Management of myasthenic crisis in a child

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ABSTRACT

Myasthenia gravis is an autoimmune disorder of peripheral nervous system, leading to fluctuating muscle weakness. It is caused by circulating antibodies that block acetylcholine nicotinic post synaptic receptors at the postsynaptic neuromuscular junction. Myasthenic crisis is a life-threatening complication, which is defined as weakness from acquired myasthenia gravis.

In this paper we described a 15-year-old boy who was admitted to the Paediatric Intensive Care Unit due to myasthenic crisis. He had suffered not only from myasthenia gravis but also hypothyroidism, cerebral palsy and epilepsy. The patient required mechanical ventilation and was successfully treated with both plasmapheresis and intravenous immunoglobulins. He recovered from the crisis and then thymectomy was performed. Perioperative period and anaesthesia passed uneventfully. Discharged home from the hospital after 2.5 month-treatment, for the last 4 years, he has only come on scheduled outpatient medical appointments.

This case reveals that myasthenic crisis, albeit rare, may occur in male adolescents. In such cases multidisciplinary care followed by surgery becomes a procedure of choice. Concomitant medical problems, if well controlled, do not affect the results of the outcome of the underlying disease.

Key words: myasthenia gravis, children; myasthenia gravis, myasthenic crisis; myasthenia gravis, general anaesthesia; myasthenic crisis, management; myasthenic crisis, plasmapheresis

CASE REPORT

A 15-year-old boy (with a weight of 70 kg and a height of 165 cm) was admitted to the ICU of the Children’s University Hospital from the neurology ward due to progressive hypodynamic respiratory failure and bulbar symptoms. His history revealed myasthenia of the lower limbs and articulation disorders that had been detected a year earlier. Based on clinical examinations, the patient was diagnosed with MG, and treatment with pyridostigmine at a dose of 60 mg day⁻¹ was initiated. Moreover, a diagnosis of hypothyroidism was made, and levothyroxine was started 1 × 50 μg day⁻¹ (euthyroidism for 3 months). The patient had a history of infantile cerebral palsy that had been diagnosed at the age of 1 year as moderate spastic paraplegia and epilepsy treated with carbamazepine.

Upon ICU admission, the patient was conscious (GCS 15) and...
had both difficulty lifting his head and blepharoptosis. He required passive oxygen therapy; his maximum SpO$_2$ was 84% and his pCO$_2$ was 78 mm Hg. Endotracheal intubation was performed and SIMV was delivered with the following parameters: IP, 30 cm H$_2$O; FiO$_2$, 0.21; f, 12 min$^{-1}$; and PEEP, 4 cm H$_2$O. SpO$_2$ increased to 97–99%. A chest X-ray (Fig. 1) performed after ICU admission revealed bilateral perihilar atelectatic changes and a dilated mediastinal shadow, which had been described as suspected thymoma six months earlier on a CT scan (approximately 45 × 20 × 25 mm). A repeat CT performed on day 2 of hospitalisation (Fig. 2) revealed residual thymus, approximately 24 × 10 × 32 mm, in the superior anterior mediastinum.

The treatment included pyridostigmine (480 mg day$^{-1}$), prednisolone and intravenous polyclonal antibodies (0.5 g kg$^{-1}$ day$^{-1}$ for 4 days). Two plasmapheresis procedures were performed on treatment days 2 and 3 (parameters in Table 1). Difficulties occurred during the first procedure: low pressure in the collecting line prevented the collection and return of blood to the double-lumen dialysis catheter (inserted through the right femoral vein). An additional central catheter was placed via the internal jugular vein. Following this complication, the first plasmapheresis was uneventful. During the second plasmapheresis (started 36 hours after the first), low pressure was observed in the collecting line, but the procedure was continued.

On day 4 after the second plasmapheresis (i.e., on day 6 of mechanical lung ventilation), once obligatory ventilation was reduced, spontaneous efficient respiration was observed and the patient was extubated. The bulbar symptoms were not observed.

Because thymectomy was indicated, a thoracic surgeon was consulted, and the patient was transferred to the Department of Thoracic Surgery. Spirometry revealed FEV$_1$ at 1950 mL (85 %) and FEV$_1$/FVC at 86 %. The risk of general anaesthesia was assessed at ASA class III. After premedication with 25 mg hydroxyzine p.o., anaesthesia was induced using atropine (0.4 mg), fentanyl (0.1 mg), thiopental (325 mg) and cisatracurium (6 mg). Endotracheal intubation was performed and IMV was initiated with a mixture of N$_2$O and O$_2$ (2:1 at FGF 6 L min$^{-1}$) with sevoflurane 1.7 vol%. During the maintenance of anaesthesia, the patient received additionally fractionated doses of fentanyl (0.3 mg in total). The neuromuscular function of the patient was monitored using the TOF-Watch® SX device. The patient did not require additional doses of muscle relaxant. The tumour was removed through sternotomy; macroscopically, it exhibited the features of thymoma. The duration of surgery was 1 h and 45 minutes, and that of anaesthesia was 2 h and 35 minutes. The circulation and gas exchange of the patient during the procedure were efficient. Thirty minutes after the completion of surgery, due to a delayed return of muscular strength, the patient was administered atropine (0.6 mg) and neostigmine (2 mg), which enabled his extubation and transfer to the postoperative ward. No complications were observed during the perioperative period.

The histopathological examinations revealed thymus hyperplasia and ectopic parathyroid glands. On day 9, the patient was transferred to the neurology ward; after 2.5 months of continuous hospitalisation, he was discharged from the hospital in good general condition. Since the operation (4 years), the patient has not been hospitalised and has only attended routine follow-ups in the outpatient neurology clinic.

DISCUSSION

Myasthenia gravis is extremely rare in children and myasthenic crisis is even rarer; therefore, it is difficult to find descriptions of treatment options, and causal studies of these issues are lacking [2].

In general, myasthenic crisis develops 2–3 years after the diagnosis and is induced by a triggering stimulus (e.g., anti-epileptic drugs or an infection) [3]. In our case, myasthenic crisis occurred one year after the diagnosis, and the only
inducing factor was the use of carbamazepine, although the pre-crisis plasma level was within reference values. Moreover, epileptic seizures were not observed. Respiratory failure caused by a weakening of the respiratory muscles and those maintaining tonus and patent upper airway was the indication for intubation and substitutive ventilation until the suitable muscle strength was regained. This condition of the patient is categorised as grade 3 or 4 on the Osserman scale and as class V according to the Myasthenia Gravis Foundation of America classification [4].

Standard management (including intubation, IPPV, steroid therapy, and pyridostigmine) is complemented with plasmapheresis and/or immunotherapy. Several cases of adult patients undergoing plasmapheresis have been described [3, 5]. Only a few studies have compared the effectiveness of non-specific antibodies and plasma exchange [6, 7]. One of the few prospective, randomised studies (which was carried out on a group of 87 patients) did not indicate a significant difference in the clinical improvement between patients treated with high doses of immunoglobulins and those administered plasmapheresis. According to advocates of immunotherapy, high doses of immunoglobulins (0.4 g kg\(^{-1}\) day\(^{-1}\) for 5 days) are equally effective and carry a lower risk of complications [6]. In our case, although polyvalent antibodies were applied, radical improvement was observed after two plasma exchange procedures. The complications that could be attributed to plasmapheresis were slight, and discontinuation of the procedures was not necessary, as in the cases described in the literature [3, 6]. Moreover, the amount of plasma exchanged at any one time was higher in our patient (3254, 2662 mL) compared to the case reported in literature, which also affected the efficacy of plasmapheresis [3].

Patients often qualify for thymectomy, which improves the prognosis and long-term treatment outcomes [5, 8, 9]. The histopathological examinations usually reveal thymus hyperplasia; thymoma is observed less frequently [9]. To date, ectopic parathyroid glands have not been described in literature as accompanying thymus removal due to myasthenia gravis. In our patient, they did not cause symptoms of hyperparathyroidism. Surgical procedures under general anaesthesia are usually uneventful when suitable doses of the non-depolarising relaxant are administered and broken down via Hofmann elimination and when the neuromuscular transmission is monitored.

### References:


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### Table 1. Parameters of plasmapheresis

<table>
<thead>
<tr>
<th>Duration (min)</th>
<th>Input/flow intensity ratio</th>
<th>Plasma (mL)</th>
<th>5% albumin (mL)</th>
<th>CES* (mL)</th>
<th>AC (mL)</th>
<th>10% Ca (mL)</th>
<th>Level of albumin Before</th>
<th>After</th>
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<td>769</td>
<td>20</td>
<td>4.42</td>
<td>3.71</td>
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</table>

CES — compound electrolyte solution, AC — anticoagulant