Anaesthetic management in a patient with progressive supranuclear palsy

Sir,

Progressive supranuclear palsy (PSP), also known as Steele–Richardson–Olszewski disease, is a neurodegenerative disorder affecting the basal ganglia and characterised by supranuclear gaze palsy and postural instability. It is the most common form of atypical Parkinsonism, with a prevalence of 6/100,000.\(^1\) Different aspects of PSP must be considered to preplan the best anaesthetic approach.

We recently administered anaesthesia to a 69-year-old female with a history of PSP undergoing total hip replacement. Physical examination showed generalised hypertonia with cervical backward tilt, dysphagia, and moderate cognitive impairment. Her condition was being symptomatically treated with carbidopa–levodopa 25 mg/100 mg PO q8h, baclofen 5 mg PO q8h, tizanidine 2 mg PO q8h, mirtazapine 30 mg/day PO, venlafaxine 75 mg/day PO, and zolpidem 5mg/day PO.

First, left femoral and lateral cutaneous nerve blocks were performed using ultrasound-guided single-shot injections of ropivacaine 0.375% (15 and 5 ml, respectively). Then, spinal anaesthesia was accomplished in a sitting position using isobaric bupivacaine 7 mg and sufentanil 2.5 mcg; three attempts were needed due to abnormal anatomical curvatures of the spine and back muscles rigidity. Propofol (10 mg) IV intermittent boluses were administered for moderate sedation given that the patient had cognitive impairment and kept moving. A total of 40 mg was needed. Surgical duration was 2 h and 30 min, during which we encountered four episodes of hypotension (two of which had occurred between spinal anaesthesia and surgical incision) despite optimal fluid intake. We used 1 l of crystalloids, one packed red blood cells unit to compensate blood losses, and phenylephrine boluses with a total of 150 µg. After 4 h of observation in the postanaesthesia care unit, the patient was returned to the ward uneventfully. Lower limbs neurological examination returned to baseline 3.5 h after spinal anaesthesia, and no prolonged effect of anaesthetics was noted.

Disadvantages of locoregional anaesthesia in PSP patients include: (1) technical difficulties due to tremor, muscular rigidity, and cognitive impairment, (2) unsuitability for the selected surgery and (3) lack of patients’ immobility. If sedation is required, it may be better to titrate short acting drugs using a start low and go slow approach; propofol seems adequate. Induced haemodynamic instability is another aspect to consider. PSP-associated dysautonomia\(^1\) can lead to exaggerated response to neuraxial blockade, and drugs such as levodopa may precipitate hypotension by causing peripheral vasodilation.\(^2\) Whenever possible, isolated peripheral nerve blocks are ideal for preventing hypotension. Low-dose spinal anaesthesia (in combination with peripheral nerve blockade\(^3\)), intravenous fluids infusion, lower legs compression, and vasopressors can be used to prevent and manage spinal anaesthesia-induced hypotension.

In case of unfeasible locoregional anaesthesia, different facets of PSP should be considered before general anaesthesia. First, severe dystonia limits cervical mobility, and retrocollis is estimated to occur in 25% of cases.\(^4\) In some cases, trismus\(^5\) and laryngeal spasm\(^6\) may cause upper airway obstruction. One PSP patient was tracheotomised after unsuccessful tracheal intubation attempts due to severe neck deformity and tracheal stenosis.\(^7\) We, therefore, suggest preplanning for difficult airway management.

Second, PSP patients commonly manifest spastic dysphagia,\(^1\) putting them at risk of aspiration pneumonia after tracheal extubation. Rapid and complete emergence from anaesthesia is hence favorable. Monitoring the bispectral index ensures avoiding excessive sedation.

Third, PSP patients are often polymedicated with antiparkinson or antidepressant drugs that are known to have significant interactions with anaesthesia.\(^2\) It is, therefore, wise to use the least amounts of anaesthetics. As for neuromuscular blocking agents, Sakai and Sumikawa reported that the effect of vecuronium was not prolonged.\(^7\) However, neuromuscular transmission measurement was difficult because of limb contractures. None have reported the use of succinylcholine on such patients, although it is not contraindicated in Parkinson’s disease.\(^8\) Finally, there is no evidence in the literature that anaesthetic drugs exacerbate PSP symptoms.\(^9\)
Letters to Editor

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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There are no conflicts of interest.

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