CASE REPORT

A Case of Myasthenia Gravis with Steroid-induced Bilateral Avascular Necrosis Neck of Femur Posted for Bilateral Core Decompression

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Abstract

Postoperative myasthenic crisis (POMC) is one of the leading causes of respiratory failure, requiring intubation or mechanical ventilation within 24 hours to 7 days following surgery in myasthenia gravis (MG) patients. Its incidence ranges from 11.5 to 18.2% in MG patients. A 32-year-old male, coming from Chengalpattu belonging to socioeconomic class III was a known case of MG for 1½ years, belongs to grade I of Osserman classification (ocular weakness without bulbar involvement) and was on oral prednisolone 30 mg/day and pyridostigmine 60 mg/day. The patient was diagnosed with a bilateral avascular necrosis neck of the femur, posted for core decompression. Various studies were aimed to develop and validate a simple clinical prediction score for POMC risk based on data from patients with MG. With this clinical prediction score, following certain anesthesia strategies will minimize its incidence and make their perioperative journey safe.

Keywords: Non-relaxant anesthesia, Perioperative steroid therapy, Postoperative myasthenic crisis.

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Introduction

Myasthenia gravis is an autoimmune disorder with autoantibodies directed against acetylcholine receptors (AChR) in the postsynaptic membrane of the neuromuscular junction, 85% will have AChR antibodies, 10% for muscle-specific tyrosine kinase (MuSK), and 5% will be seronegative (Fig. 1). Optimization of regular medication like pyridostigmine with MG patients is critical as overtreatment leads to an increase in muscarine effect that leads to cholinergic crisis and undertreatment causes weakness myasthenic crisis.[1] Myasthenic crisis is defined as weakness of pharyngeal muscles causing airway collapse and larynx obstruction leading to respiratory failure, requiring intubation or mechanical ventilation, which is common postoperative hypoxia in MG patients.

Case Report

A 32-year-old male, coming from Chengalpattu belonging to socioeconomic class III was a known case of MG for 1½ years, belongs to grade I of Osserman classification (ocular weakness without bulbar involvement) and was on oral prednisolone 30 mg/day and pyridostigmine 60 mg/day (Fig. 2). The patient was diagnosed with a bilateral avascular necrosis neck of the femur, posted for core decompression (Fig. 3). The patient was assessed and fit under ASA II (MG) planned under regional anesthesia with a BMI of 27.2. As per the POMC prediction score, patients fall into score 1 as duration of illness falls under 1–2 years and all other clinical predictions are absent (score <2.5 has 10% of POMC). Following anesthesia strategies have been used to reduce the chance of POMC:

- A morning pyridostigmine dose of 20 mg was given on the day of surgery.
- Regional anesthesia (Fig. 4)—combined spinal and epidural anesthesia was administered.
- Amide group of local anesthetic agents (bupivacaine) was used.
- Intraoperative high-dose steroid (injection hydrocortisone 100 mg IV) was administered as per guidelines of perioperative steroid management.
- Postoperative pain was managed with epidural infusion (0.125% bupivacaine).
- Vigilant postoperative monitoring for cough during deglutition, hoarseness of voice, shallow respiration, restlessness, and anxiety was done.

Discussion

Postoperative myasthenic crisis is defined as weakness of pharyngeal muscles causing airway collapse and larynx obstruction leading to respiratory failure, requiring intubation or mechanical ventilation within 24 hours to 7 days.[2] Its incidence ranges from 11.5 to 18.2% in MG patients. It is characterized by anxiety, cough while swallowing, weak voice, difficulty in counting 20 Nos, shallow respiration, and respiratory distress.[3] Various

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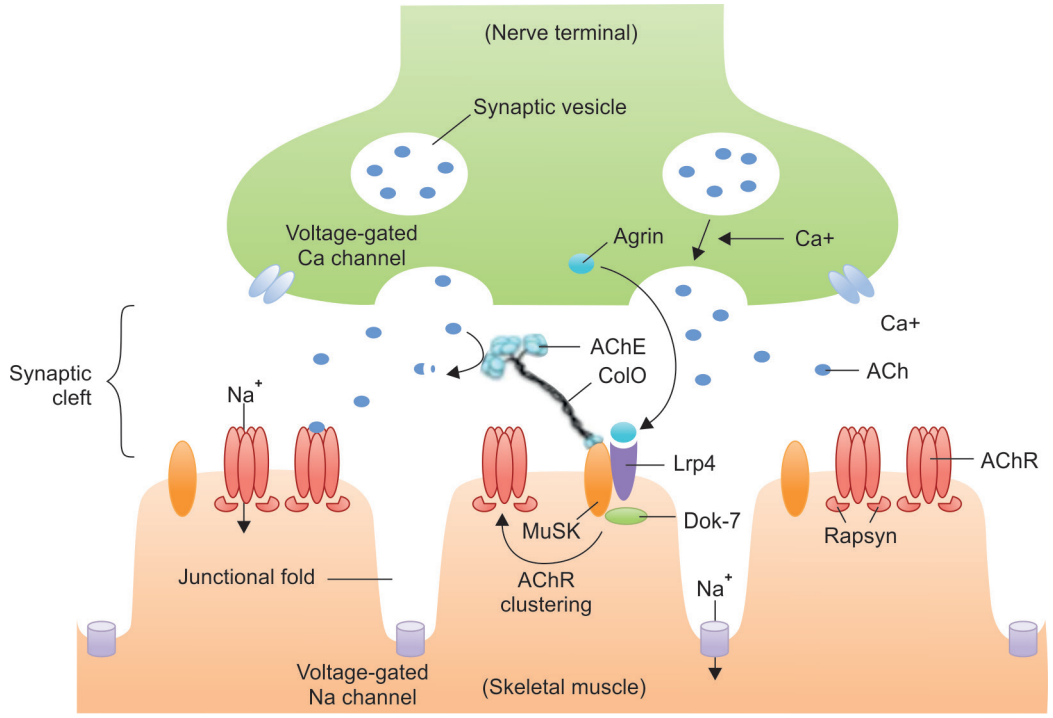


Fig. 1: Structure and molecular architecture of a neuromuscular junction

Fig. 2: Patient

studies were aimed to develop and validate a simple clinical prediction score for POMC risk based on data from patients with MG.

One such accepted prediction score for POMC (Table 1) states that if the score is less than 2.5 associated with 10% of myasthenic crisis and a score of four predicts a 50% chance of POMC.[4,5]

Anesthesia Strategies

Preop

- Proper clinical evaluation and grading of disease.
- Preop pulmonary function test and scoring.
- Continue the regular medication of anticholinesterase on the day of surgery.
- Consider for plasmapheresis if bulbar or generalized weakness is present.
- Preop respiratory muscle training like incentive spirometry usage.

Intraop

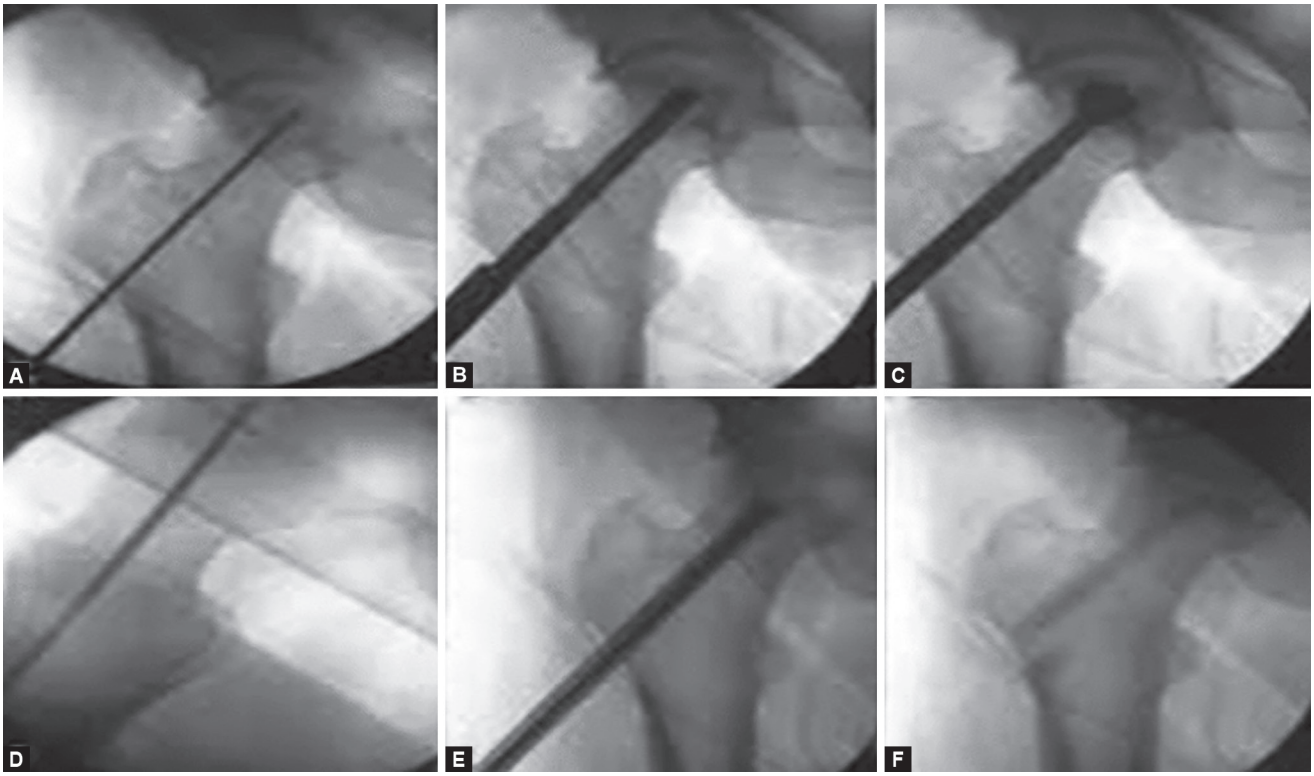
- Avoid narcotics and sedatives as premedication.
- General anesthesia can be safely given by the combination of intravenous/inhalation anesthetic agents and avoiding the use of muscle relaxants (non-relaxant anesthesia).
- Whenever possible, promote regional anesthesia and nerve blocks.
- Prefer for an amide group of local anesthetic agents than esters.
- High-dose steroids should be administered (Table 2).

Postop

- Postoperative pain management without opioids and narcotics in view of respiratory depression.
- Vigilant postoperative monitoring for signs and symptoms of myasthenic crisis.

Conclusion

Myasthenia gravis patients need a multidisciplinary approach by understanding the pathophysiology of the illness, meticulous planning, wise choice of anesthesia technique and drug, intraoperative management, and postoperative monitoring makes their perioperative journey safe.



Figs 3A to F: C-arm image of core decompression (intraoperative)

Fig. 4: Administration of spinal anesthesia to patient

Table 1: The predictive score for POMC[4]

Variables associated with POMC	*Assigned points (range: 0.0–8.5)*
Osserman stage	
Stage I–II A	0
Stage II B	1
Stage III–IV	3
Duration of MG (years)	
<1	0
1–2	1
>2	2
Lung resection	
No	0
Yes	2.5
BMI	
<28	0
>28	1

Table 2: Perioperative guidelines for steroid therapy[5]

Patients who have received a regular daily dose of more than 10 mg prednisolone or equivalent in the last 3 months	
Minor surgery (hernias, extremity surgeries)	25 mg hydrocortisone at induction
Moderate surgery (hysterectomy)	Usual preop steroids +25 mg hydrocortisone at induction +100 mg hydrocortisone/day
Major surgery (Major trauma, prolonged surgery, or surgery where there is delayed oral intake)	Usual preop steroids +25 mg hydrocortisone at induction +100 mg hydrocortisone/day for 2–3 days Resume normal oral therapy when the gastrointestinal function has returned
For all other patients—no additional steroids are required	

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