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A CASE REPORT OF CHRONIC INFLAMMATORY DEMYELINATING POLYRADICULONEUROPATHY (CIDP)

General Medicine

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ABSTRACT

Case presentation of a 21-year-old guy with a likely diagnosis of Chronic Inflammatory Demyelinating Polyradiculoneuropathy is provided in this article. (CIDP).CIDP usually presents with lower limb weakness (difficulty in walking, foot drop), numbness, abnormal sensation (tingling, pain), and even loss of reflexes. CIDP does not have a definitive diagnosis, and it requires a combination of thorough history taking, clinical examination and a series of investigations (both invasive and non-invasive). The pathophysiological mechanisms of CIDP are not fully yet understood. Currently, 1st line treatment for CIDP includes- IVIG therapy and IV Steroid therapy.

KEYWORDS

INTRODUCTION:-

Peripheral nerves and their roots are particularly vulnerable to the immune-mediated neuropathy known as Chronic Inflammatory Demyelinating Polyradiculoneuropathy (CIDP). Around 10 instances per 100,000 persons are reported annually. In CIDP, men outnumber women by a ratio of around 4:1. CIDP has many variants namely- Multifocal, Focal, Motor, Sensory and Distal CIDP. Risk factors for development of CIDP are not yet known. The case presented below appears to be a mixed motor-sensory type.

Case Presentation:-

Male, age 21, arrived with symptoms of lower back discomfort dating back six months, bilateral knee pain for 6 months and lower limb weakness for 3 months. Patient was apparently asymptomatic 6 months back following which he developed lower back pain and bilateral knee pain which was insidious in onset, gradually progressive, aggravated on movement, relieved on rest and on taking medication.

Patient also had lower limb weakness which was insidious in onset and gradual in progression, not involved with any loss of sensations or abnormal sensations over lower limbs. Patient has no known co-morbidities. The patient's family history was nonexistent. The patient was alert, cooperative, and afebrile throughout the examination. There were no signs of malnutrition or dehydration, such as pale skin, icterus, clubbing, cyanosis, or lymphadenopathy.

No abnormalities were found in the lungs, stomach, or heart during the systemic examination. On examination of central nervous system- B/L Proximal lower limb power- 3/5, B/L Distal lower lower limb power- 3/5. Tenderness present over L4-L5 vertebrae. Plantar- B/L Flexor. Sensations on lower limbs are intact.

Electrocardiogram done showed a regular sinus rhythm and normal rate with no ST-T changes. MRI C-Spine with whole spine screening: The ventral thecal sac is indented by a little disc bulge at the C4-C5, C5-C6 levels, although there is no major neurological impairment ; Loss of cervical lordosis, Sacralisation of lumbar vertebra. Neurologist opinion was sought. Neurologist opined with clinical findings that it could be Chronic Inflammatory Demyelinating Polyneuropathy (CIDP). Additionally, distal acquired demyelinating symmetric neuropathy (DADS) and multifocal motor neuropathy (MFN) are also possible diagnoses. (MMN).

ESR was found to be mildly elevated (26 mm/1st hr), CPK-Total was found to be decreased (39 U/L).
Hb- 13.2, PCV- 39
RBC- 4.79, TLC- 10000, Platelet count- 2.74L
Cerebrospinal fluid showed elevated protein- 1.38 g/L (0.15-0.45 g/L) and normal glucose- 3 mmol/L (2.5-3.5 mmol/L) and normal pressure 10 mmCo2 (5-20 mmCO2), normal WBC count with normal appearance of fluid. Nerve conduction Study showed- Left median mild motor involvement, Right ulnar motor involvement, Both median sensory involvement, Right superficial peroneal involvement thereby confirming a mixed motor and sensory demyelinating neuropathy with peripheral neuropathy leading to a probable diagnosis of CIDP.

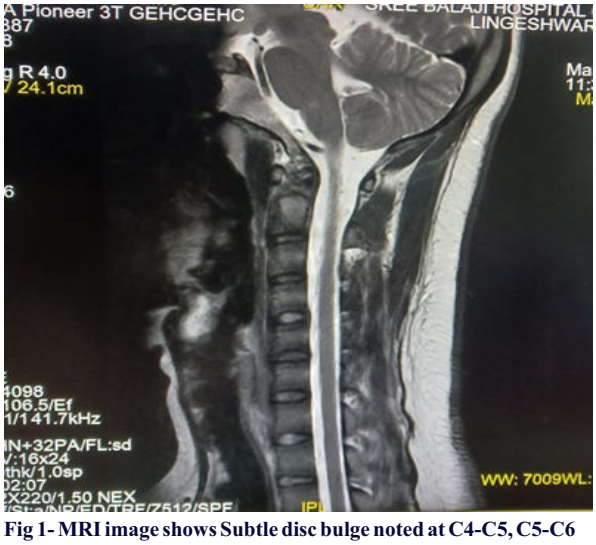

Fig 1- MRI image shows Subtle disc bulge noted at C4-C5, C5-C6

Stim Site	NR	Onset (ms)	P-T Amp (mV)	Site1	Site2	Delta-0 (ms)	Dist (cm)	Vel (m/s)
Left Median Motor (Abd Poll Brev)								
Wrist		3.4	4.1	Elbow	Wrist	4.9	24.0	49
Elbow		8.3	3.6					
Right Median Motor (Abd Poll Brev)								
Wrist		3.6	9.3	Elbow	Wrist	4.1	24.0	59
Elbow		7.7	8.5					
Left Ulnar Motor (Abd Dig Minimi)								
Wrist		2.1	7.5	B Elbow	Wrist	4.1	26.0	63
B Elbow		6.2	6.7					
Right Ulnar Motor (Abd Dig Minimi)								
Wrist		2.4	4.5	B Elbow	Wrist	5.0	26.0	52
B Elbow		7.4	4.4					
Left Peroneal Motor (Ext Dig Brev)								
Ankle		3.8	7.3	B Fib	Ankle	7.0	34.0	49
B Fib		12.8	6.7					
Right Peroneal Motor (Ext Dig Brev)								
Ankle		4.0	8.3	B Fib	Ankle	7.7	34.0	44
B Fib		11.7	7.4					
Left Tibial Motor (Abd Hall Brev)								
Ankle		3.5	8.9	Knee	Ankle	7.2	36.0	50
Knee		10.7	7.7					
Right Tibial Motor (Abd Hall Brev)								
Ankle		3.4	9.4	Knee	Ankle	7.9	36.0	46
Knee		11.3	7.6					

Anti Sensory Summary Table

Stim Site	NR	Onset (ms)	P-T Amp (μV)	Site1	Site2	Delta-0 (ms)	Dist (cm)	Vel (m/s)
Left Median Acr Palm Anti Sensory (2nd Digit)								
Palm		0.9	61.6	Wrist	Palm	0.9	7.0	78
Wrist		2.6	59.9	Wrist	Palm	2.6	7.0	41
Right Median Acr Palm Anti Sensory (2nd Digit)								
Palm		1.1	65.1	Wrist	Palm	1.0	7.0	64
Wrist		2.7	54.1	Wrist	Palm	2.7	7.0	44
Left Ulnar Anti Sensory (5th Digit)								
Wrist		2.4	87.2	Wrist	5th Digit	2.4	14.0	58
Right Ulnar Anti Sensory (5th Digit)								
Wrist		2.0	56.4	Wrist	5th Digit	2.0	14.0	70
Left Sup Fibular Anti Sensory (Ant Lat Mall)								
14 cm		2.2	11.5	14 cm	Ant Lat Mall	2.2	14.0	64
Right Sup Fibular Anti Sensory (Ant Lat Mall)								
14 cm		3.0	4.3	14 cm	Ant Lat Mall	3.0	14.0	47
Left Sural Anti Sensory (Lat Mall)								
Calf		2.1	25.8	Calf	Lat Mall	2.1	14.0	61
Right Sural Anti Sensory (Lat Mall)								
Calf		2.1	19.4	Calf	Lat Mall	2.1	14.0	67

F Wave Studies

NR	F-Lat (ms)	L-R F-Lat (ms)
Left Median (Mrkrs) (Abd Poll Brev)		
	27.50	5.00
Right Median (Mrkrs) (Abd Poll Brev)		
	22.50	5.00
Left Ulnar (Mrkrs) (Abd Dig Min)		
	23.05	5.48
Right Ulnar (Mrkrs) (Abd Dig Min)		
	28.53	5.48
Left Peroneal (Mrkrs) (EDB)		
	47.57	0.47
Right Peroneal (Mrkrs) (EDB)		
	48.75	0.47
Left Tibial (Mrkrs) (Abd Hallucis)		
	47.70	2.65
Right Tibial (Mrkrs) (Abd Hallucis)		
	44.52	2.65

H Reflex Studies

NR	H-Lat (ms)	L-R H-Lat (ms)	H-Amp (mV)
Left Tibial (Mrkrs) (Gastroc)			
	30.83	0.51	2258.13
Right Tibial (Mrkrs) (Gastroc)			
	30.31	0.51	1500.00

Fig 2- Findings of Nerve Conduction Study

Patient was initially started on oral painkillers and IV Thiamine infusion. Patient was then started on IV Immunoglobulins and IV Steroids following which the patient reported marked symptomatic improvement. IVIG therapy was completed and IV steroid therapy was gradually tapered following which he was discharged. Patient was advised to be in regular follow-up with his primary care physician for further evaluation if necessary.

DISCUSSION:-

A uncommon condition affecting the roots and peripheral nerves is known as chronic inflammatory demyelinating polyneuropathy (CIDP).[1] Clinical features of CIDP include- Weakness (difficulty in walking, foot drop), numbness, abnormal sensation (tingling, pain), and even loss of reflexes.[2] The pathophysiology of CIDP is not very well understood, Although some studies suggest that both cellular and humoral immunity can assume a part in the pathogenesis of CIDP.[3] The variants of CIDP include;- Asymmetric sensorimotor (Multifocal), Focal, Pure Motor, Pure sensory, Distal and sensory predominant, Proximal (Polyradiculopathy), Nodal and Paranodal disorders[4]. Diagnosis of CIDP requires a combination of detailed history, physical examination and series of investigations(both non-invasive and non-invasive). CSF analysis would show elevated protein in patients with CIDP.[5] CSF investigation showing an expanded CSF white cell count of >10 cells/mm3 ought to propose a considering other than CIDP, such as infection (ex- Lyme's disease), inflammatory disease (ex- sarcoidosis), or neoplasms (ex- lymphoma, leukemia). Nerve biopsy can be done which may show solid evidence of demyelination and is primarily used when more research has been insufficient to make the diagnosis of CIDP(however, the use of nerve biopsy is controversial). In a nerve biopsy, CIDP can be diagnosed based on the presence of endoneurial edema, demyelinated and remyelinated nerve filaments, macrophage-associated demyelination, onion bulb arrangement, endoneurial mononuclear cell penetration, and inhomogeneous involvement of different fascicles and different areas within the same fascicle.[6] A limitation of nerve biopsy is it's sub-par sensitivity and specificity. In order to diagnose CIDP, there is currently no gold standard. Current diagnostic criteria for CIDP are those established by the European Academy of Neurology/Peripheral Nerve Society (EAN/PNS) in 2021.[7] Some differential diagnoses include Guillain Barre Syndrome, Acute Demyelinating Polyneuropathy both of which were ruled out since symptoms were that of chronic nature and not acute nature.[8] anti-nodal and anti-paranodal autoantibodies have been discovered in around 10% of individuals with a clinical diagnosis of CIDP and are thought to be harmful.[9] Antibodies of the IgG4 subclass, the majority of which are directed against proteins at or near the node of Ranvier. Usually for cases of CIDP, IVIG therapy and Corticosteroid therapy is indicated. Other potential diagnosis include distal acquired demyelinating symmetric neuropathy and multifocal motor neuropathy (MMN). (DADS). The patient has to be evaluated further by carrying out further investigations.

CONCLUSION:-

A complicated condition of the peripheral nerve system is called CIDP. It's pathophysiology is not very well understood as of yet. Treatment options for CIDP are as of now limited to IVIG therapy[10] and IV Steroid therapy. As future studies reveal newer pathophysiological mechanisms, newer treatments will be available to treat patients with this disorder. Patients with lower limb weakness have to be evaluated at the earliest with detailed history and thorough physical examination before starting any vital treatment. For the above mentioned patient, we came to a differential diagnosis of CIDP, however the patient has to be evaluated further.

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Ethical Consent:-

The participant gave informed permission to participate in the research.

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Conflict of Interest:-

According to the writers, they have no conflicts circumstances.

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