Guillain - Barre Syndrome (Acute Motor and Sensory Axonal Neuropathy) and Acute Transverse Myelitis Overlap Syndrome - A Rare Neurological Post COVID Complication

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Abstract: Concomitant acute transverse myelitis (ATM) and Guillain - Barre syndrome (GBS) is described as GBS and ATM overlap syndrome. Its presentation varies widely, thus making the diagnosis difficult. Overlap syndrome is more commonly associated with acute motor axonal neuropathy (AMAN), a variant of GBS. However, we present a case of a young man with combined ATM and acute motor and sensory axonal neuropathy (AMSAN) subtype of GBS in a patient with post COVID - 19 infection. This combination is quite rare and only few cases are available in literature.

Keywords: acute transverse myelitis, guillain - barre syndrome, COVID - 19, overlap syndrome

1. Introduction

Guillain - Barre syndrome (GBS) is an acute immune-mediated polyradiculoneuropathy of the peripheral nervous system which comprises of several subtypes, while acute transverse myelitis (ATM) is an immune mediated disorder of the central nervous system (CNS) [1 - 2].

The occurrence of Guillain Barre syndrome (GBS) and acute transverse myelitis (ATM), either concurrently or sequentially, is defined as GBS and ATM overlap [3]. This overlap is quite rare and its diagnosis is challenging. Till now around twenty four cases of overlap syndrome has been reported [4]. These cases were mostly preceded by gastrointestinal infections such as Campylobacter jejuni while other cases were associated with Zika virus, Mycoplasma pneumoniae, Legionella pneumophila, Bartonella henselae, Influenza virus, and Paramyxovirus [5 - 10]. Most of the cases were reported in the paediatric age group and were commonly associated with acute motor axonal neuropathy, but only few cases of post covid ATM and GBS overlap have been reported so far. [11]

This was a patient of 28 years old male post covid - 19 infection presented with overlap syndrome (AMSAN and GBS)

2. Case Presentation

A 28 years young male presented with quadriaparesis and retention of urine with girdle like sensation in cervical region. History wise patient had acute onset flaccid paralysis of both lower limb for 20 days, after two days he marked weakness of both upper limb and left upper limb was weaker than right upper limb. Weakness was static and after 10 days, he developed retention of urine and girdle like sensation in lower cervical and upper thoracic region. Patient had history of moderate covid - 19 infection two months back, for which he was hospitalised and recovered.

On general examination patient was conscious well oriented to time, place and person. Pulse rate was 86 per minute, regular in rhythm. Blood pressure was 120/70 mm of mercury measured in right arm supine position with no postural hypotension. Respiratory rate was 24 per minute and abdominothoracic type. Examination of cranial nerves were normal except 7th nerve palsy (bilateral LMN type). On motor system examination bilateral power 1/5 around hip, knee and ankle joint, 3/5 in right upper limb around shoulder, elbow and wrist joint and 1/5 in left upper limb around shoulder, elbow and wrist joint. All reflexes in both upper limb and lower limb were absent and bilateral plantar was non - responsive. All modalities of sensation were lost below T1 spinal segment. Patient was shifted to ICU and observed closely for any respiratory depression and autonomic instability but patient was not having added symptoms and signs.

3. Investigation

Along with routine biochemical and haematological investigation, screening test like HIV, HBV, HCV, Anti SARS Cov - 2 (IgG and IgM) done, and Anti SARS Cov - 2 IgG was found to be positive (639.17). Other specific investigation like cerebrospinal (CSF) fluid analysis revealed Albumino - cytological dissociation, and negative
bacteriological study. Nerve conduction studies shows Axonal demyelinating sensory motor polyneuropathy, normal F wave. Other investigation including GBS serology like serum aquaporine - 4 antibody, Anti MOG antibody, ACE was negative. The Magnetic Resonance Imaging (MRI) of the cervical spine without contrast was showing long segment intramedullary lesion in the cervical cord extending from C2 to C6 suggestive of transverse myelitis and MRI of brain was within normal limit.

4. Discussion

The overlap of Guillain Barre syndrome (GBS) and acute transverse myelitis (ATM) is defined as the concurrent or sequential occurrence of GBS and ATM. This may be explained by the presence of a common epitope of myelin in the peripheral and central nervous system [3]. Overlap syndrome is generally considered to be rare and more commonly associated with acute motor axonal neuropathy (AMAN) subtype of GBS [4]. However, we present a case of 28 years old male with post COVID - 19 infection concurrent acute motor and sensory axonal neuropathy (AMSAN) and ATM. During our literature review, we identified only 1 report of similar overlap [14].

The clinical presentation of GBS and ATM overlap varies extensively. One review broadly classified the clinical presentation of overlap syndrome into three categories: patients with positive pyramidal signs and are flexia or hyporeflexia, those who suffered pain at the onset of the disease and those with respiratory compromise requiring ventilator support. Sensory loss and incontinence indicate concurrent ATM [4]. In our case, the patient had motor weakness (more in the lower limbs), hyporeflexia, sensory loss, and extreme flaccidity of lower limbs.

During the early stages of overlap syndrome, the diagnosis is challenging. Hence, most of the patients are initially diagnosed with GBS or ATM alone. Similar observation has been made by other researcher like Gou et al had marked that electrophysiological studies help with the diagnosis of GBS, while spinal cord MRIs are fundamental for the identification of ATM [4].

Currently, the first - line therapy for GBS and ATM overlap is not well defined [4]. Corticosteroid therapy and plasmapheresis are effective for ATM [12]. The first - line therapy for GBS is intravenous immunoglobulins (IVIG) or plasmapheresis while corticosteroids alone have not proven to be beneficial in the management of GBS and may worsen the weakness [5]. If patients of ATM do not show improvement with initial corticosteroid therapy, the next treatment options are plasmapheresis or IVIG [13]. Combination therapy of corticosteroids and IVIG has not proven to have a beneficial outcome; a review study revealed that less than 50% of patients benefited from the combined therapy [4]. In our patient also treated with IVIG and corticosteroid therapy combinedly and patient shows improvement in motor power, more in upper limb in comparison to lower limb.

5. Conclusions

Overlap of ATM and GBS is rare complication in post COVID - 19 infection and can often be missed due to varied clinical presentations. Hence, it should be considered in patients who have an acute progressive weakness with combined upper and lower motor neuron signs on examination. However, more studies are required to identify the exactetiology and first - line therapy for effective management of overlap syndrome.

6. Conflict of interest

None

References


[14] Inam, s. H. A; Ayesha, Malik, Manahil, Chaudhry, Mohommad, Abuddla, zimar, bismah, RiazAnnals of International Medical and Dental Research; 7 (1), 2021. Article in English |GIM| ID: covidwho – 1116203

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**MRI REPORT – CERVICAL SPINE**

**OBSERVATION:**

Long segment intramedullary lesion is seen in the cervical cord extending from C2 to C6 appearing hyperintense on T2WI and hypointense on T1WI. No cord atrophy seen.

Loss of normal cervical lordosis.

Hemangioma/focal fatty infiltration is seen at C2.

Mild posterior disc bulges are seen at C4-5 and C5-6 indenting anterior subarachnoid space without nerve root or cord compression.

Rest of the cervical vertebral bodies, pedicles, laminae, transverse and spinous processes show normal alignment, MR morphology and signal pattern.

Rest of the IV disc spaces show normal height and signal pattern. The facet joints and neural foraminae appear normal.

The cervical cord is well displayed and it is of normal size. No evidence of any abnormal signal intensity lesions seen within the cord. The cervicomedullary junction is normal.

The CSF displays normal signal intensity in all sequences and there is no evidence of any mass in and around the CSF pathway.

Sagittal dimension

C2-3: 10.1 mm
C3-4: 10.0 mm
C4-5: 10.2 mm
C5-6: 10.0 mm
C6-7: 10.0 mm
IMPRESSION:

This examination reveals long segment intramedullary lesion in the cervical cord extending from C2 to C6 suggestive of transverse myelitis.

Suggest: CSF Analysis

Dr. Digesh Devodi Shah
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Consultant Radiologist

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Motor Nerve Studies
UPPER LIMB
Nerve: Right Medial Ulnar + Pin + Cmp

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Sensory Nerve Studies
UPPER LIMB
Nerve: Right Ulnar Wrist

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VER LIMB

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