

5-year-old boy presented with progressive asymmetric weakness of distal extremities, without sensory or cranio-bulbar involvement. NCS showed pure motor demyelinating neuropathy with conduction blocks across non-entrapment sites. He was evaluated on the lines of immune-mediated causes of neuropathy. CBC, RBS, RFT, LFT, thyroid profile, CK and serum electrolytes were normal. Serum vitamin B12, folate, and homocysteine were normal. ESR was normal and CRP was negative. Viral markers (HIV, HBsAg, HCV) and VDRL were negative. ANA profile and ANCA were negative. Ganglioside antibody panel (IgM and IgG) were negative. Serum ACE levels were normal. Urine screening for abnormal electrolytes, serum ammonia and lactate as well as tandem mass spectrometry were non-contributory. CSF analysis showed 1 cell (lymphocyte) and normal protein. MRI (19th August, 2022) showed mild thickening of left brachial plexus. Echocardiogram was normal. CECT of chest, abdomen, and pelvis was normal. Pure tone audiometry was normal. Copy number variation analysis of PMP22 is awaited.

The patient was empirically treated with intravenous immunoglobulins (2 gm/kg, total dose = 30 grams). He developed headache, abdominal pain, vomiting and one episode of fever on second day of IVIG infusion. This subsided with symptomatic treatment (oral ibuprofen). He reported mild subjective improvement in walking difficulty. It is planned to continue monthly IVIG (1 gm/kg, dose = 15 grams/month), assess the clinical response and then decide on the further course of treatment. The patient was clinically and haemodynamically stable and is being discharged with following medical advice.

Course In Hospital

As in discussion

Treatment

Premedication with

Inj Avil 10 mg IV
Inj Paracetamol 150 mg IV
Inj Pantop 15 mg IV

followed by

Inj IVIG (10 grams) as slow infusion on 20 th August.
Inj IVIG (10 grams) as slow infusion on 21 st August.
Inj IVIG (5 grams) as slow infusion on 25 th August.
Inj IVIG (5 grams) as slow infusion on 26 th August.

Advice At Discharge

1) Inj Intravenous Immunoglobulin (5 grams) once daily slow infusion for three consecutive days a month (September, October and November 2022)

Premedications before IVIG

-Inj Avil 10 mg IV
-Inj Paracetamol 150 mg IV
-Inj Pantop 15 mg IV

(to take IVIG under the supervision of pediatrician/ neurologist, if there are no contraindications)

2) To do the following tests before IVIG infusion

- Hemogram: hemoglobin, total leucocyte count, platelet count
- RFT: serum urea and creatinine
- LFT: serum bilirubin, SGOT, SGPT, total protein and albumin
- Fasting blood sugar
- Urine microscopy

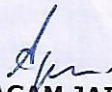
3) Rehabilitation measures to continue.

4) Referred to Bangabandhu Sheikh Mujib Medical University (BSMMU)/ National Institute of Neurosciences and Hospital, Bangladesh, for further management.

In case of Emergency Contact Phone No -

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Signature Of Senior Resident/Consultant

Date & Time