

Anaesthetic management of a patient with multiple system atrophy

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Multiple system atrophy (MSA) is a rare adult-onset neurodegenerative disease. Symptoms vary from autonomic dysfunction to Parkinsonism and cerebellar ataxia, in any combination. MSA affects many organ systems with many possible complications and makes perioperative management of a patient with this condition very challenging. Successful management of a patient with MSA posted for laparoscopic nephrectomy under general anaesthesia is reported.

Keywords: laparoscopy, multiple system atrophy, Parkinsonism

Introduction

Multiple system atrophy (MSA) is a progressive, adult-onset neurodegenerative disease characterised clinically by autonomic dysfunction, Parkinsonism, and cerebellar ataxia in any combination. MSA was originally thought to be three different diseases: Shy-Drager syndrome, striatonigral degeneration, and sporadic olivo-pontocerebellar atrophy. These terms are no longer in use, but are now considered to be components of MSA.¹

Perioperative and postoperative haemodynamic and respiratory management are important considerations in these patients. We report successful management of a patient with MSA posted for laparoscopic nephrectomy under general anaesthesia.

Case report

A 62-year-old female, with controlled hypertension and diabetes mellitus, developed symptoms of imbalance while walking, frequent falling, intention tremors, incoordination, altered speech, swallowing difficulty, choking and coughing bouts while feeding, recurrent lower respiratory tract infections and loss of bladder control for the past two years. She was diagnosed as a case of MSA type C on clinical picture and investigations. She was treated with levodopa plus carbidopa, piracetam, citicholine and multivitamins. She underwent bone marrow derived multinuclear cell (BM MNC) transplantation for the same condition with little change in symptoms. She was then posted for a laparoscopic right partial nephrectomy due to an exophytic mass in the lower pole of right kidney.

Preoperatively blood pressure (BP) in both sitting and supine position revealed mild hypertension. Cardiac, respiratory and abdominal systems were normal.

Neurological examination revealed slurring of speech, tremors, hypotonia, hyper-reflexia, inability to walk without support, frequent falling, dysarthria, dysphagia, nystagmus, involuntary movements in all four limbs, and ataxic gait with loss of coordination.

Airway examination revealed adequate mouth opening, neck extension and Mallampati class 2.

Routine blood investigations, chest X-ray and arterial blood gases were normal. A pulmonary function test showed a mild restrictive element. Two-dimensional transthoracic echo revealed grade 1 diastolic dysfunction with preserved ejection fraction.

Computed tomography scan of the brain revealed patchy hypodensities in bilateral fronto-parietal white matter and right ganglio-capsular regions, suggestive of ischaemic changes.

Magnetic resonance imaging scan of the brain revealed multiple small ischaemic foci in both cerebral hemispheres with periventricular white matter neurodegenerative changes and diffuse cerebral atrophy.

The patient was advised to take levodopa and carbidopa, plus antihypertensives, on the morning of surgery with sips of water.

In the operating room, all routine monitors (five-lead ECG, non-invasive blood pressure monitor [NIBP], pulse oximeter) were attached. In addition, the right internal jugular vein and right radial artery were cannulated for central venous and continuous blood pressure monitoring. Prior to induction, her pulse was 90 beats/min and BP 150/90 mmHg, with saturation of 98% on room air. The patient was pre-oxygenated with 100% oxygen and sedated with midazolam 2 mg and fentanyl 150 µg. Induction was done by propofol 130 mg and atracurium 25 mg. The airway was secured with a 7.5 mm endotracheal tube with the help of a C-Mac videolaryngoscope.

The patient was placed in the left lateral position for surgery. Dependant parts were adequately padded. Anaesthetic depth was maintained with desflurane at 1 mac with O₂:air at 50:50. Blood pressure rose to 170/100 mm of Hg on insufflation of carbon dioxide, which was managed by increasing desflurane concentration. Twenty-five minutes into the surgery, BP fell from 140/90 mmHg to 75/35 mmHg, which was managed with fluid boluses and 12 mg of ephedrine. The pulse rate stayed steady at 70 beats/min without much change even during periods of hypotension. The surgery lasted for two hours with 100 ml blood loss. The patient was given 1500 ml of Ringer's lactate during the procedure with 200 ml of urine output. Pain was managed with fentanyl top-ups hourly and intravenous paracetamol 1 gm.

The patient was reversed and extubated after adequate attempts. After extubation, there was an episode of airway obstruction, which was managed by inserting a nasal airway. The patient was moved to the ICU for observation, where she was haemodynamically stable. She was transferred to the ward the next day.

Discussion

First described by Gram and Oppenheimer in 1969, MSA is characterised by autonomic failure, cerebellar ataxia and Parkinsonism in various combinations.² Two phenotypes are distinguished clinically by the predominant motor symptom: the Parkinsonian variant (MSA-P) and the cerebellar variant (MSA-C).³

The prevalence of MSA is very low (1.94 to 4.4/100 000) with onset in the fifties, independent of gender.⁴ It is diagnosed by symptoms, progression and brain imaging. Treatment consists of management of supine hypotension, hypertension, movement disorders and anti-Parkinsonian medications. BM MNC is also being used as a modality of treatment for MSA. It exerts neuroprotective effects against dopaminergic neuronal death.

Disease progression is much faster in MSA than in Parkinson's disease (PD) without any drug treatment that provides MSA patients with consistent long-term benefits.⁵ The clinical diagnosis of MSA is difficult and there are no pathognomonic features to discriminate the Parkinsonian variant (MSA-P) from PD. Hence some warning signs, also called red flags, may be helpful to corroborate MSA as the effective diagnosis. They are divided into six categories: early instability, rapid progression, abnormal posture, bulbar dysfunction, respiratory dysfunction, and emotional incontinence.⁶

Perioperative management of MSA remains critical. Disturbances in the autonomic and central nervous systems result in defective baroreflex and vasoconstriction of capacitance and resistance vessels, with an unpredictable response to vasoconstrictor agents. Blood loss has to be anticipated and dealt with efficiently as these patients do not respond with appropriate tachycardia in response to hypotension. Hence blood and blood products should be kept on standby. Also, positive pressure ventilation could lead to blood pressure instability due to a decrease in venous return. Autonomic involvement of the gut leads to higher risks of aspiration under anaesthesia, especially if patients have a full stomach. Involvement of the laryngeal muscles may lead to increased chances of laryngospasm and laryngeal dystonia. Interaction of opioids and muscle relaxants in these patients may be complicated.⁷ The presence of obstructive sleep apnoea in these patients makes the use of sedation difficult. Dangerous airway obstruction upon induction of or emergence from general anaesthesia may occur, but this obstruction is relatively easy to overcome by applying minimal positive pressure. Vocal cord abductor paralysis (VCAP) is well described and may cause nocturnal sudden death in patients with multiple system atrophy. Nocturnal stridor is the sole symptom of VCAP and can make the diagnosis very difficult.⁸ The chance of vocal cord paralysis is aggravated in the postoperative period due to insufficient depth during anaesthesia or unnecessary suction. Bronchoscopic intubation can be used in these patients, and a tracheostomy kit should be kept on standby.⁹ These patients have a high incidence of postoperative ventilatory failure, poor recovery from general anaesthesia and ventilator

dependence, hence ventilator back-up is advised. Despite several reported cases of hypotension, general anaesthesia is considered the gold standard in multiple system atrophy.⁷ We decided on general anaesthesia due to non-cooperation, involuntary movements and regional anaesthesia not being suitable for laparoscopic surgery. Rigidity, tremors and involuntary movements may make regional anaesthesia difficult to perform. In addition, autonomic imbalance after subarachnoid block may be more pronounced. Successful management of MSA patients under regional anaesthesia has been described. A recent article describes the management of a patient with MSA posted for transurethral resection of the prostate being managed under spinal anaesthesia.¹⁰

In conclusion, perioperative management of a patient with MSA can prove challenging for the anaesthetist, with many possible complications. However, these cases can be successfully managed and complications can be minimised with appropriate knowledge of the disease, thorough preoperative examination and tailoring the anaesthetic technique according to the extent of disease and type of surgery.

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