Anaesthesia management in anti-synthetase syndrome, a rare orphan disease – A case study

Dear Editor,

Anti-synthetase syndrome (ASS) is a rare orphan autoimmune condition marked by interstitial lung disease (ILD), polyarthralgia, myopathy, skin hyperkeratosis and the presence of autoantibodies against aminoacyl-t ribonucleic acid (RNA) synthetases. The prevalence of ASS has been reported to be 1.5 per 100,000 population.[1]

A middle-aged female (weight- 64 kg, height- 156 cm), who was a known case of ASS for 5 years, was scheduled for revision total knee replacement (TKR). She had been treated with oral prednisolone (30 mg once daily [OD]) and azathioprine (100 mg OD). She had undergone right-sided TKR 4 years back, followed by implant removal and cement spacer insertion for the infected implant under spinal anaesthesia 3 years back. Pulmonary function testing revealed a forced vital capacity (FVC) of 66% and forced expiratory volume in the first second of 74% predicted, suggesting mild restrictive disorder. The patient was educated about energy conservation, reconditioning exercises and breathing techniques. She was advised to continue her medications till the day of surgery, and written informed consent was taken. Intraoperatively, intravenous hydrocortisone (100 mg) was administered, followed by spinal anaesthesia using hyperbaric bupivacaine (12.5 mg) and fentanyl (25 μg).

A T8 spinal level was achieved, and the surgery was uneventful. The patient was advised to continue incentive spirometry and reconditioning exercises postoperatively. Twenty days later, the patient developed a septic knee, for which gastrocnemius flap coverage and debridement were performed under spinal anaesthesia.

Autoantibodies against aminoacyl-tRNA synthetases, such as anti-Jo-1, anti-PL-12, anti-EJ anti-Ro, anti-La and anti-double-stranded deoxyribonucleic acid (dsDNA), are the commonly detected antibodies in ASS.[2] The beginning of pulmonary symptoms indicates an advanced disease with 5-year mortality of up to 45%.[3,4] The best predictors of restrictive spirometry patterns in ILD in ASS are FVC <80% predicted and diffusing capacity of the lungs for carbon monoxide <70% of the predicted value.[5] Regional anaesthesia is a commonly used anaesthesia technique for these patients as ILD patients often have limited respiratory reserve. However, high neuraxial technique in ILD patients may exacerbate the underlying pulmonary pathology by changes in intrathoracic pressure and alter the distribution of pulmonary blood flow, resulting in reduced inspiratory capacity and expiratory reserve volume.[4] Postoperative pulmonary complications such as acute lung injury, pneumonia, atelectasis, pneumothorax, pulmonary embolism and ventilator-induced lung injury are all risks associated with general anaesthesia, increasing morbidity and mortality.[4] A stress dose of steroids must be provided to lessen the risk of adrenal insufficiency in individuals receiving high-dose corticosteroids.

To conclude, sound knowledge and a multidisciplinary approach are required for the perioperative management of this rare syndrome.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient consented to her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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