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A case of Guillain Barre Syndrome (GBS) variant: Bilateral tonic eye pupils with autonomic dysfunction in AMAN

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Abstract

Guillain-Barre Syndrome (GBS) is an acquired acute polyradiculoneuropathy characterized by ascending weakness and are flexia and it can has different clinical manifestation and presentations. Here we observe a case of GBS who presented with bilateral tonic pupils associated with autonomic dysfunction and symmetrical weakness of the extremities. His GBS disability scale was 4 upon admission and required intubation for respiratory failure. Nerve conduction study was consistent with Acute Motor Axonal Neuropathy (AMAN). Lumbar puncture showed features of albumin-cytologic dissociation. He was treated with intravenous immunoglobulin 2 g/kg and he was able to be weaned from ventilatory assistance. His autonomic dysfunction recovered at day 29 of admission except bilateral tonic pupils remained persisted. He was successfully discharged after one month of hospitalisation. His GM1a antibody reported as borderline. Hence this case demonstrated ascending flaccid paralysis associated with bilateral tonic pupils and autonomic dysfunction in AMAN variant.

Introduction

GBS is an acquired acute polyradiculoneuropathy with typically characterized by ascending weakness and areflexia which is often preceded with common infections. It occasionally involves the oculomotor nerves but rarely involves the pupils. Here we reported a case of GBS who presented with bilateral tonic pupils associated with autonomic dysfunction and symmetrical weakness of the extremities. His serum GM1a was positive. This showed the association of the bilateral tonic pupils in a case of AMAN variant GBS.

Case Report

A 13 years old boy initially presented with diarrhea and vomiting followed by ascending limbs weakness with acute urinary retention. His GBS disability scale was 4 upon presentation. On neurological examination, he was alert with normal mental function. Both pupils were dilated 6/6 bilaterally and light

reflexes were hardly detected bilaterally. He had bilateral eye ptosis with bilateral facial weakness. He was flaccid tetraplegic and deep tendon reflexes were all absent. Plantar responses were negative and there was no objective sensory impairment. He was tachycardic since admission. Electrocardiogram (ECG) showed sinus tachycardia. Commuted Tomography (CT) brain upon arrival was normal. Nerve conduction study revealed acute motor axonal neuropathy. Intravenous immunoglobulin was immediately initiated. At day 3 of admission he was intubated for respiratory failure due to progressive weakness of respiratory muscle. Complete optic nerve function assessment and pharmacological studies for the pupils was unable to be done as patient was intubated. Lumbar puncture two weeks into the illness revealed albumin-cytologic dissociation. He subsequently needed tracheostomy in view of prolonged ventilation. Treatment with intravenous immunoglobulin 2 g/kg however had led to clinical improvement and he was able to be weaned from ventilatory assistance at day 24 of admission. His autonomic dysfunction recovered at day 29 of admission except bilateral tonic pupils remained persisted. He was then able to be discharged after one month of hospitalisation. Subsequently the result of serum GM1a came back as positive. During clinic follow up which was into 3 months of the illness his muscle power improved MRC grading of at least 3 over all four limbs. His autonomic disturbances such as tachycardic and tonic pupils gradually recovered in the subsequent follow up.

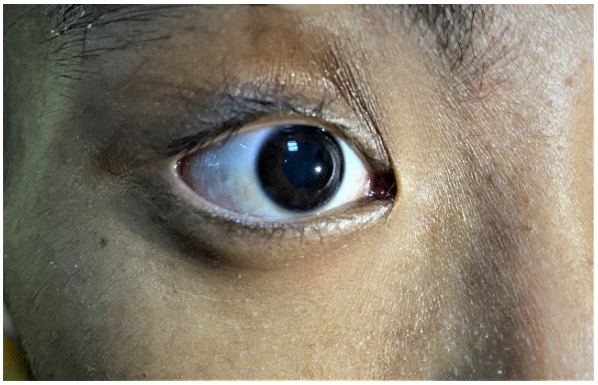

Figure 1a: Right eye (upon admission).

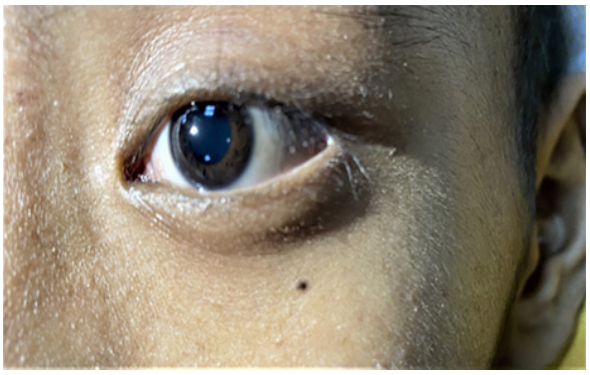

Figure 1b: Left eye (upon admission).

Discussion

Guillain-Barré syndrome is an autoimmune disorder encompassing a heterogeneous group of pathological and clinical entities. Molecular mimicry has resulted in triggering the immune response which subsequently cross reacts with the nerves leading to demyelination or axonal degeneration. There are many subtypes of GBS which include Acute Inflammatory Demyelinating Polyradiculoneuropathy (AIDP), Acute Motor Axonal Neuropathy (AMAN), Acute Motor And Sensory Axonal Neuropathy (AMSAN), dysautonomia, paraparetic, Miller Fisher Syndrome (MFS). The prevalence of each subtype varies greatly based on geographical location with the prevalence of the axonal forms (AMAN and AMSAN) being around 15-35% and the MFS variant being around 1-25%. However case where GBS accompanied by bilateral tonic pupils is very rare. In general, the causes of tonic pupils varies. These etiologies include Adie tonic pupil syndrome, infection and inflammation, local lesion affecting ciliary ganglion or nerve, paraneoplastic causes.

In reference to the pathogenesis in GBS, bilateral tonic pupils could be caused by abnormalities of the ciliary ganglion or a postganglionic parasympathetic branch of the ciliary nerve. In GBS, demyelination

of the postganglionic ciliary nerve can be manifested as tonic pupils as demonstrated here. The sympathetic post ganglionic nerve contains nonmyelinated fibers which are derived from superior cervical ganglion of the sympathetic trunk and reach the dilator pupillae through the short ciliary nerves. These postganglionic fibers might be damaged indirectly by the inflammatory demyelinating process of the short ciliary nerve which resulted in tonic pupils which characterized by poor pupillary light reaction, accommodation paresis, tonic pupillary near response with light-near dissociation and segmental palsy of the sphincter.

Most patients with a tonic pupil do not require any treatment. Patients with an underlying systemic cause for their tonic pupils should have treatment directed at their other autonomic neuropathies. The accommodative paresis tends to resolve spontaneously over several months.

In a more severe case especially with severe motor deficits and respiratory failure especially in AMAN variant, they are associated with autonomic dysfunction as present in this case where he had persistent sinus tachycardia and urinary retention when he presented to us. The widespread demyelination could affect the entire autonomic nervous system with bilateral tonic pupils. The prognosis for recovery is worse for patients with secondary axonal degeneration or when the primary attack is against the axon as in the acute motor axonal neuropathy (AMAN) variant. Our patient showed improvement in motor function gradually but disability scale remained at 4 where he still being wheelchair bound at 4 months into the illness. Aggressive rehabilitation and physiotherapy is important and detrimental for the subsequent recovery.

Conclusion

In conclusion, this is a case of GBS-AMAN variant associated with bilateral tonic pupils. It shows the importance of including it as the differential diagnoses in patients presented with bilateral tonic pupils especially those came with atypical presentation. Heightened suspicious in dealing with neuro ophthalmic presentation in GBS allow us to diagnose early and hence proper and effective treatment can be given to expedite the disease recovery.

Declarations

Acknowledgement: I would like to thank to the family of the patient for allowing us to publish the case report. My gratitude goes to Department of Internal medicine, Hospital Tawau, Sabah.

Consent: Informed consent for the publishing of this case report was obtained from the patient's parents.

Conflict of interest: There was no conflict of interest.

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