatory vasoconstriction , ischemia and subsequent brain edema (vasogenic/ cytotoxic)

Moderate to severe hypertension is encountered in 70%patients with PRES . Nevertheless, a significant number of PRES patients present without hypertension including this case.

A case of obstructive hydrocephalus

Case history :

32 year old female , no previous comorbidities , presented with :

- Holocranial headache (lasting throughout the day) since 2 months
- Transient visual obscurations ,multiple episodes since 2 months
- Imbalance while walking since 2 months

No history of diplopia/ facial asymmetry/dysphagia

No history of incordination / tremors

No history of loss of consciousness /convulsions / vomiting

No history of fever /weight loss /drug intake

Examination findings :

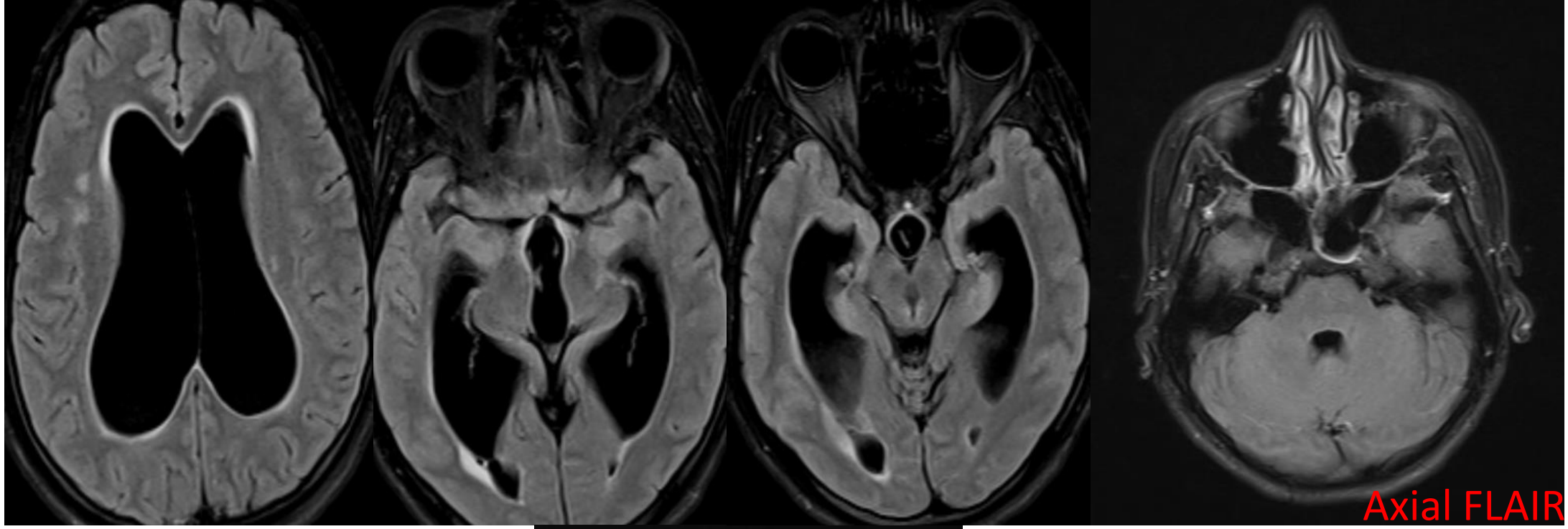
- Normal BMI
- Visual acuity – Normal
- Normal pupils
- Bilateral papilledema
- Perimetry -constriction of bilateral visual fields
- Ataxic while walking
- No finger nose ataxia

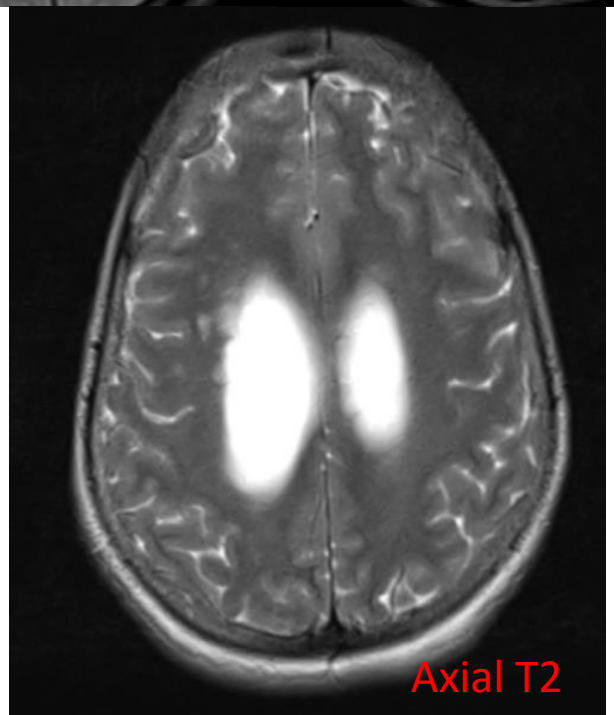
Differential diagnosis :

Gait ataxia with evidence of raised ICT :

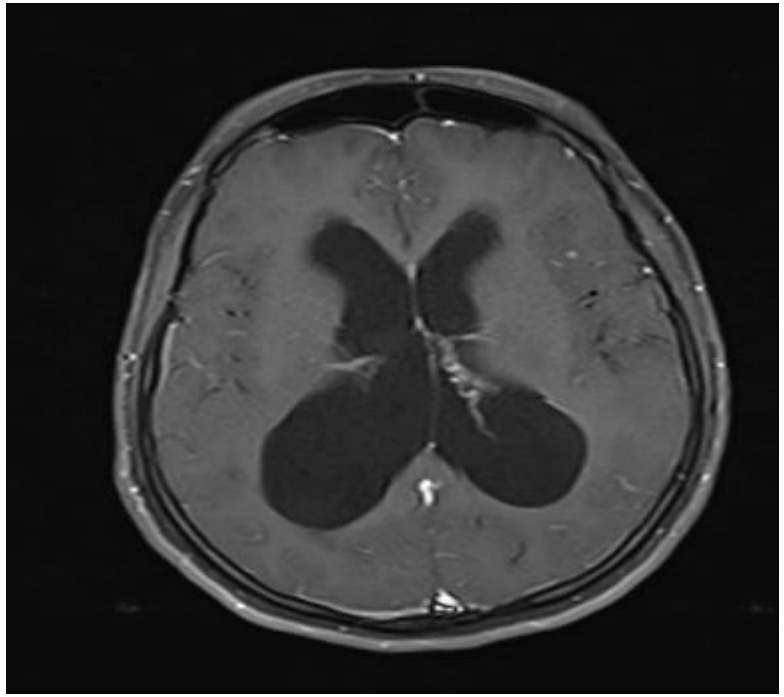
-Hydrocephalus

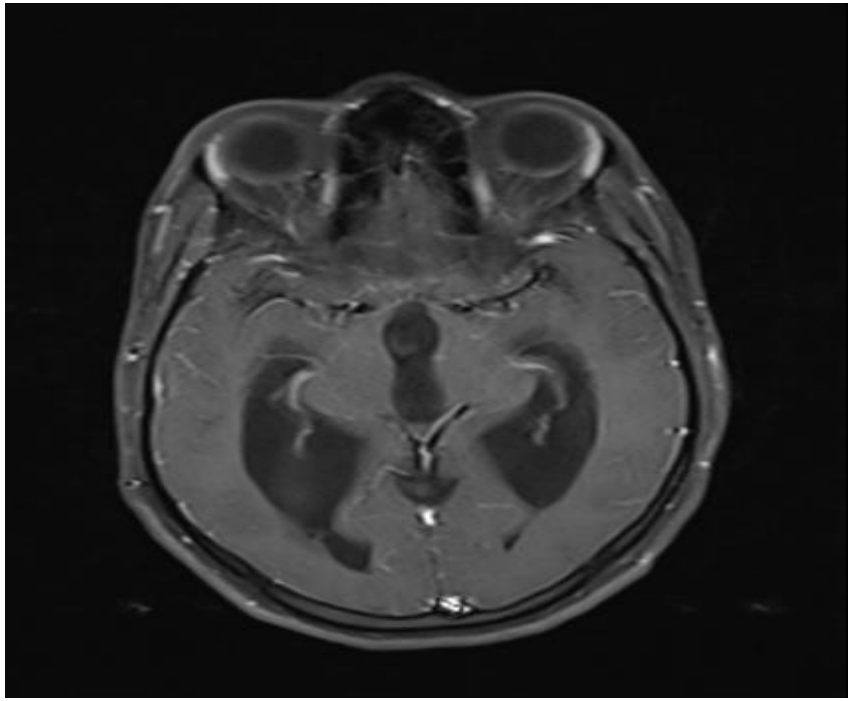
-Space occupying lesion

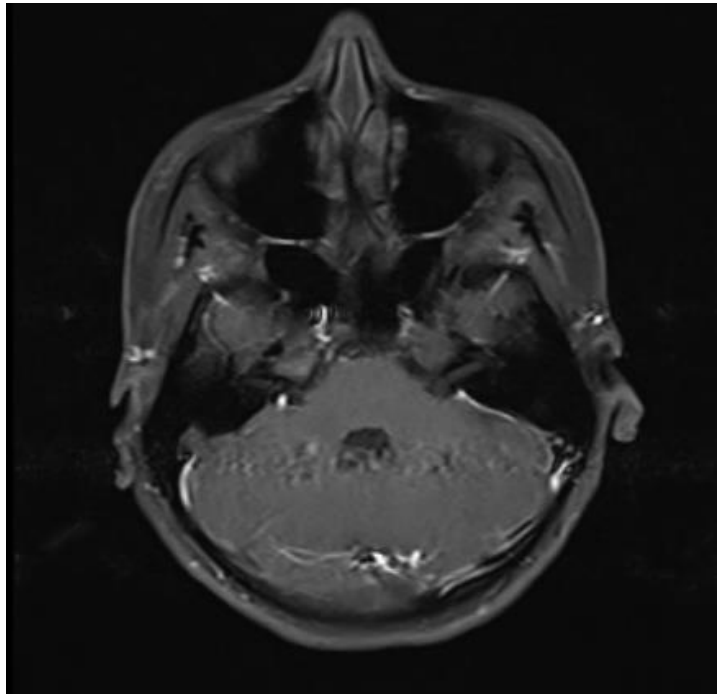

Axial FLAIR


Axial T2

Post contrast axial -T1 :







Investigations :

- ESR- 45
- HIV - Negative
- ANA-IFA – negative
- CSF –R/M : proteins – 50 mg / dl , glucose – 78 (corresponding 112), no cells

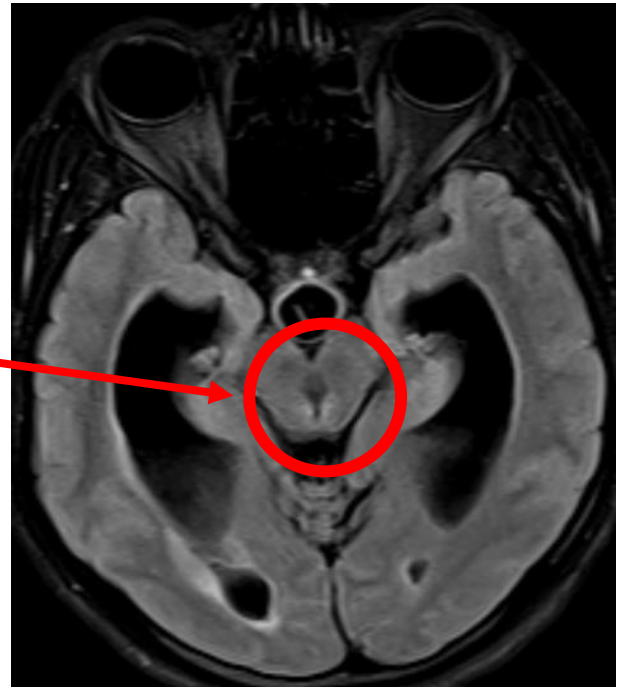
  CB-NAAT – negative

bacterial and fungal cultures – negative

- Chest x ray PA view , HRCT thorax –normal , Mantoux test – negative

? Etiology of hydrocephalus

FLAIR hyperintensities in periaqueductal region



AQP4 antibodies – positive

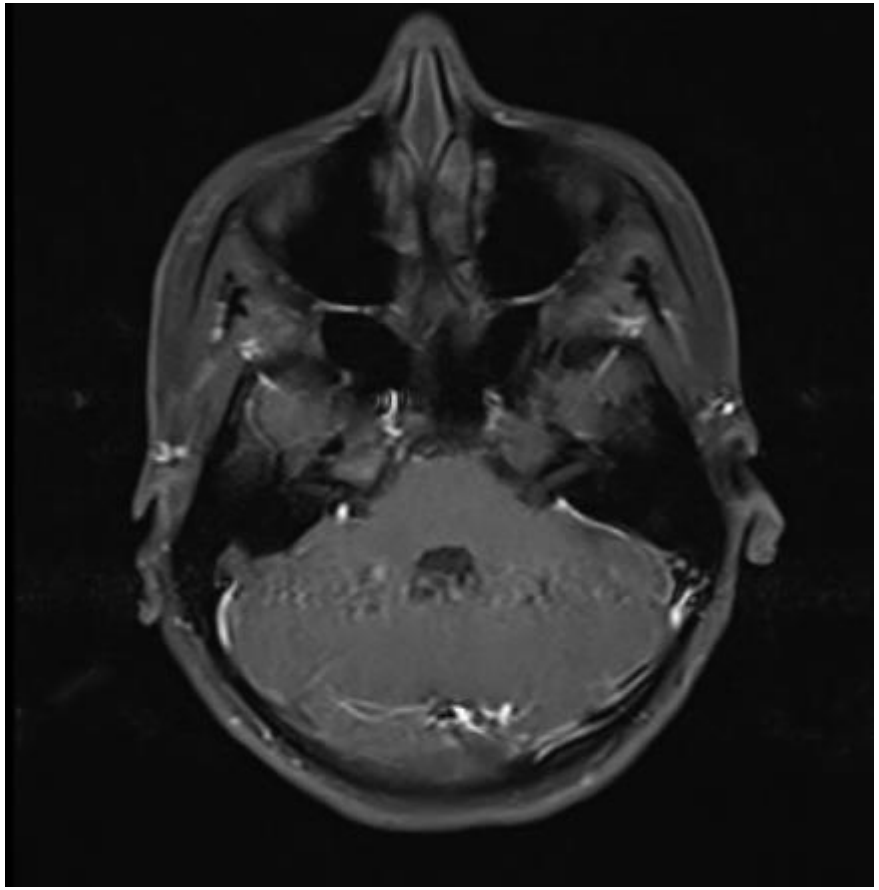
Diagnosis :

- NMO with hydrocephalus

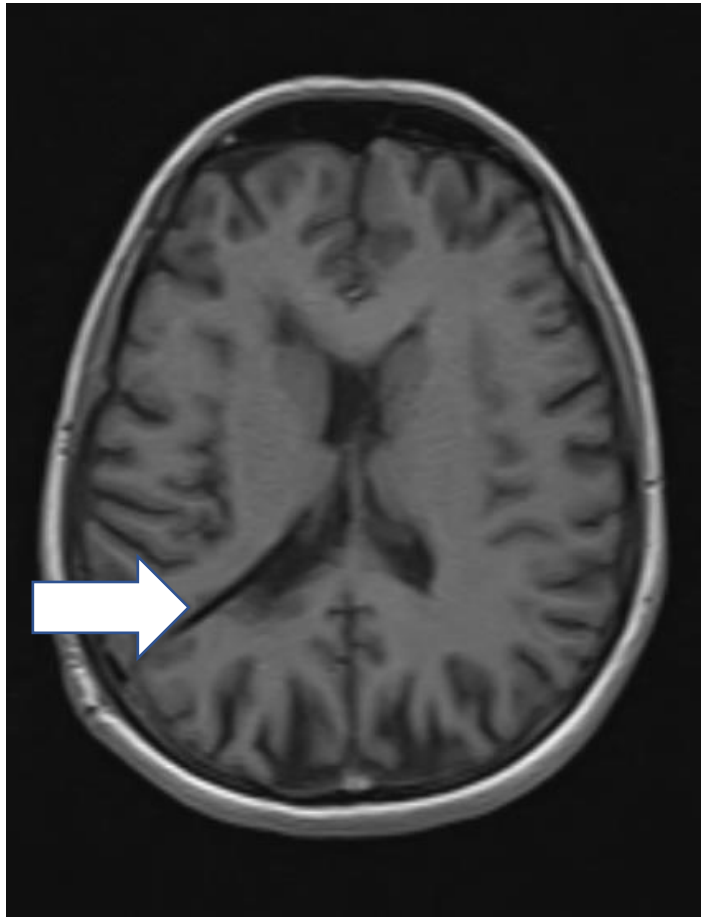
Treatment during first visit :

- IV-MPS for 5 days followed by Azathioprine and oral steroids
- VP shunting for hydrocephalus
- Gait imbalance and headache improved

Post VP shunting :



Axial post contrast
T1



Axial T1 :

8 months later…

- Acute DOV both eyes
- RE– Only PL+, Grade-3 RAPD
- LE -3 ft FC, Grade-2 RAPD
- VEP B/L prolonged P100 (R> L)
- IV-MPS for 5 days →No improvement → plasmapheresis and rituximab
- Significant improvement in vision and no relapse since 1 year

NMO and hydrocephalus :

- Cerebral aqueduct, an anatomic bottleneck of the ventricular system, is lined by ependymal cells that express AQP4
- Inflammatory sequelae of IgG binding to AQP4 in this region causes scarring, occlusion, stenosis, or reduced compliance of the aqueductal channel leading to obstruction [1]

- NMO, altered cerebrospinal fluid resorption could lead to acute hydrocephalus by a non-obstructive mechanism[2]

Take home message :

- NMO can present as hydrocephalus secondary to periventricular inflammation and ventricular scarring

References :

1. Hydrocephalus in neuromyelitis optica. Neurology. 2014 May Clardy SL, Lucchinetti CF, Krecke KN, Lennon VA, O'Toole O, Weinshenker BG, Boyd CD, Krieger S, McGraw C, Guo Y, Pittock SJ. 20;82(20):1841-3. doi: 10.1212/WNL.0000000000000428. Epub 2014 Apr 23. PMID: 24759842; PMCID: PMC4035709
2. Acute Hydrocephalus Resulting from Neuromyelitis Optica: A Case Report and Review of the Literature Liesl N. Close1 , Mario Zanaty, Patricia Kirby , Brian J. Dlouhy; WORLD NEUROSURGERY, https://doi.org/10.1016/j.wneu.2019.05.177

Review > World Neurosurg. 2019 Sep;129:367-371. doi: 10.1016/j.wneu.2019.05.177.
Epub 2019 Jun 11.

Acute Hydrocephalus Resulting from Neuromyelitis Optica: A Case Report and Review of the Literature

Liesl N Close [1], Mario Zanaty [1], Patricia Kirby [2], Brian J Dlouhy [3]

Affiliations

Unusual case of headache

50-year,male

Presented with :-

Right hemi-cranial headache

Right ear tinnitus

Double vision on right horizontal gaze

} 15 days

Transient visual obscurations +

No History : –

Decrease vision, seizure, LOC, fever,

altered sensorium, ear discharge or pain,

weakness or sensory loss.

Examination

Vitals - Normal

Fundus examination – B/L early papilledema

Right eye 6th cranial nerve paresis.

Rest nervous system examination within normal limit

Summary

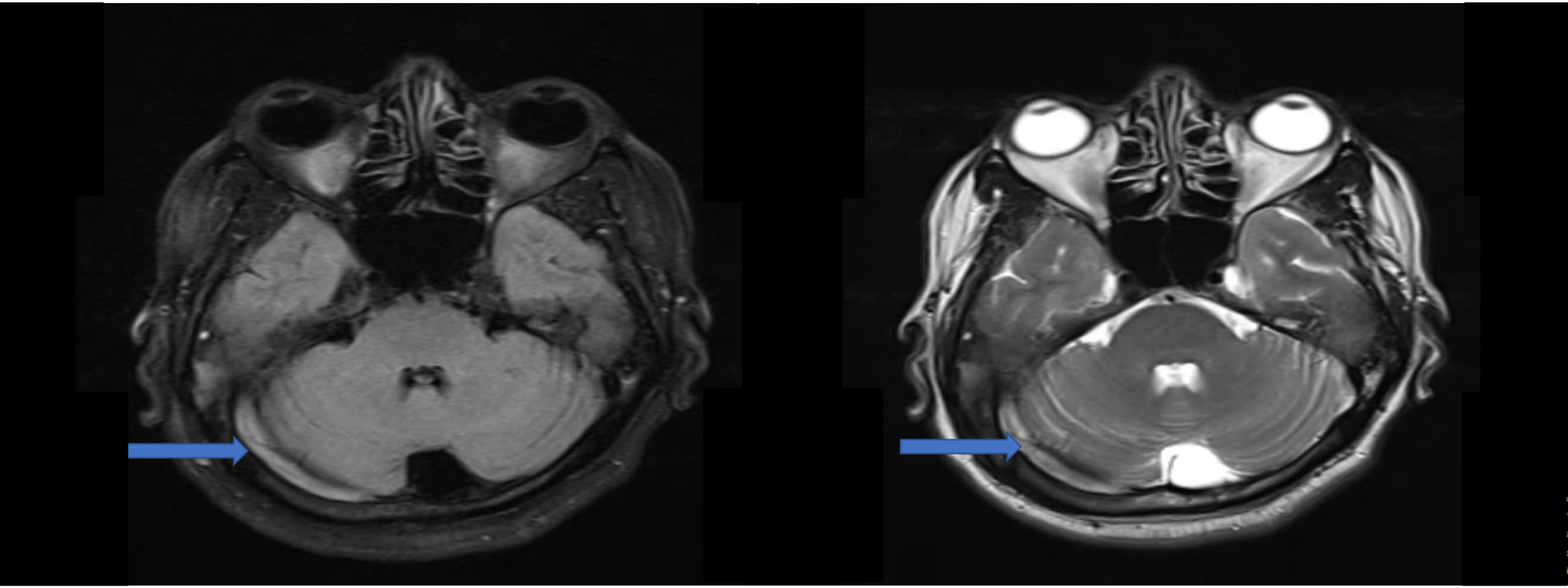
50-year male presented with right hemi-cranial headache, tinnitus, bilateral early papilledema, Right eye 6th nerve paresis.

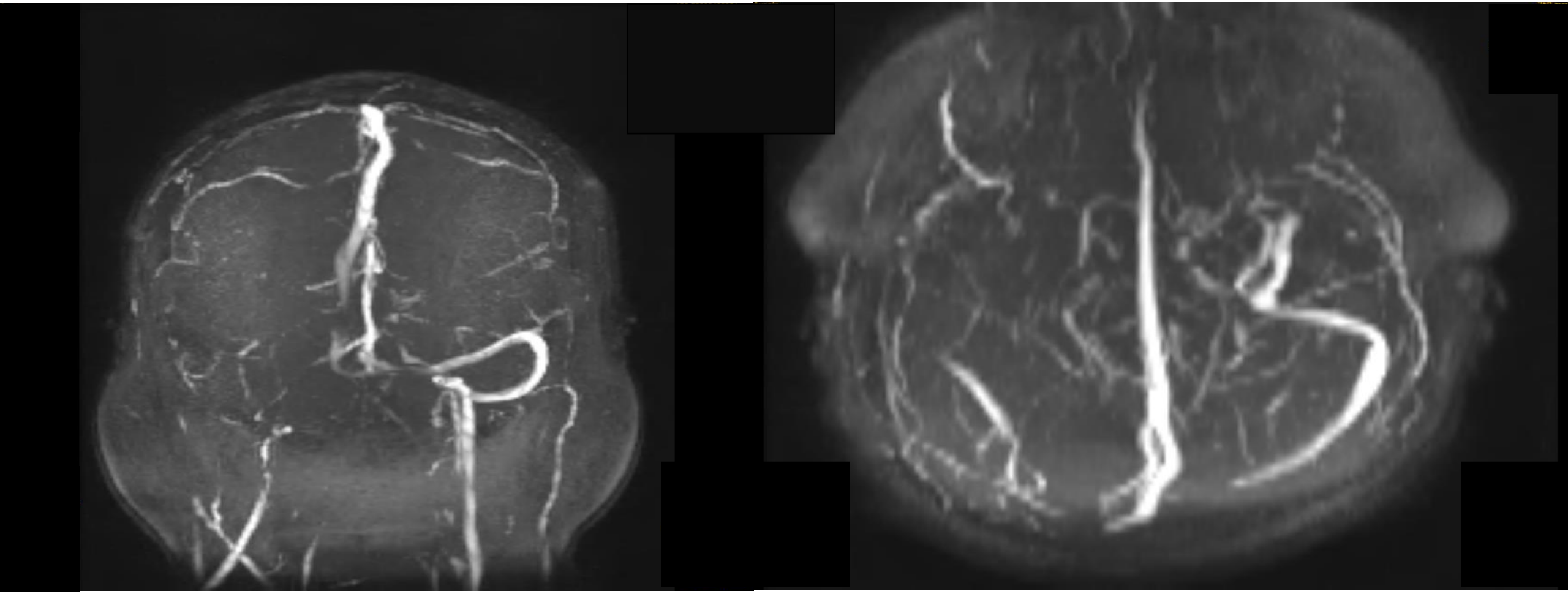
Differential diagnosis:-

Cerebral venous sinus thrombosis

Space occupying lesion .

ROTUINE LAB INVESTIGATION WERE WITHIN NORMAL LIMIT



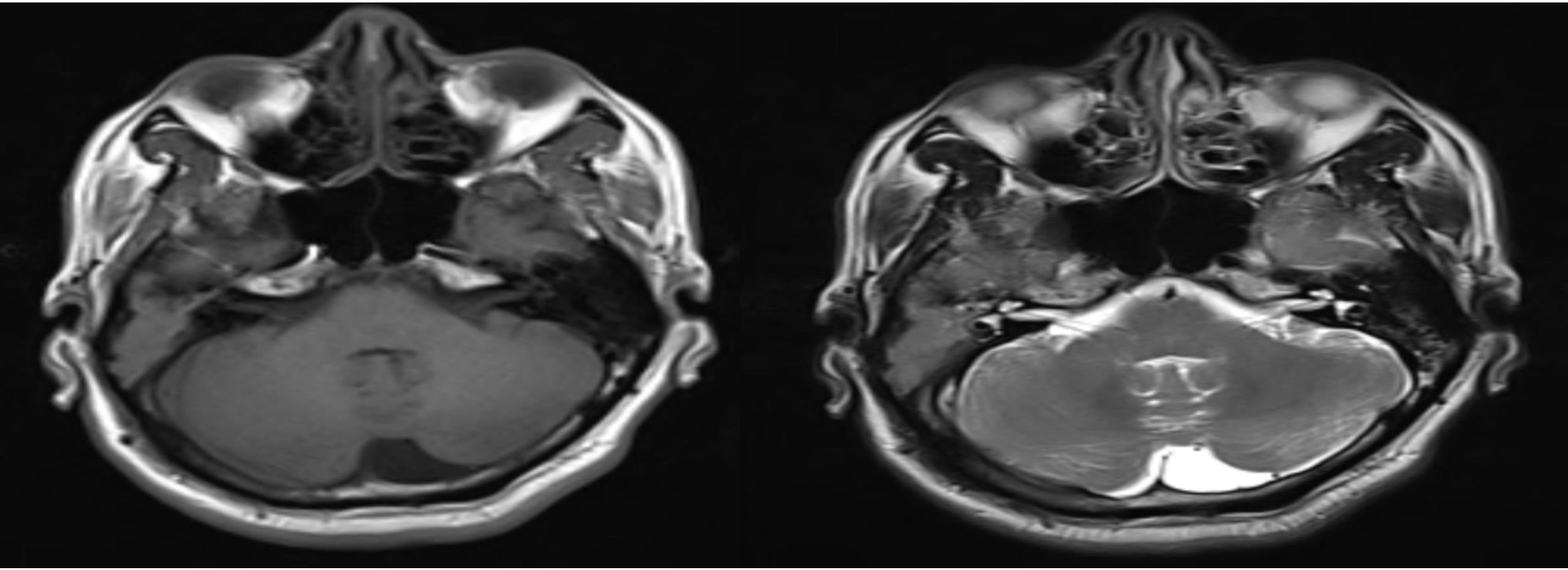


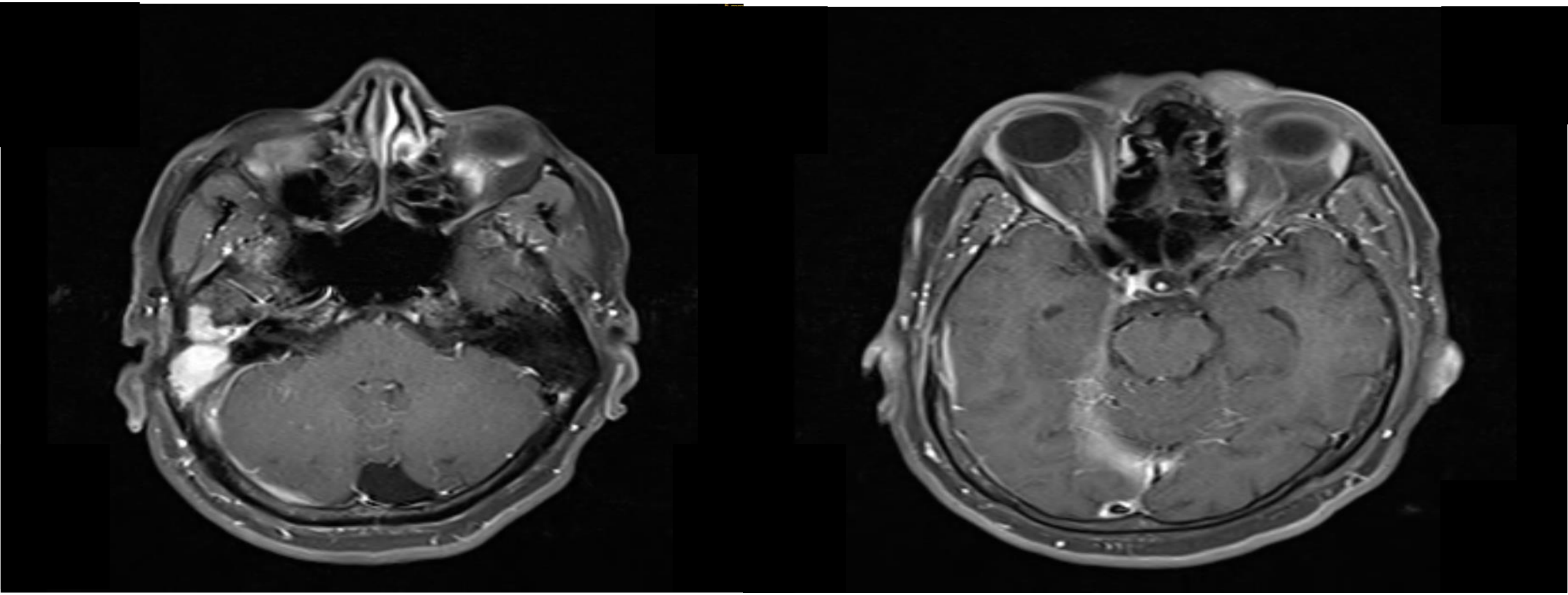
Investigations :-

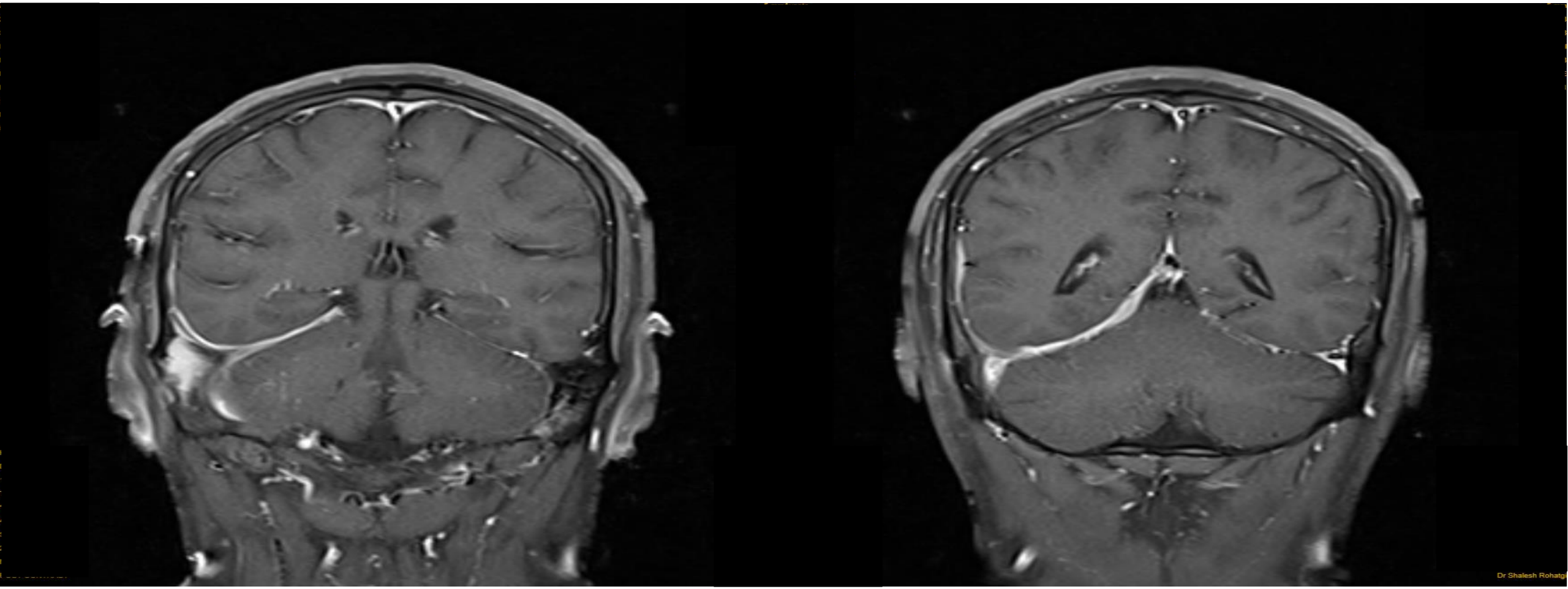
ANA IFA / APLA PROFILE – Negative

Vit B-12 level – 686 pg/ml

Sr. Homocysteine – 7.32

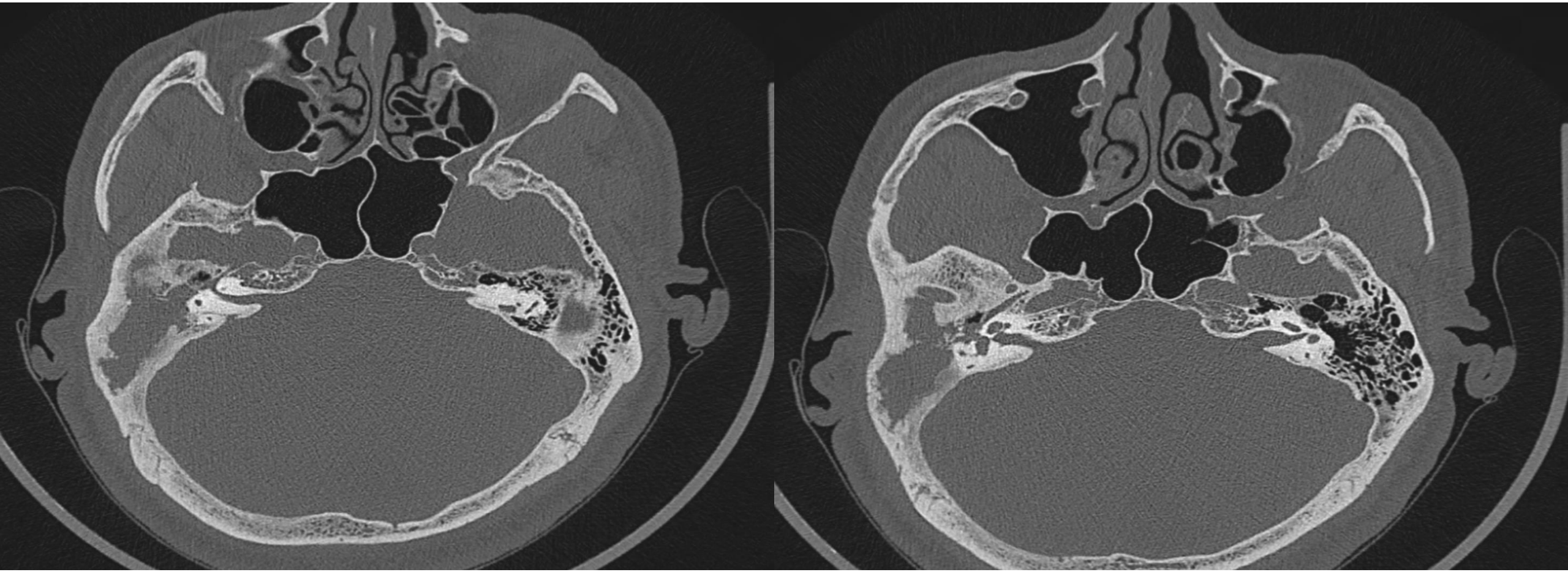





Dr Shalesh Rohatgi

CSF :-

PROTEINS - 61.1 mg/dl
GLUCOSE- 50 mg/dl (CORROSPONDING BSL -94mg/dl)
2 LYMPHOCYTE



ENT Opinion :-

Cholesteatoma

Right intact wall modified radical mastoidectomy with ossicular reconstruction with type 2 tympanoplasty

[redacted]
Pune

PID: 35009169

Age: 50.00 Year[redacted]

Reference:Dr.--
Sample Collected At
--
Dr. D. Y. Patil Medical College,
Hospital & Research Centre,
Sant Tukaram Nagar, Pimpri Colony, Pune,

SID: 322604929
322604929
Collection Date
24-01-2023 03:03 PM
Registration Date
24-01-2023 03:03 pm
Report Date
31-01-2023 04:59 PM

HISTOPATHOLOGY REPORT	
HPE no. :	B/358/23
Clinical details :	
Nature of specimen	**HPE of soft tissue from right middle ear mastoid.**
Gross Examination :	Received multiple (03) grey white, soft to firm tissue bits and pieces ranging in size from 0.4 to 0.7 cm and aggregating to 2 x 1.7 x 0.3 cm. 01 - All.
Microscopy :	[redacted] Section studied shows fibrocollagenous tissue consisting of dense inflammation comprising of lymphocytes, plasma cells, foamy macrophages and few neutrophils. Areas of hemorrhage noted. Tiny bony spicules also seen. There is no evidence of fungal elements, granuloma or malignancy. GMS- No fungal elements seen. PAS- No fungal elements seen.
Diagnosis :-	**Chronic Inflammatory pathology.**

Note :-

If Specimen preserved, it will be retained in the laboratory for 12 weeks from the date of receipt.

IHC & Special stains are not included in the NABL scope.

HISTOPATHOLOGY REPORT

Sections studied show multiple fragments of fibro-collagenous tissue with dense infiltrate composed Predominantly of plasma cells, admixed with a few lymphocytes. There is diffuse and dense fibrosis with a

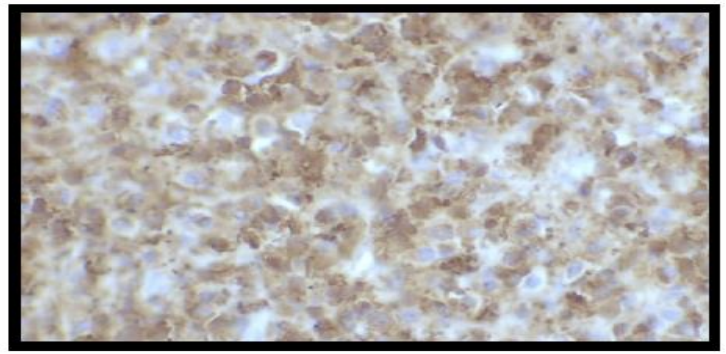
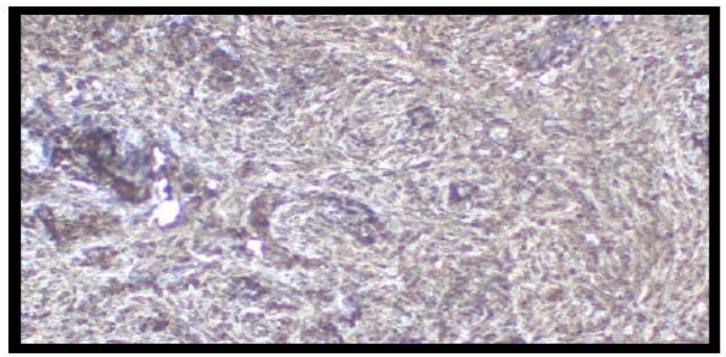
'storiform' pattern.

Occasional vessels show obliteration of the lumina .

Few bony bits and scanty necrosis is seen.

There are no granulomas or fungal elements.

There is no evidence of malignancy.

MARKERS (CLONES)	RESULT	IMAGES
IGG4	DIFFUSELY POSITIVE (REPEATED TWICE)	
IgG	DIFFUSELY POSITIVE (REPEATED TWICE)	

Sr.IgG4 :– 2.35g/L (0.03-2.0)

Final diagnosis :-

IgG-4 Disease

(Cerebral venous sinus thrombosis with pachymeningitis and Right sided mastoiditis)

Treatment :- **Tab. Prednisolone 1mg/kg**

+

Anti-coagulation

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Short Communication

IgG4-related pachymeningitis and mastoiditis, associated with cerebral venous thrombosis: A case report

Christelle Nilles [a], Guillaume Poillon [b], Lydia Deschamps [c], Mary Daval [d], Ibrahima Mariam Diallo [e], Romain Deschamps [a], Marine Boudot de la Motte [a]

Annals of Indian Academy of Neurology
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Ann Indian Acad Neurol. 2021 May-Jun; 24(3): 432–436.
Published online 2021 Jan 8. doi: 10.4103/aian.AIAN_411_20
PMCID: PMC8370182
PMID: 34447016

Cerebral Venous Sinus Thrombosis and Pachymeningitis in IgG4 Related Disease: Report of Two Cases and Review of Literature

Varun K. Singh, Jayantee Kalita,[1] Usha K. Misra,[1] and Sunil Kumar[2]

Discussion

Immunoglobulin disease (IgG4-D) is an immune-mediated fibroinflammatory condition that is capable of affecting multiple organs.

Common forms of presentation include:-

- Autoimmune pancreatitis (AIP),
- IgG4-related sclerosing cholangitis,
- Orbital disease, often with proptosis,
- Retroperitoneal fibrosis, frequently with chronic peri-aortitis and often affecting the ureters.
- Major salivary gland enlargement or sclerosing sialadenitis.

Discussion

- CNS involvement is very rare.
- IgG4-D in the nervous system most commonly manifests as hypertrophic pachymeningitis
- IgG4-D can mimic typical symptoms of other diseases

Discussion

- The hallmarks of IgG4-related disease (IgG4-RD) are dense lymphoplasmacytic infiltrations with a predominance of IgG4-positive plasma cells in the affected tissue, usually accompanied by some degree of fibrosis and often by obliterative phlebitis and an increased number of eosinophils.

- Glucocorticoids are the first-line agent for remission induction in all patients with active, untreated IgG4-RD

- Other :- Rituximab, azathioprine or mycophenolate.

Take Home message

- ✓Presence of CVST + Pachymeningitis → raised suspicion of IgG4 disease.
- ✓IgG4 presenting as mastoiditis and CVST is a mimicker of sepsis.

References :-

- *Sireesha Y, Uppin MS, Ganti S, et al. A Series of Biopsy-proven Patients with Immunoglobulin G4-related Neurological Disease. Ann Indian Acad Neurol. 2019;22(1):73-78*

- *Khosroshahi A, Wallace ZS, Crowe JL, et al. International Consensus Guidance Statement on the Management and Treatment of IgG4-Related Disease. Arthritis. Rheumatol 2015; 67:1688.*

- *Kamisawa T, Zen Y, Pillai S, Stone JH. IgG4-related disease. Lancet 2015; 385:1460.*

Thank You