Case Report: Challenges in Diagnosis and Management of Myasthenic Crisis in Resource-Limited Health Care Setting

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ABSTRACT

Myasthenia gravis, the most common autoimmune neuromuscular disorder, is characterised by muscle weakness and fatigability. A 23-year-old-lady with background history of breathing and swallowing difficulty for six months was presented with respiratory distress to the hospital which is without an in-house neurology service. Her diagnosis remained a challenge as patient presented as an emergency without detailed medical history. She was subjected to bedside ice pack testing and subsequently managed along the diagnosis of myasthenia crisis. She responded to the treatment and survived the critical period. So, diagnosis of myasthenia gravis by ice pack test and managing a case of myasthenic crisis would be possible in limited heath care setting.

Keywords: myasthenia gravis, ice pack test, myasthenic crisis

INTRODUCTION

Myasthenia gravis (MG), the most common autoimmune neuromuscular disorder, is characterised by muscle weakness and fatigability. There are two patterns described; ocular myasthenia gravis and generalised myasthenia gravis.[1] MG is diagnosed based on appropriate clinical context supported by diagnostic tools like serology testing, edrophonium testing, repetitive nerve stimulation testing and single fibre electromyography.[2] Myasthenia gravis is easily diagnosed in tertiary centre with neurology service and adequate resources. However, it is a diagnostic challenge to doctors in smaller hospitals who are mostly the novices in medical hierarchy due to limited resources. Ice Pack Test has long been described as an alternative method in aiding myasthenia gravis diagnosis. Unfortunately, this simple and safe bedside diagnostic tool has been under-utilised and remains unknown to majority of the health care providers.

CASE PRESENTATION

A 23-year-old-lady without any known medical illness was brought in by her family, complaining of difficulty in breathing associated with high-grade fever and cough for two days. She has background of breathing and swallowing difficulties for six months with worsening a day prior to admission. She has been seen and treated by different primary health care providers without any improvement of her symptoms. Her medical problem remains unresolved. As she presented acutely, a detailed history and examination were not possible.

Upon admission she was cyanosed with low Glasgow Coma Scale (Eye: 2, Verbal: 1, Motor: 1). Her blood pressure was 128/67 mmHg, with heart rate of 118 beats/min and SpO2 88% under room air temperature of 38.5°C. Her blood investigations revealed that Hb concentration was 12.7 gm/dl, total white cell count 20.6× 10^9 /L, platelet count 399 × 10^9 /L, ESR > 140 mm/H. Her biochemical profile for urea 2.0 mmol/L, sodium 142 mmol/L, potassium 2.84 mmol/L and creatinine 61 µmol/L. Her total bilirubin was 14.3 µmol/L, albumin 40.6 g/L, ALT 8.4 U/L, AST 17.8 U/L and ALP 35 U/L. C-reactive protein (CRP) was 11.62 mg/L. Her arterial blood gases (ABG)

analysis were pH 7.32, pO2 173 mmHg, pCO2 38.4 mmHg, HCO_3 19.7 mmol/L. Chest x-ray showed heterogeneous opacity over both lungs with lower lung field more prominent over the right side. Computer tomography (CT) of brain was unremarkable without any features suggestive of stroke. Blood and sputum specimen sent for culture and sensitivity but no organism was isolated.

She was intubated for respiratory failure, and treated for aspiration pneumonia with intravenous Ceftriaxone in ICU setting.

During her stay in ICU, two attempts of extubation were tried by anaesthetist but in vain. Upon weaning her off from sedation, there were copious amount of oral secretion needing regular suctioning and obvious bilateral ptosis noted. There was bilateral facial paresis with pupillary sparing ptosis. Neurology assessment of limbs revealed tetraparesis with medical research council power grading 4 out of 5 for all 4 limbs accompanied by intact tendon reflexes and flexor plantar responses.

She was initially planned for a transfer to tertiary centre for further management by neurology team. However this plan appeared unwise given that the land transport (the only mode available in our setting) takes approximately four hours and she was deemed unstable for transfer.

Hence she was subjected to ice pack test based on the clinical findings of bulbar weakness and bilateral ptosis . Figure 1 shows her eyelids position before ice pack test. The ice pack result is as in Table 1. Figure 2 shows her eyelids position after application of ice pack test. There was a drastic improvement in term of her ptosis after two minutes of ice pack application.

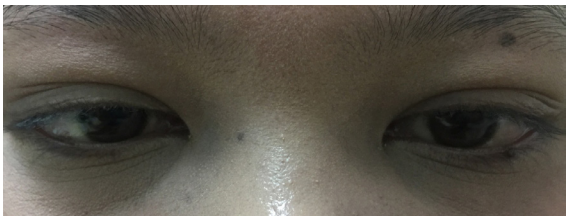

Figure 1 Eyelids position before ice pack test

Figure 2 shows her eyelids position after application of ice pack test. There was a drastic improvement in term of her ptosis after two minutes of ice pack application.

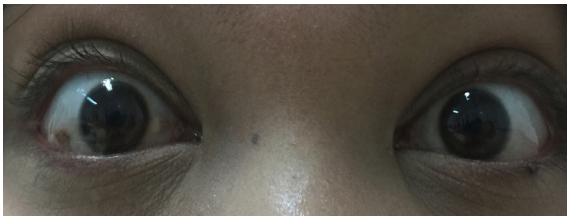

Figure 2 Eyelids position after ice pack test

Table 1 Ice pack test result

Interpalpebral distance	Right	Left
Before ice pack	10 mm	10 mm
After ice pack	12 mm	13 mm

As her ice pack test was positive, blood specimen was taken and sent to tertiary hospital for acetylcholine receptor antibody testing.

There are few differential diagnoses to be considered in a young lady with bulbar weakness and bilateral ptosis, namely myasthenia gravis, Guillain Barre Syndrome (GBS), botulism, and acute stroke with brain stem involvement.

She was treated as myasthenic crisis. This is because she presented with respiratory distress requiring intubation and ventilatory support. Tracheostomy was done for her in view of repeated failed extubation. She was treated with antibiotic (Ceftriaxone for one week duration) for aspiration pneumonia, intravenous immunoglobulin (IVIG), high-dose Prednisolone at 1 mg/kg/day (initiated at day four of IVIG), subcutaneous Enoxaparin for thromboembolic prophylaxis and tablet

Pyridostigmine 30 mg 5 times/day (initiated prior to discharge). Clinically, she progressively showed improvement and was discharged well after 2 weeks of inpatient treatment.

Six weeks after discharge, she achieved full resolution of her weakness and was independent in carrying out activity of daily livings. Tracheostomy tube had been removed. Her Acetylcholine receptor antibody came back as positive during clinic follow-up (> 8.4 nmol/L; normal < 0.4 nmol/L) which confirmed the diagnosis of myasthenia gravis.

DISCUSSION

This is a challenging case seen in a resource-limited health setting. Patient was presented acutely requiring ventilatory support and ICU care without much clinical history. Thus doctors were to treat her based on best clinical judgement and using all the slightest clues available. The unavailability of neurology service added onto this challenge. Escalation of care to tertiary centre was impractical given patient's instability for transfer. Neurological consult with neurologist of neighbouring tertiary hospital was available via phone consultation; yet this is not of much help given that the neurologist did not have the opportunity to assess the patient and had to rely on assessment of the referring doctor. Serological investigations such as acetylcholine receptor antibody take approximately three to four weeks to be ready as the specimen has to be sent to a tertiary centre laboratory for processing. Neurophysiological study and edrophonium test were also not available given the similar reason as above.

According to the clinical judgement, she had myasthenia gravis based on the presence of bulbar weakness, bilateral ptosis and positive ice pack testing. As she presented acutely with respiratory distress requiring intubation and ventilatory support. So, her case was considered as myasthenic crisis. This case was managed based on the latest recommendation.[3]

The initiation of high-dose steroid may worsen myasthenia gravis in 50% of patients and even precipitate a crisis in another 10%.[3] The quick action onset of IVIG helps to prevent the transient worsening of MG associated with steroid initiation.[3] In this case, IVIG was used as acute therapy for myasthenia crisis. Total IVIG given to her was 2 g/kg over the course of 5 days (0.4 g/kg/day). Steroid was introduced at day 4 of IVIG when infection was well controlled with antibiotic treatment based on clinical and microbiological parameter.

Ice pack testing in myasthenia gravis is well described in many literatures.[4, 5, 6, 7, 8] This test is performed by objective measurement of interpalpebral distance before and after the application of ice packs. The ice used should be packed and placed on closed eyes for 2 minutes as to minimise the risk of cold-induced injury. Two independent observers should be available to measure the interpalpebral distance as to prevent bias in result interpretation. Ice pack test is considered positive if there is objective improvement of the ptosis by at least 2 mm of the interpalpebral distance. The principles governing Ice Pack Testing are that acetylcholinesterase activity of skeletal muscle reduces with lower temperature. The risk of this procedure is minimal compare to edrophonium where the later has the risk of precipitating heart block. Edrophonium test can only be conducted by an experienced neurologist with standby resuscitation trolley in case of acute cardiac event.

In limited health care setting, this was the only alternative available. Ice pack test can be conducted at the bedside by non-neurologist, much safer compare to edrophonium test and cost effective. Moreover, a study comparing ice pack test and edrophonium test showed that ice pack test had sensitivity and specificity of 100% in myasthenia gravis patient.[4] However, ice pack test may not be reliable in cases of isolated diplopia without ptosis.[4] In this patient, ice pack test is reliable as she had ptosis.

CONCLUSION

Myasthenic crisis need to be suspected in a patient presented with weakness, difficulty in breathing and swallowing. Ice Pack Test is a simple, reliable, safer bedside test that can aid in the diagnosis of myasthenia gravis especially in resource-limited health setting.

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CONFLICT OF INTEREST

The author declares that he has no competing interests.

CONSENTS

Written informed consent was obtained from the patient to publish the case with its related pictures. A copy of the written consent is available for review by the Chief Editor of this journal.

REFERENCES

1. Bird SJ. (2017). Clinical manifestations of myasthenia gravis. UpToDate.
2. Philllips LH, Melnick PA. (1990). Diagnosis of myasthenia gravis in the 1990s. Semin Neurol 10: 62 – 69.
3. Bird SJ, Levine JM. (2017). Myasthenic crisis. UpToDate.
4. Tabassi A, Dehghani A, Saberi B. (2005). The ice test for diagnosing myasthenia gravis. Acta Medica Iranica 43 (1): 60 – 62.
5. Lertchavanakul A, Gamnerisiri P, Hirunwiwatkul P. (2001). Ice test for ocular myasthenia gravis. J Med Assoc Thai 84 (Suppl 1): S131 – S136.
6. Sethi KD, Rivner MH, Swift TR. (1987). Ice pack test for myasthenia gravis. Neurology 37 (8): 1383 – 1385.
7. Czaplinski A, Steck AJ, Fuhr P. (2003). Ice pack test for myasthenia gravis; a simple, noninvasive and safe diagnostic method. J Neurol 250: 883 – 884.
8. Liu WW, Chen A. (2016). Diagnosing Myasthenia Gravis with an Ice Pack. N Engl J Med 375: e39. DOI: 10.1056/NEJMicm1509523
9. Larner AJ, Thomas DJ. (2000). Can myasthenia gravis be diagnosed with the 'ice pack test'? A cautionary note. Postgrad Med J 76: 162 – 163.