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Anesthesia management in a case of Turner syndrome with anti-NMDA limbic encephalitis and multiple co-morbidities for repair of fracture femur

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We present the case of a 65-year-old female, a known case of Turner syndrome with multiple co-morbidities, namely, long-standing coronary triple vessel disease, diabetes, hypertension, hypothyroidism, gastro-esophageal reflux disease, psychiatric illness, severe osteoporosis, and severe sensorineural hearing loss. She was scheduled to undergo repair of an intertrochanteric fracture of the femur. Evaluation of her symptoms revealed episodes of autonomic dysfunction in relation to stressful events and illness, characterized by hypotension and tachycardia, which were responsive to fluids. Her medications at the time included clopidogrel, ecosprin, ramipril, metoprolol, atorvastatin, quetiapine, zoledronic acid, vitamin D3, prednisolone, L-thyroxine, metformin, and multivitamins. She was admitted to the neurology department with complaints of hallucinations, memory disturbances, and agitation. During the course of her hospital stay, she experienced a fall in the bathroom that resulted in a fracture of the right femur. Her cerebrospinal fluid examination was within normal limits; magnetic resonance imaging of the brain revealed features suggestive of limbic encephalitis, and infectious etiologies and other possible causes of encephalitis were ruled out. Considering her background of Turner syndrome, an autoimmune workup was done. She was found to have anti-N-methyl D-aspartate receptor (NMDA) receptor limbic encephalitis; treatment was started with steroids and intravenous immunoglobulin therapy. Subsequently, she demonstrated marked improvement in symptomatology.

On examination, she was found to be short-statured (140 cm), weighing 48 kg, with webbed neck, and webbed toes. Her heart rate was 88 beats per minute, blood pressure 100/60 mmHg, respiratory rate 14 cycles per min, and peripheral oxygen saturation 97%. Auscultation of her chest revealed diffusely reduced air entry, particularly in the lung bases. Her biochemical investigations revealed the following: hemoglobin 11.2 g/dl and platelet count 220×10^9/L with high titers of anti-thyroid peroxidase, anti-thyroglobulin, and anti-NMDA antibodies; the results of other investigations (liver and kidney function tests, serum electrolytes etc.) were grossly within normal limits. High resolution computed tomography of her thorax suggested bilateral pleural effusion with adjacent subsegmental lung collapse and moderate pericardial effusion. The condition of the patient was explained to the patient's relatives, and written informed consent was obtained. Clopidogrel was stopped seven days prior to surgery, and ramipril and metformin were stopped on the day of surgery.

Turner syndrome is a chromosomal disorder with XO genotype and affects females. Phenotypically, it is associated with short stature and multiple autoimmune diseases such as type 1 diabetes mellitus, Hashimoto's thyroiditis, ulcerative colitis, rheumatoid arthritis, and Addison's disease. Limbic encephalitis

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is characterized by subacute impairment of memory, psychotic behavior, and seizure disorder; autoimmune anti-NMDA encephalitis is a rare entity that has been described since 2007 [1]. Anti-NMDA encephalitis exclusively involves limbic regions and is characterized by psychotic behavioral changes, autonomic dysfunction, and movement disorders [1]. There is a paucity of literature on the anesthetic concerns in a case of autoimmune limbic encephalitis; many authors have recommended general anesthesia [2,3], and we could find only one case in which authors had given a transverse abdominis plane block [4]. Most general anesthetics, including the volatile anesthetics, act via the NMDA receptor. NMDA antagonists like ketamine, nitrous oxide, xenon, tramadol, methadone, and phencyclidine are avoided in this condition because their effects are unpredictable and may worsen the course of the disease [2-4]. Pascual-Ramírez et al. have used a combination of propofol, sevoflurane, and fentanyl in a case with no complications [2]. Although propofol is considered a safe drug as it does not act via NMDA receptors, and a few authors have used it with no complications, some case reports document severe unexpected hypotension in the patient on using propofol induction [5]. Therefore, a single drug cannot be labeled as safe for use in limbic encephalitis. We did not prefer general anesthesia owing to the risk of aspiration, gastro-esophageal reflux disease, known theoretical atypical response to volatile agents and induction drugs, delay in the reversal of airway reflexes, and postoperative delirium. Moreover, the patient already had impaired lung function, which could have led to post-operative pulmonary complications, and ventilator support may have been required in the postoperative period. Spinal anesthesia alone was also not safe owing to the risk of a high level of blockade and autonomic instability; therefore, we preferred combined low dose spinal-epidural anesthesia, so as to titrate the level of blockade and prevent profound hypotension. Hemodynamic instability was expected owing to autonomic instability and associated anti-limbic NMDA encephalitis, and we preferred fluid bolus and dobutamine because of her triple vessel disease in order to maintain adequate cardiac output.

Management of patients having Turner syndrome with multiple co-morbidities and associated anti-NMDA limbic encephalitis is a challenge for attending anesthesiologists. These patients are at risk of perioperative complications such as aspiration, post-operative respiratory complications, and delirium, and may show abnormal responses to general anesthetic agents. Combined spinal-epidural anesthesia is a safe alternative to spinal and general anesthesia in these patients, but one must carefully maintain the level of blockade and the hemodynamics.

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References

1. Dalmau J, Gleichman AJ, Hughes EG, Rossi JE, Peng X, Lai M, et al. Anti-NMDA-receptor encephalitis: case series and analysis of the effects of antibodies. Lancet Neurol 2008; 7: 1091-8.
2. Pascual-Ramírez J, Muñoz-Torrero JJ, Bacci L, Trujillo SG, García-Serrano N. Anesthetic management of ovarian teratoma excision associated with anti-N-methyl-D-aspartate receptor encephalitis. Int J Gynaecol Obstet 2011; 115: 291-2.
3. Liao Z, Jiang X, Ni J. Anesthesia management of cesarean section in parturient with anti-N-methyl-D-aspartate receptor encephalitis: a case report. J Anesth 2017; 31: 282-5.
4. Chaw SH, Foo LL, Chan L, Wong KK, Abdullah S, Lim BK. Anesthesia in anti-N-methyl-d-aspartate receptor encephalitis - is general anesthesia a requisite? A case report. Rev Bras Anestesiol 2017; 67: 647-50.
5. Splinter WM, Eipe N. Anti-NMDA receptor antibodies encephalitis. Paediatr Anaesth 2009; 19: 911-3.