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Autoimmunity strikes neuromuscular junction: Myasthenia gravis & achalasia cardia

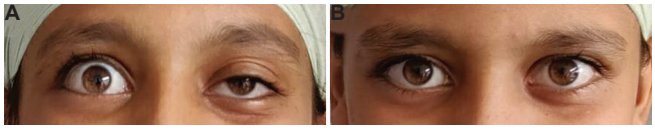

Fig. 1. (**A**) Ptosis of left eye at the time of presentation. (**B**) Improvement in ptosis of left eye after treatment with Pyridostigmine.

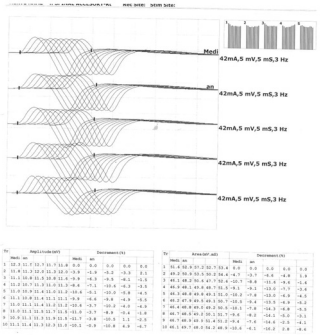

Fig. 2. Repetitive nerve stimulation of median nerve at 3 Hz showing significant decrementing responses in Compound Muscle Action Potential (>10%) suggestive of post-synaptic neuromuscular transmission disorder-Myasthenia Gravis.

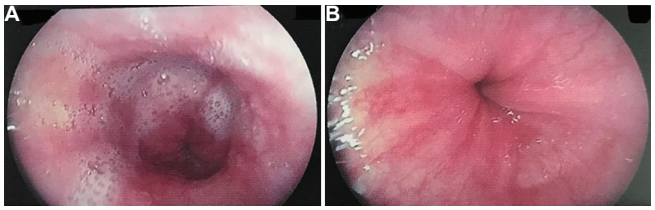

Fig. 3. (**A**) Upper gastrointestinal endoscopy showing dilated oesophagus and (**B**) pooled secretions with tight lower oesophageal sphincter.

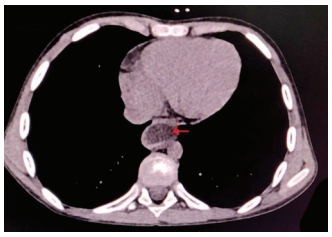

Fig. 4. Arrow in the axial view of computed tomography of the chest showing dilated oesophagus.

A 11 yr old male child† presented to division of Neurology, Guru Gobind Singh Medical College & Hospital, Faridkot, India, in September 2019, with persistent vomiting, difficulty swallowing and regurgitation since one year. Examination showed partial ptosis of the left eye with fatigability (Fig. 1).

Acetylcholine receptor antibody was positive [0.69 nmol/l (Serum reference value <0.25 nmol/l)] and repetitive nerve stimulation test of median and spinal accessory nerve was suggestive of postsynaptic neuromuscular junction disorder - myasthenia gravis (Fig. 2).

Upper gastrointestinal endoscopy (Fig. 3) and computed tomography of the chest showed dilated oesophagus (Fig. 4). Barium swallow X-ray and oesophageal manometry showed the failure of the

†Consent to publish clinical information and images obtained from the patient's parent.

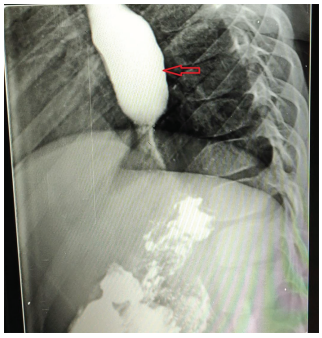

Fig. 5. Arrow in the Barium swallow X-ray showing a prominent bird beak sign, oesophageal dilatation and incomplete relaxation of the lower oesophageal sphincter.

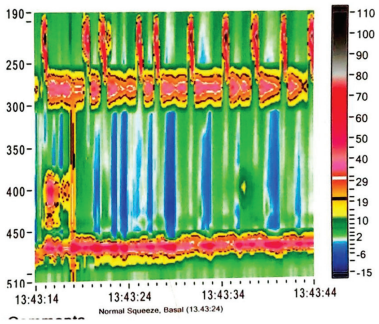

Fig. 6. Oesophageal manometry showing the failure of the lower oesophageal sphincter (LES) to relax, elevated basal lower oesophageal sphincter pressure (51.6 mm of Hg) and aperistalsis of the oesophageal body suggestive of achalasia cardia-type 1.

lower esophageal sphincter LES to relax, elevated basal LES pressure and aperistalsis of the oeasophageal body suggestive of achalasia cardia (Figs 5 and 6). The patient was started on steroids, acetylcholinesterase inhibitor (pyridostigmine) and nifedipine. He responded well to the treatment (Fig. 1B) upon follow up after one month.

Coexistence of myasthenia gravis and achalasia cardia in children has not been reported before. Clinicians should keep an open mind to these rarities as these are amenable to treatment with good prognosis.

***Conflicts of Interest*:** None.

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