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Research Paper

A Case Series on Guillain Barré Syndrome

Medicine

KEYWORDS :

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Introduction:
Gullain barré syndrome is an acute,frequently severe and fulminant polyradiculopathy that is autoimmune in nature. It typically presents as a progressive weakness of variable degree from mild paresis to complete paralysis and generalized hypo or areflexia.

Here by we are discussing some of the cases of GBS,in order to understand clinical features and to understand the importance of early intervention which will not only save the life but also improves the quality of life in very young patients.

Case 1:
A 14 year old male hindu patient residing in Ahmedabad presented to VSGH with complain of sudden weakness of lower limb.Patient was very apprehensive and described that he was completely normal before going to school but was not able to walk or even stand after school hours. Patient was thoroughly examined in presence of senior neurologist and it was found that patient was well oriented to time,place and person and examination was normal except for the power in lower limbs was bilaterally 2/5(proximally as well as distally)and complete absence of all deep tendon reflexes. Patient's vitals were normal and there was no bladder or bowel involvement. Past history was not significant except for the episode of self limiting diarrhoea 7 days ago.

Patient's blood was sent for investigations such as complete blood count,renal function tests,liver function tests,electrolyte,B12 level,thyroid profile and lumbar puncture performed for CSF examination.

Results of investigation were inconclusive and did not reveal any abnormality. All investigations were within normal physiological limit. So, based on clinical features diagnosis of GBS made and patient was subjected to further investigations such as electromyography and nerve conduction velocity and urine for porphobilinogen for 3 consecutive days. All causes of differential diagnosis were eventually ruled out and patient was given 5 cycles of plasmapheresis on alternate day and supportive treatment in terms of physiotherapy, nutritional support and prophylactic antibiotic. Further recovery was uneventful and patient was discharged at home without residual abnormality.

Case 2:
A 28 year old married male hindu patient from Jamnagar district of Gujarat state was presented to VSGH(V.S General Hospital) with weakness of lower limb since 3 days. Patient was relatively asymptomatic before 3 days then he developed gradual weakness which progressed over 3 days and patient has problems with micturation also in terms of increased frequency. Patient also recalls history of self limiting upper respiratory tract illness 10 days back. On examination power in lower limb was 3/5(bilaterally symmetrical involving proximal as well as distal muscles equally)knee jerk and ankle jerk were absent and supinator jerk was also absent bilaterally and biceps and triceps jerk were showing hyporeflexia.Plantars were flexors bilaterally. Patient was subjected to various tests as described in previous case .Patient's NCV report showed involvement of common peroneal nerve bilaterally and increased motor latency. Diagnosis of GBS made and patient received 7 cycles of plasmapheresis on alternate day along with supportive care and was sent back to home without any residual abnormality.

Case 3:
An 18 year old male hindu patient from north Gujarat presented to VSGH with complain of lower limb weakness which was bilaterally symmetrical and all deep tendon reflexes were absent. Patient was subjected to tests mentioned earlier but before the receipt of report patient became severely breathless and complained of facial weakness also. Patient's single breath count was only 12.respiratory and cardiovascular system examination were normal.Patient was immediately shifted to intensive care unit where urgent elective intubation done.Reports turned out to be normal.NCV showed conduction slowing in all lower limb nerves.Diagnosis of GBS made and patient was given 2 cycles of plasmapheresis on alternet day but patient did not improve. So,on the advice of neurologist patient was given plasmapheresis everyday for 5 more days.But patient did not improve at all.So,again the advice from neurologist taken and patient was given injectable steroids for 3 more days.patient was maintained on assisted controlled mode of ventilator for more than 200 days but ultimately unfortunately patient died because of ventilator associated pneumonia.

Case 4:
A 29 year old male hindu patient of higher socioeconomical class from Rajkot presented to VSGH with complain of lower limb weakness which was bilaterally symmetrical and there was arflexia.Patient also gave interesting history that he had been diagnosed with GBS 12 years back when he had similar complaints and was given IVIg for the treatment for 5 days.Patient was also tachypneic and his SBC was around 20.Patient was admitted to intensive care unit but he refused central line insertion. Patient was explained about plasmapheresis as a treatment option but patient insisted for more expensive IVIg therapy .Patient was given 2g/kg IVIg over 5 days. Patient improved dramatically within a week and was discharged.

Case 5:
A 13 year old male hindu patient presented with complain of breathlessness and lower limb weakness .Based on diagnostic criteria listed below in discussion session and thorough investigation and detailed investigational work-up, diagnosis was confirmed as GBS. Patient was intubated electively and eventually tracheostomised after consultation with an ENT surgeon.Patient was given 7 cycles of plasmapheresis everyday.It was followed by a short course of injectable steroids. Patient was kept in ICU for more than 100 days and after that was managed in general ward for another week after which he was discharged with full functional recovery.

Discussion:
Diagnostic criteria for AIDP

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1)Required for diagnosis
- Progressive weakness of variable degree from mild paresis to complete paralysis
- Generalized hypo-areflexia

2)Supportive criteria
- Symptom progression: motor weakness rapidly progresses initially but ceases by 4 weeks.Nadir attained by2 weeks in 50%,3weeks in 80%,and 90%by 4 weeks.
- Demonstration of relative symmetry regarding paresis.
- Mild to moderate sensory signs
- Frequent cranial nerve involvement(cranial nerve VII in 50% and typically bilateral but asymmetric)occasional involvement of cranial nerves III,IV,VI,X,XI,XII.
- Autonomic dysfunction can include tachycardia,postural hypotension,hypertension
- A preceding gastrointestinal illness(eg diarrhoea)or upper respiratory tract infection are common.
- Elevated or serial elevation of CSF protein.
- CSF cell counts<10 mononuclear cell/cumm.
- 80%of patients having evidence of NCV slowing/conduction block at some time during disease process.
- Patchy reduction in NCV attaining values<60% of normal.
- Distal motor latency increase may reach 3 times of normal values.
- F wave indicates proximal NCV slowing.
- About 15-20% patients have normal findings.

3)Findings reducing the possibility of diagnosis
- Asymmetric weakness.
- Failure of bowel/bladder dysfunction at initiation of the disease.
- >50 mononuclear cells/cumm in CSF.
- Well-demarcated sensory level.

4)Exclusion criteria
- Diagnosis of other acute neuromuscular weakness(e.g. myasthenia gravis,botulism,poliomyelitis,toxic neuropathy)
- Abnormal CSF cytology suggesting carcinomatous invasion of nerve roots.

Pathogenesis

GBS is an autoimmune disorder which results due to molecular mimicry between antigens of various infectious agents(most common is c.jejuni but others are HSV(herpes simplex virus),CMV(cytomegalo virus), and mycoplasma pneumonia)and self antigens of nervous tissue(GM1 and GQ1b etc).

Differential Diagnoses
- Botulism
- Cauda Equina and Conus Medullaris Syndromes
- Chronic Inflammatory Demyelinating Polyradiculoneuropathy
- Myasthenia Gravis
- Heavy Metal Toxicity
- Lyme Disease
- Metabolic Myopathies
- Multiple Sclerosis
- Nutritional Neuropathy
- Vasculitic Neuropathy
- Acute intermittent porphyria

Treatment

Treatment should be started as early as possible.After two weeks of first motor symptom, immunotherapy may not be effective. High dose of IVIg(2g/kg over 5 days) or plasmapheresis(40-50 ml/kg of plasmapheresis over a week) are equally effective but for some variants of GBS such as acute motor axonal neuropathy(AMAN)and Miller Fischer syndrome will respond better to IVIg therapy. Role of glucocorticoid is controversial. Few neurologists use them based on their experience without obvious clinical evidence.

Progonosis and recovery

Approximately 85% patients will have full functional recovery within a year. However ,minor findings on examination(e.g.areflexia) may persist longer .The mortality rate is<5% in optimal settings, death mostly results from secondary pulmonary infections. Between 5-10%of the patients typical GBS have one or more late relapses. Such cases are then classified as chronic inflammatory demyelinating neuropathy.

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