Flail arm syndrome (FAS), also known as ‘man-in-the-barrel’ syndrome or brachial amyotrophic diplegia, is an atypical presentation of amyotrophic lateral sclerosis (ALS). It is differentiated from ALS by lower motor neuron type proximal muscle weakness of only the upper limbs. The lower limbs may show mild upper motor neuron type weakness, but the bulbar and respiratory muscles are not affected. It has a progressive course with a male-to-female ratio of 4 : 1 [1]. FAS is one of the spectra of motor neuron disease (MND) and shares many of the pathophysiological features of MND. MND is a degenerative disease of the spinal cord tracts and leads to respiratory muscle atrophy due to the ongoing, chronic process of denervation and re-innervation of muscles.

We describe the anesthetic management of a 50-year-old male (164 cm tall and 54 kg) that had FAS with bilateral proximal upper limb weakness and chronic obstructive respiratory disease. The patient was a chronic smoker. He had been recently diagnosed with an epigastric hernia and was scheduled for a mesh hernioplasty. The patient had gradually progressive and symmetrical weakness of his proximal muscles of both upper limbs, for 2 years, with no wasting or weakness in the distal muscles. His symptoms had worsened over the previous 6 months. There was severe wasting of the bilateral shoulder and arm muscles, with the power grading being 2/5. The patient’s neurophysiological assessment was normal. A muscle biopsy suggested neurogenic atrophy. Pulmonary function assessment indicated moderate obstruction on spirometry. The patient was at risk of acute respiratory failure due to his respiratory status and the general aspiration risk of MND patients. Hence, we decided to adopt a transverse abdominis plane (TAP) block as our first choice of anesthesia. If the block failed, we were prepared to administer total intravenous anesthesia (TIVA) with propofol and dexmedetomidine without a muscle relaxant. Bronchodilator nebulization and anti-aspiration prophylaxis were done in the preoperative preparation. Under all aseptic precautions, the patient was given an ultrasound-guided bilateral subcostal TAP block, using 15 ml of 0.5% ropivacaine on either side. The patient was also given mild sedation with dexmedetomidine bolus of 50 μg over 10 minutes, followed by 25 μg/h. The total duration of the procedure was ninety minutes. Intra-operative management was uneventful, and the patient was shifted to the postoperative anesthesia unit with stable hemodynamics and respiratory status. There were no adverse effects on the patient’s vitals, respiratory depression, sedation, and muscle weakness observed during the postoperative period.

There is a lack of literature in the anesthetic management of patients with FAS. In FAS patients, we have the same anesthetic considerations as those with ALS or MND. Neuromuscular monitoring and the need for postoperative mechanical ventilation must be anticipated where muscle relaxants cannot be avoided [2]. Depolarizing muscle relaxants such as succinylcholine are not recommended because of reported rhabdomyolysis and...
hyperkalemia from denervated muscles, possibly leading to ventricular arrhythmias and fibrillation. Their use in patients with ALS has also been associated with neuro-myotonia. Inhalational agents are known to potentiate a neuromuscular blockade as they possess intrinsic neuromuscular blockade activity. Regional anesthesia is preferred in patients with MND to avoid aspiration and the possibility of a prolonged neuromuscular blockade following general anesthesia. However, the use of a central neuraxial blockade may also exacerbate the progression of the disease course. Demyelination of the nerve fibers makes the patients more susceptible to the neurotoxic effects of local anesthesia [3]. Therefore, the preferred anesthetic technique in these patients is TIVA or regional anesthesia. Thampi et al. [4] described the use of TIVA with an obturator nerve block in the successful anesthetic management of a patient with ALS for transurethral resection of the prostate. The TAP block targets the myocutaneous nerves to the anterior abdominal wall (T6 to L1). The subcostal approach blocks the sensory nerves from T6 to T10, which is sufficient for surgeries with an incision above the umbilicus. The major advantages that a subcostal TAP block offers over general anesthesia include a lower opioid requirement, decreased incidence of postoperative respiratory depression, nausea, and vomiting. As compared to central neuraxial blocks, the advantages include the absence of a sympathetic block and avoidance of disease progression. The primary challenge in the anesthetic management of patients with FAS is to guarantee optimal surgical conditions while preserving the perioperative neuromuscular and respiratory function of the patient close to the pre-operative baseline status. Mild sedation and a bilateral subcostal TAP block ensured adequate anesthesia, analgesia, and immobility. Besides, we were able to efficiently manage the case while avoiding the aggravation of neuromuscular weakness and aspiration. To the best of our knowledge, this is the first safe use of dexmedetomidine along with a TAP block in an MND case.

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Conflicts of Interest

No potential conflict of interest relevant to this article was reported.

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References