SJOGRENS SYNDROME : A TRUE CHAMELEON

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SENIOR RESIDENT
DEPARTMENT OF NEUROLOGY

- 40/F (housewife) admitted with
- Imbalance while walking (sways to either side) since 2 days
- Difficulty in swallowing (nasal regurgitation)
- Double vision

She then complained of bilateral dropping of eyelids on day 3

- NO history of
- Weakness in limbs
- Tingling , numbness , pain.
- Urinary incontinence/ constipation
- Fever
- Upper respiratory tract infections
- Breathlessness, palpitations
- No Diurnal variation
- No history of toxin consumption/ medications/ addictions .

- P- 80 /min
- BP - 110/70 mmhg
- SPO2 - 98 % On Room air
- Single breath count - 38
- General examination - normal - (No bite marks)

- NEUROLOGICAL EXAMINATION
- Higher Mental Functions- Normal
- Bilateral ptosis +
- Pupils bilaterally equal reactive to light
- Fundus Examination - Normal
- Extra Ocular Movements - Complete bilateral ophthalmoplegia.
- Bilateral LMN Facial Paresis present
- GAG Reflex - Weak

- MOTOR EXAMINATION -
- Tone - Normal
- Power-

	RIGHT	LEFT
UPPER LIMBS	5/5	5/5
LOWER LIMBS	5/5	5/5

- Planter - Flexor
- Reflexes- Absent
- Sensory Examination - Pinprick normal, Impaired Joint Position.
- Sensory Ataxia- Present
- No cerebellar signs
- Bowel / Bladder - Normal.

Summary

- 40/F
- Acute onset
- Ataxia
- Opthalmoplegia and bilateral ptosis
- Dysphagia
- Bilateral Facial Paresis
- Areflexia

- ? MILLER FISHER SYNDROME
- ?Pharyngeal cervical brachial variant of GBS
- ?? Myasthenia Gravis

- LABORATORY INVESTIGATIONS
- Haemogram, electrolytes , liver function test , Renal function test, vit B12 , CPK total , seromarkers was normal.
- Csf (done on day 7)

Proteins	51
Cells	2 (100%L)
Glucose	87(Bsl 110)

- NERVE CONDUCTION STUDY - NORMAL
- MRI BRAIN AND WHOLE SPINE SCREENING WAS NORMAL

	Observed Value	Reference Range	Disease association
GM1	Negative,1	Negative	Multifocal motor neuropathy Guillian Barre syndrome
GM2	Negative,0	Negative	Multifocal motor neuropathy, Guillian Barre syndrome & variants
GM3	Negative,1	Negative	Multifocal motor neuropathy
GD1a	Negative,4	Negative	Guillian Barre syndrome & variants
GD1b	Negative,0	Negative	Sensory neuropathy
GT1b	Negative,0	Negative	Guillian Barre syndrome & variants
GQ1b	Negative,0	Negative	Miller Fisher syndrome

- Started on IVIG 2g/kg over 5 days
- However patient did not have any improvement even after 2 weeks.

- ANA by IF - 1+ , Speckled

ANA Blot test

The assay detects ANA of IgG class against 17 different antigens, each of which has been shown to be associated with specific autoimmune disorders as listed below -

Sample : Serum

Test Description	Observed value	Biological Reference Interval	Disease association
Antigens			
nRNP/Sm	**Negative**	Negative	1. Systemic lupus erythematosus (SLE) 2. MCTD (Sharp syndrome) 3. Systemic Sclerosis 4. Polydermatomyositis.
Sm	**Negative**	Negative	Systemic Lupus Erythematosus (SLE)
SS-A	**Negative**	Negative	1. Sjogren syndrome(40-80%) 2. SLE 3. Neonatal lupus erythematosus
Ro-52	**Positive**	Negative	Sjogren syndrome
SS-B	**Negative**	Negative	1. Sjogren syndrome(40-80%) 2. SLE 3. Neonatal lupus erythematosus
Scl-70	**Negative**	Negative	Progressive systemic sclerosis - diffuse & limited
PM-Scl	**Negative**	Negative	1. Systemic sclerosis including overlap syndrome 2. Polymyositis, dermatomyositis 3. Progressive Systemic sclerosis
Jo-1	**Negative**	Negative	1. Polymyositis(25-35%) 2. Dermatomyositis
CENP-B (Centromere Protein B)	**Negative**	Negative	1. Systemic sclerosis diffuse & limited 2. Primary billary cirrhosis
PCNA (Proliferating Cell Nuclear Ag)	**Negative**	Negative	SLE(3%)
ds DNA	**Negative**	Negative	SLE(40-90%)
Nucleosomes	**Negative**	Negative	SLE
Histones	**Negative**	Negative	1. Drug induced lupus(95%) 2. SLE 3. Rheumatoid arithritis (15-50%)
Ribosomal P-protein	**Negative**	Negative	SLE
AMA-M2 (Mitochondrial)	**Weak Positive**	Negative	Primary biliary cirrhosis
Mi-2	**Negative**	Negative	Dermatomyositis
Ku	**Negative**	Negative	SLE/Myositis/SSC

Suggested clinical correlation and ANA by IF.

Ro- 52 POSITIVE

HISTOPATHOLOGY REPORT	
HPE no. :	B/174/24
Clinical details :	
Nature of specimen	**HPE of 4 mm Punch biopsy from lower lip mucosa.**
Gross Examination :	Received disc of skin measuring 0.4 cm in diameter. 01 - ALL
Microscopy :	Section shows stratified squamous epithelium with underlying fragment of salivary gland tissue. The salivary gland shows mild focal ductal dilatation with mild to moderate lymphocytic infiltrate in the periductal region. No evidence of basal cell hyperplasia or parenchymal fibrosis. No evidence of dysplasia / malignancy.
Diagnosis :-	**Chronic Sialadenitis favouring Sjogren's syndrome.** Advice: Kindly correlate with antibody titres to confirm the diagnosis of sjogren's syndrome.

- SCHIMERS TEST- WETTING >10 MM IN BILATERAL EYES (NORMAL)
- TEAR BREAK UP TIME - NORMAL

- Inj methylprednisolone 1gm iv 1-0-0 for 5 days followed by oral steroids.
- Patient clinically improved after steroids.
- Planned for starting immunosuppression
- Currently in improving phase.

CASE REPORT

Neuropathy similar to Miller Fisher syndrome associated with primary Sjogren's syndrome

Response to intravenous immunoglobulins

Genç, Emine; Genç, Bülent Oguz; Avunduk, Mustafa Cihat*; Kozak, Hasan Hüseyin; Ilhan, Nurhan

Author Information

Annals of Indian Academy of Neurology 9(2):p 116-118, Apr–Jun 2006. | **DOI:** 10.4103/0972-2327.25985

CASE 2

- 36/F
- Presented with
- Tingling and numbness in bilateral hands and feet since 15 days
- Progressive proximal lower limb weakness

- History of fever for 3 days (20 days back)
- No history of cranial nerves or bladder bowel involvement.

PAST HISTORY

August 2012- GBS (**paraparesis**) received IVIG took 2 months for recovery

September 2014- GBS (**quadriparesis**) had hypoxemia , required BIPAP ventilation —received IVIG —recovered in 4 months

April 2015- GBS (**paraparesis**) Received IVIG recovered in 4 months

MAY 2022 (DY PATIL) - GBS (**quadriparesis**) Received IVIG - recovered in 4 months

All episodes preceded by fever (first 3 episodes had URTI, 4th episode had UTI)

No residual weakness /sensory symptoms after each episode .

- P - 80 / MIN
- BP - 120/80 mmhg
- SPO2 - 98 % On room air
- Single breath count - 34

- CRANIAL NERVES- Normal
- MOTOR EXAMINATION
- TONE - Normal

POWER	RIGHT	LEFT
UPPER LIMBS	5/5	5/5
LOWER LIMBS	PROXIMAL4/5 DISTAL 5/5	PROXIMAL 4/5 DISTAL 5/5

- AREFLEXIA IN BILATERAL LOWER LIMBS
- PLANTERS - FLEXOR
- SENSORY - Impaired joint position and vibration sense in bilateral upper limbs and lower limbs
- NO cerebellar signs/ bowel bladder involvement

- **PREVIOUS REPORTS (MAY 2022)**
- NCS - Demyelinating sensorimotor polyneuropathy with secondary axonal degeneration involving all four limbs.
- CSF-R/M

Proteins – 45

Glucose – 78 mg/dl (corresponding BSl- 112)

Cells -2 (both lymphocytes)

Previous Nerve Biopsy Report -

NERVE BIOPSY - Date: 05/05/2022 09:47 AM

Nature Of Specimen:

Received a curved flattened nerve segment measuring 2cm in length. All procssed -A1,A2.

Grossed by Dr. Sridevi. Dt: 29/4/22

Histopathology Report:

Section from nerve biopsy shows hyperosmolar artefacts. There is mild subperineurial edema. There is no acute axonal breakdown or onion bulb formation. There is no inflammation or vasculitis. Kpal stain shows mild non uniform loss of myelinated fibres, with several thinly myelinated fibres and occasional regenerating clusters.

Final Impression:

Demyelinating neuropathy; right sural nerve biopsy

MAY 2022

ANA BY IFA - 1+

ANA – BLOT - **Ro52 positive**

SCHIMERS TEST and Tear Break up time - Normal

Advised for minor salivary gland biopsy (not willing)

- During the current admission she was treated with Inj methylprednisolone 1gm IV OD for 5 days ———> **Improved** .

- Advised for salivary gland biopsy and planned for immunomodulatory therapy .

Case Report/Olgu Sunumu

İstanbul Med J 2020; 21(Suppl 1): 9–11
DOI: 10.4274/imj.galenos.2020.52933

Rheumatic Diseases Presenting with Guillain-Barré Syndrome: Sjögren's Syndrome and Systemic Lupus Erythematosus

Guillain-Barré Sendromu ile Prezente Olan Romatolojik Hastalıklar: Sjögren Sendromu ve Sistemik Lupus Eritematozus

Ebru Aytekin[1], Hüdanur Coşkun[1], Yasemin Pekin Doğan[1], Burak Tayyip Dede[1], Özer Burnaz[1], Ufuk Emre[2]

[1]University of Health Sciences Turkey, İstanbul Training and Research Hospital, Clinic of Physical Medicine and Rehabilitation, İstanbul, Turkey
[2]University of Health Sciences Turkey, İstanbul Training and Research Hospital, Clinic of Neurology, İstanbul, Turkey

However sjogrens syndrome presenting as Recurrent GBS has not yet been reported

CASE 3

29/Female

Episodic muscle contractions and abnormal posturing in bilateral upper limbs and lower limbs

Since 5 months

Symptoms used to last for 2-3 min with 10-12 episodes per day.

DYSTONIA

No Behavioral abnormalities.

No Diurnal variation

No symptoms during sleep.

ON EXAMINATION

P – 78/min
BP – 138/70
RR – 18 /min

NEUROLOGICAL EXAMINATION

Tone was increased in all 4 limbs.
Generalized Hyperreflexia
Plantars were b/l extensor

INVESTIGATIONS

- Routine investigations were normal
- Serum ceruloplasmin , serum iron profile,ionised calcium levels, serum Magnesium , phosphorous, vitamin D, total CPK Nac ,Thyroid profile was normal
- Nerve conduction study - Normal
- MRI Brain - Normal

Ana by IFA =+1 positive.

Ana blot

Anti Ro 52 antibodies positive

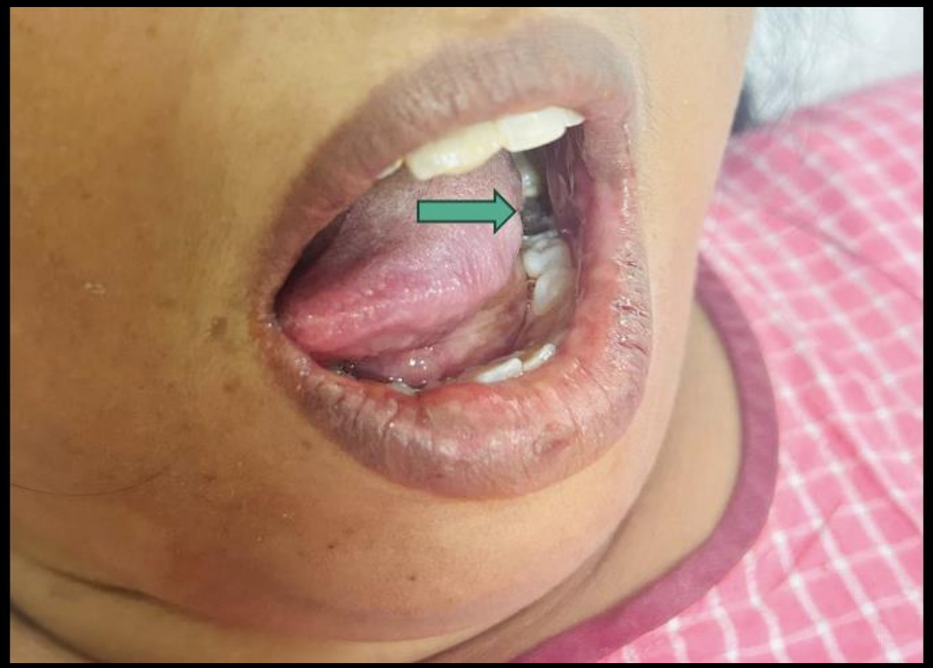
Anti SSB antibodies positive.

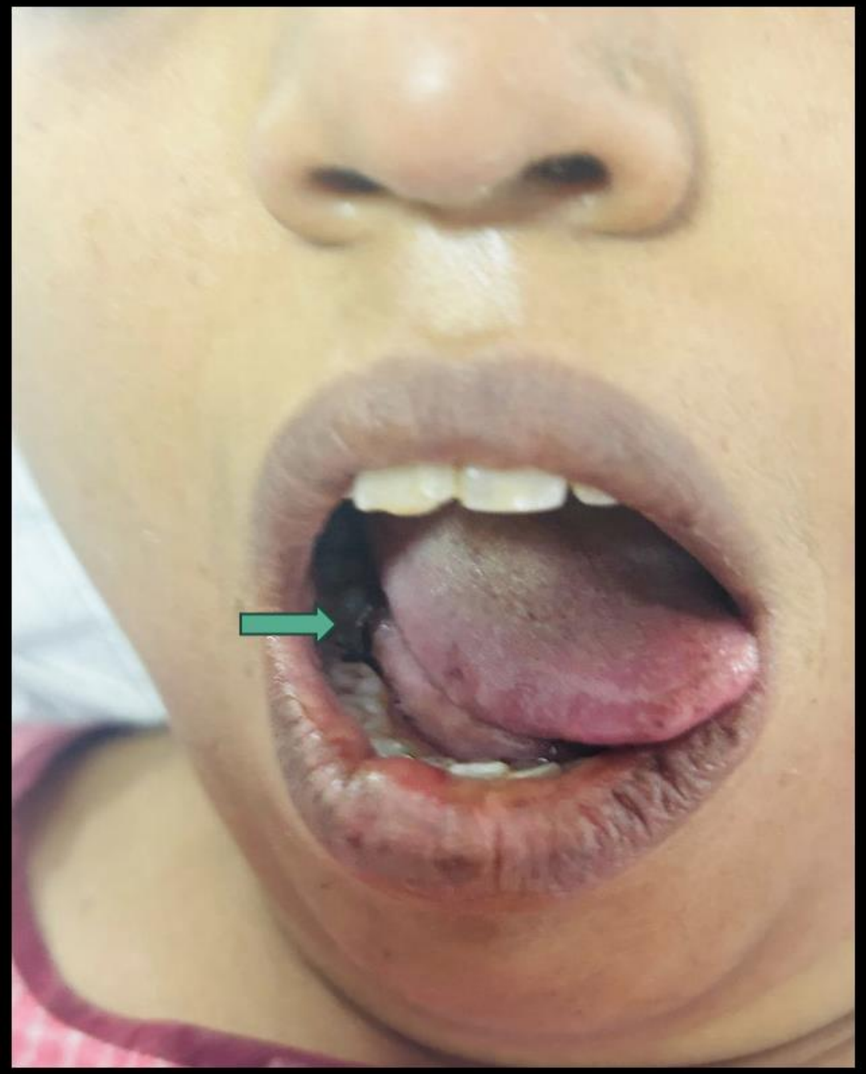
Minor salivary gland Biopsy - no evidence of inflammation.

Schimers test- wetting >10 mm in bilateral eyes (normal)

Tear break up time - normal

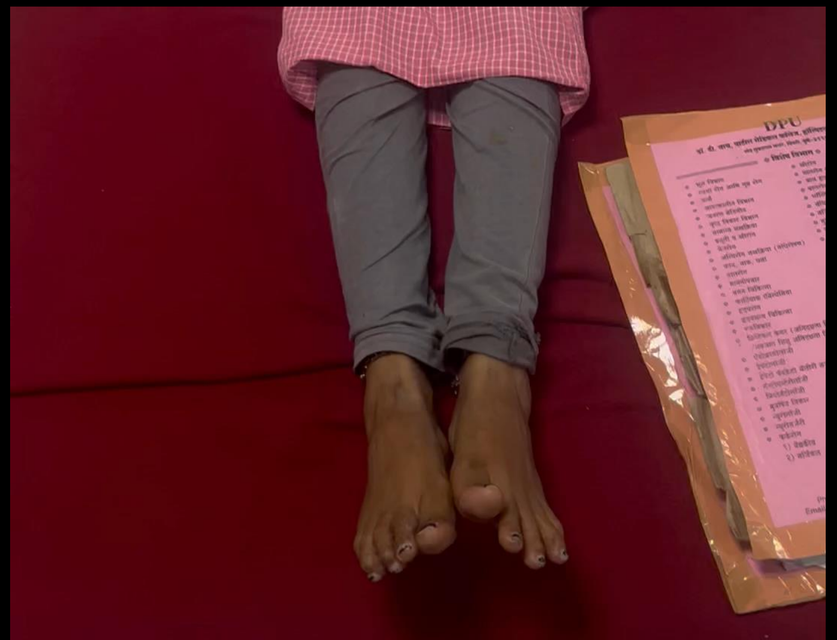
Dental caries





- TREATMENT
- Patient was treated with IV steroids and symptomatic management .

ON DISCHARGE



BMJ Journals

CASE REPORT

Case of primary Sjogren's syndrome preceded by dystonia

Kerime Ararat,[1] Idanis Berrios,[2] Anas Hannoun,[1] Carolina Ionete[3]

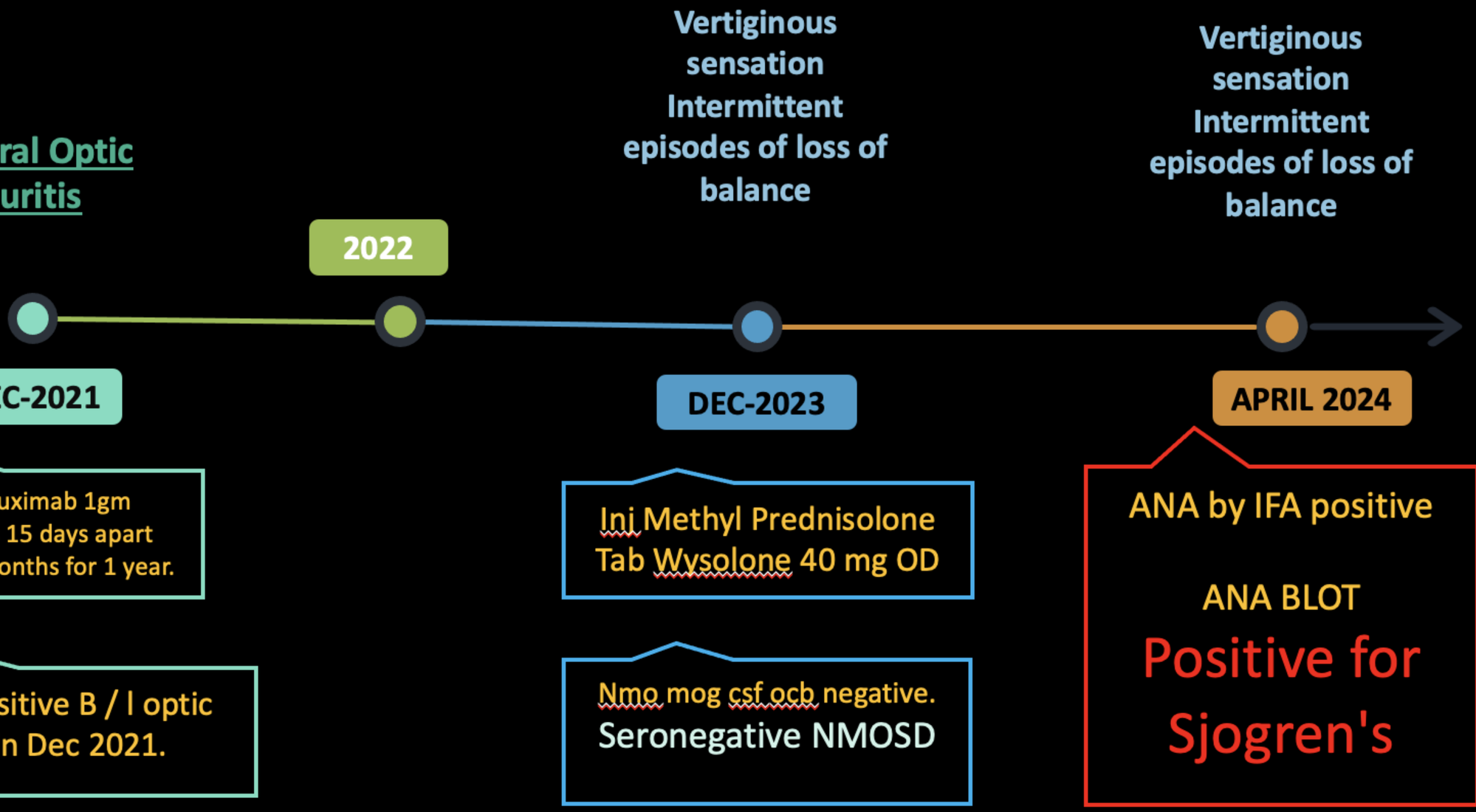
CASE 4

24 Year old female

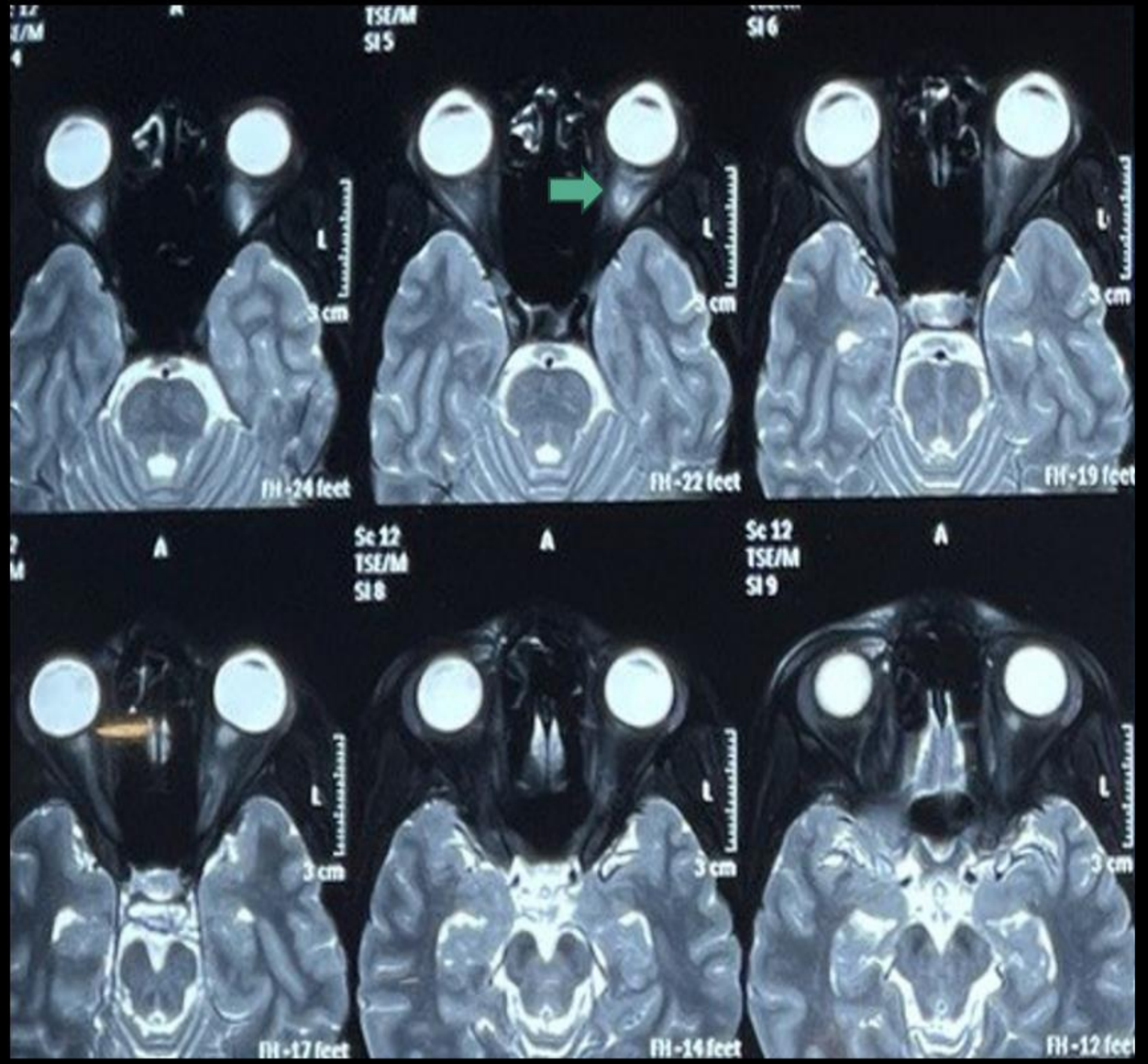
- Imbalance while walking since 3 months

- Dizziness and multiple episodes of vomiting. since 1 month

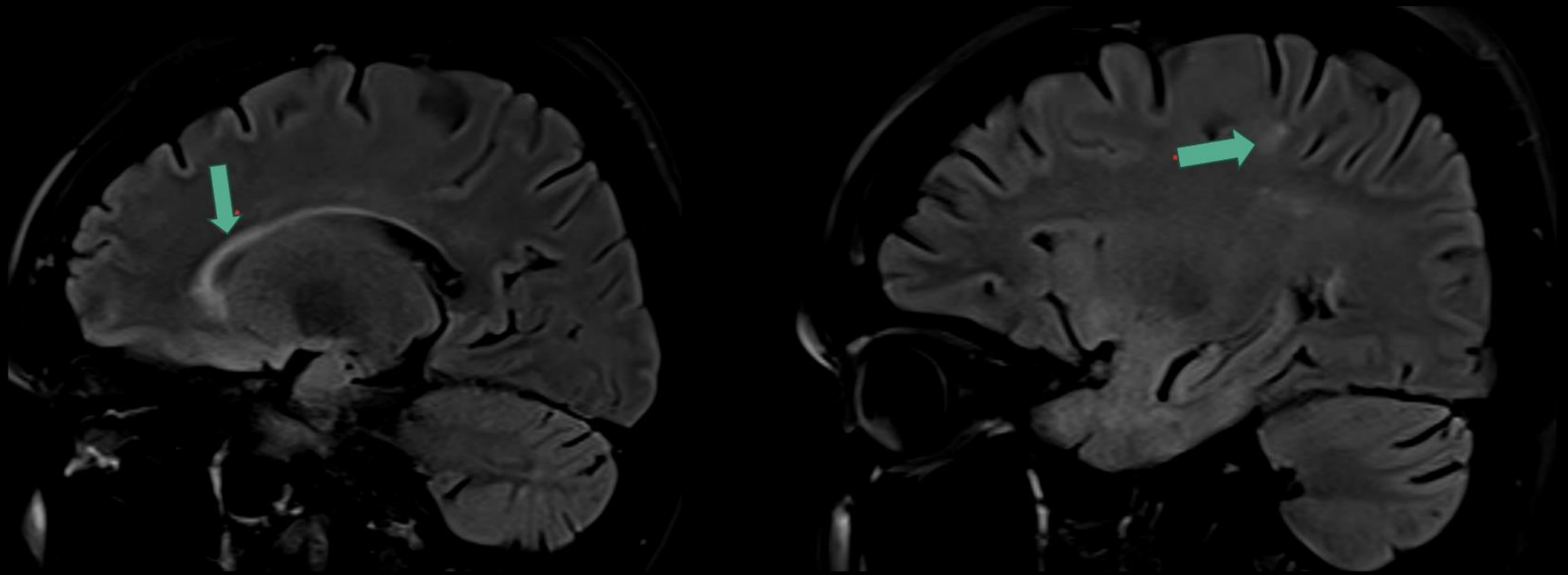
Chronology of clinical events



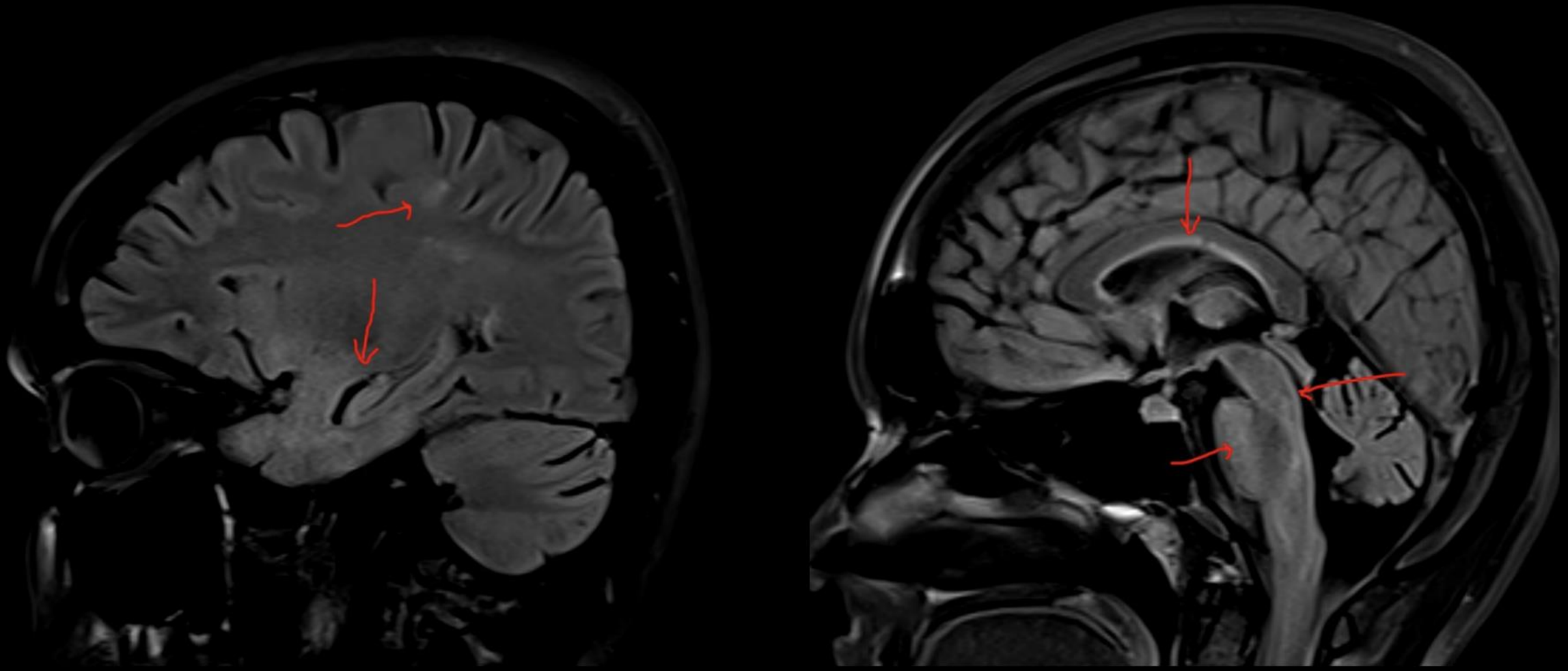
MRI Orbit - DEC-2021



MRI Brain – DEC -2023



MRI JAN 2024



Treatment given

- Inj Methyl Prednisolone 1gm in 300 ml NS over 5 days
- Plasmapheresis – 5 cycles
- Inj Rituximab 1 gm
- Tab Wysolone 40 mg OD
- Symptomatic Treatment
- Physiotherapy

CNS demyelinating events in primary Sjögren's syndrome: A single-center case series on the clinical phenotype

TAKE HOME MESSAGE

- The co occurrence of both Primary Sjogrens's Syndrome and symptoms mimicking GBS and MFS suggest that there is a clinical rationale for searching for occult Sjogrens's Syndrome in such patients.
- Neurologic manifestations may precede sicca symptoms in 33 to 93 percent of patients.
- Sicca symptoms can be so subtle while dystonia can be the only presenting symptoms in primary Sjogren's syndrome.
- Sjogren's can present as a demyelinating disease like NMOSD.

NEUROLOGICAL PRESENTATION OF SJOGRENS SYNDROME

CENTRAL NERVOUS SYSTEM

Spinal cord dysfunction
- (i) Chronic progressive myelopathy
- (ii) Lower motor neuron disease
- (iii) Neurogenic bladder
- (iv) Acute transverse myelitis

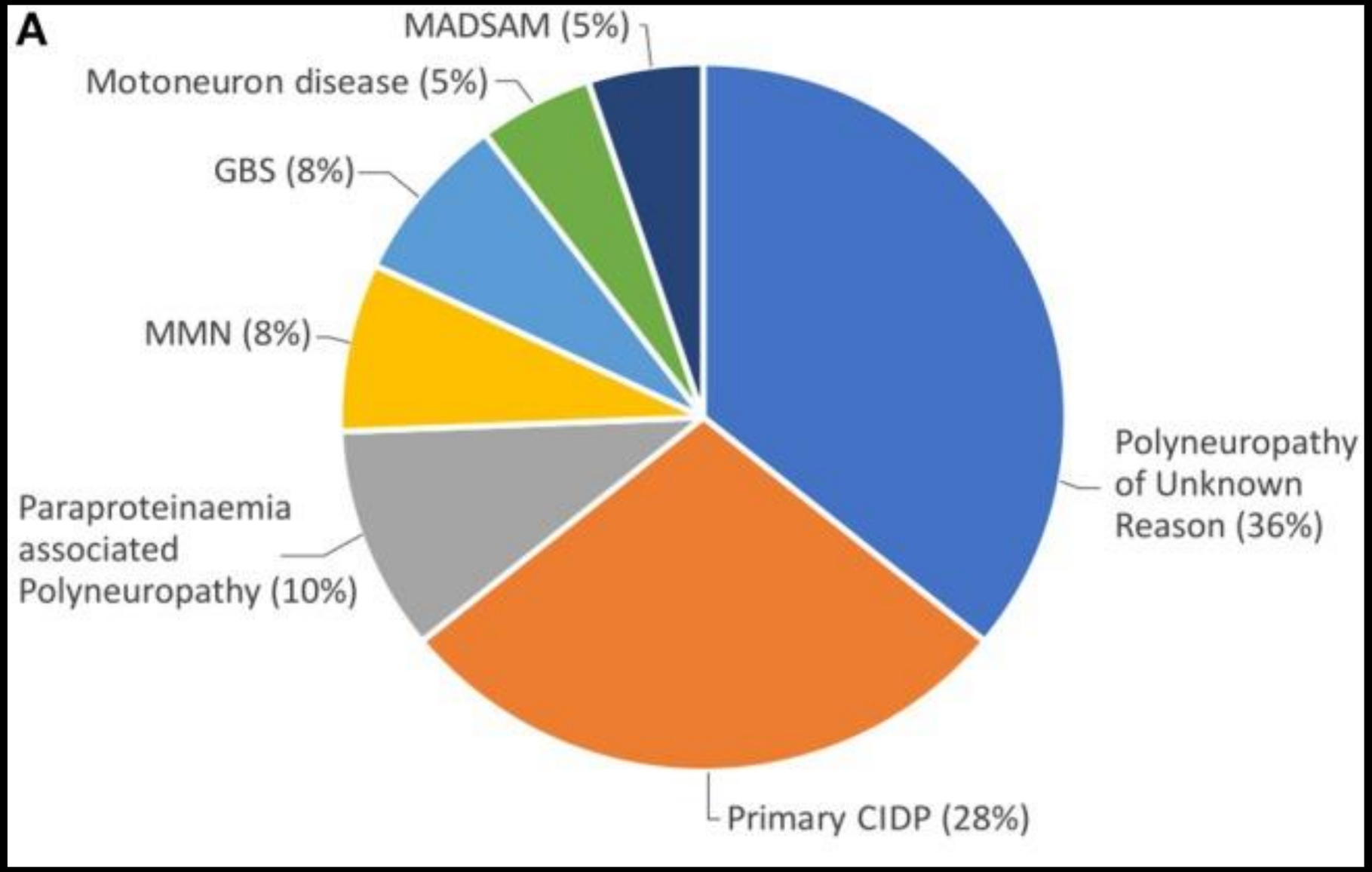
Progressive-multiple sclerosis-like syndrome

Central nervous system vasculitic involvement

Focal
- (i) Seizures
- (ii) Movement disorders
- (iii) Cerebellar syndrome
- (iv) Optic neuropathies
- (v) Pseudotumor lesions
- (vi) Motor and sensory loss

Multifocal disease
- (i) Cognitive impairment
- (ii) Encephalopathy
- (iii) Dementia
- (iv) Psychiatric abnormalities
- (v) Aseptic meningoencephalitis

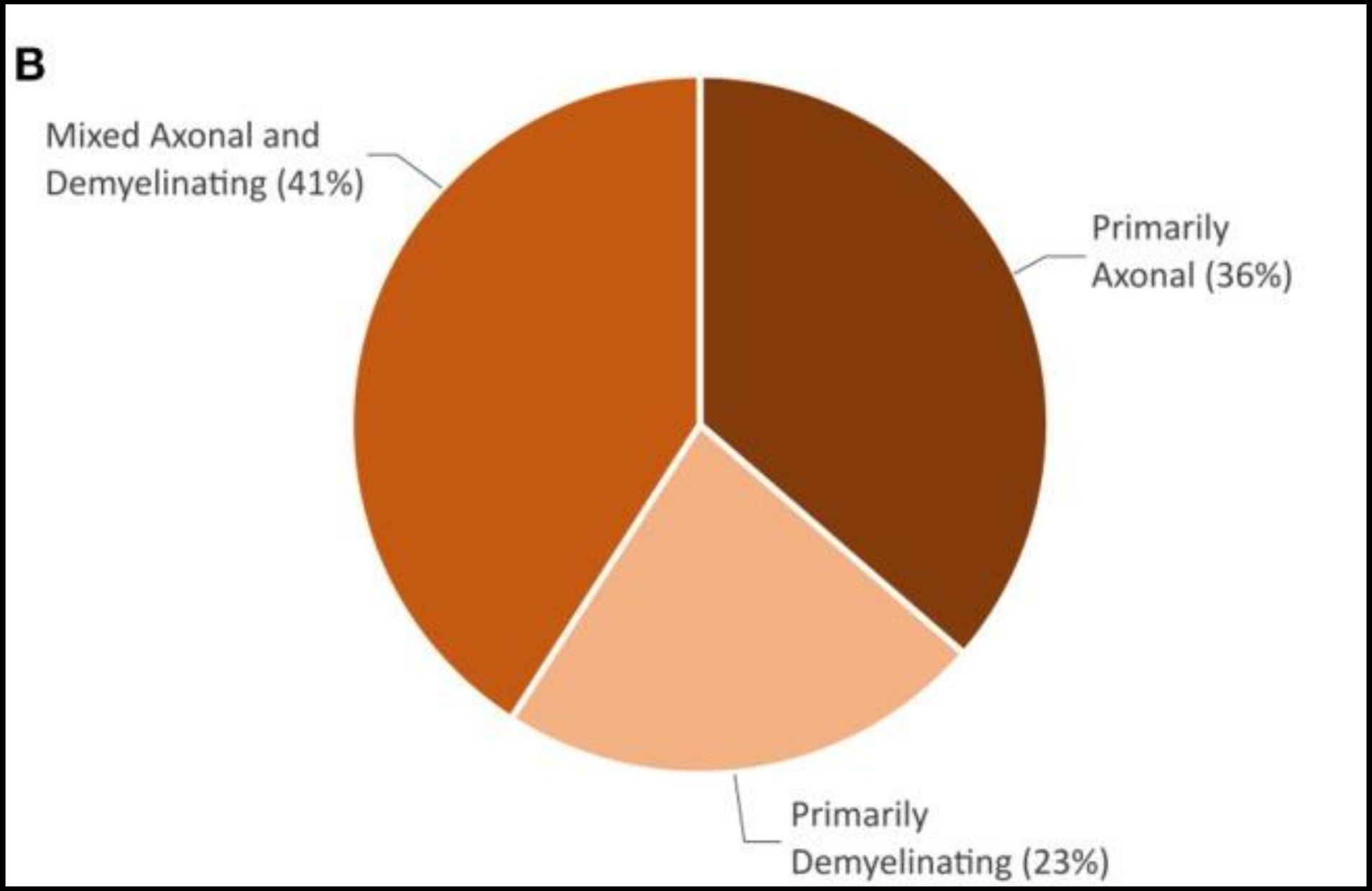
Neuropathy Diagnosis prior to sjogrens syndrome diagnosis



Small-fiber neuropathy
Multiple mononeuritis
Trigeminal and other cranial nerves neuropathies
Autonomic neuropathies
Sensory ganglioneuronopathy

REFERENCE - FRONTIERS IN IMMUNOLOGY JUL 2019

Distribution of electrophysiological damage pattern at initial neurographic analysis



REFERENCE - FRONTIERS IN IMMUNOLOGY JUL 2019

THANK YOU