Management of a parturient with Sjogren’s syndrome and its associated complications undergoing emergency caesarean section

Sir,

Sjogren’s syndrome is an autoimmune disorder,[1] characterised by the presence of dry eyes (keratoconjunctivitis sicca) and dry mouth (xerostomia) as a consequence of lymphocytic infiltration into the lacrimal and salivary glands and may present as a multisystem involvement. We present a case of a parturient with incidental finding of neonatal bradycardia leading to the diagnosis of Sjogren’s syndrome with renal involvement, who presented for emergency caesarean section.

A 42-year-old pregnant [G5P3, 37 + 2 weeks gestational age] female patient [weight 86 kg, height 167 cms; body mass index (BMI) 30.8 kg/m²] presented for emergency caesarean section for non-reassuring foetal status. She was diagnosed with Sjogren’s syndrome about 3 months before admission. Her foetus was found to have congenital heart block, and this led to maternal autoimmune evaluation. She showed anti-Ro, anti-La, and anti-ds-DNA antibodies with negative rheumatoid factor, which suggested the diagnosis of Sjogren’s syndrome.

The patient developed severe symptomatic hypokalaemia in response to intravenous dexamethasone given for foetal lung maturity, which manifested as proximal muscle weakness as per physician’s examination. Concurrently, she was found to have renal tubular acidosis with nephrocalcinosis, and developed rhabdomyolysis diagnosed by raised creatinine kinase enzyme. Potassium replacement was started to treat hypokalaemia and sodium bicarbonate for correcting acidosis. There was no clinical neurological impairment at the time of surgery.

We explained the anaesthesia plan for spinal anaesthesia with informed consent. Spinal anaesthesia was performed under strict aseptic technique. A male baby boy was born after 8 minutes of skin incision with an APGAR score of 8 at 1 and 5 minutes. The neonate was found to have a heart rate of 50 beats/min and peripheral oxygen saturation (SpO₂) 92% with free-flow of oxygen and did not require intubation. The neonate was transferred to the neonatal intensive care unit (NICU) for further management. Estimated blood loss was about 500 ml. Her post-operative investigations were unremarkable. She was given an appointment with renal medicine and rheumatology for her follow-up after discharge. Neonatal bradycardia warranted placement of a cardiac pacemaker.

Sjogren’s syndrome is an autoimmune disease that, if not well controlled in preconception, has the potential to deteriorate during pregnancy. The obstetric patient with Sjogren’s syndrome is at risk of worsening renal function, disease relapse, and foetal loss. Sjogren’s syndrome also predisposes the foetus to adverse perinatal outcomes, such as congenital heart block, growth restriction, and...
Sjogren's syndrome is characterized by the presence of a plethora of autoantibodies. Of the various autoantibodies present in a patient with Sjogren's syndrome, Anti Ro/SSA and Anti La/SSB are the most common antibodies present in 50-70% of the patients. Patients may not have significant symptoms and are only brought to medical attention solely on the identification of foetal heart block or rash in a neonate. Hence, the presence of foetal congenital heart block should prompt further maternal autoimmune disorder evaluation.

Our patient was found to have renal tubular acidosis diagnosed upon medical review for further evaluation of significant maternal hypokalaemia. Investigations showed a positive urine anion gap with nephrocalcinosis and rhabdomyolysis suggestive distal renal tubular acidosis. Only minority of the patients may have severe manifestations of proximal renal tubular acidosis (RTA), even presenting with profound hypokalaemia, as in this patient. The therapy of type 2 RTA should be instituted including potassium replacement and sodium bicarbonate treatment. Our patient developed severe hypokalaemia (2.5-2.8 mEq/L) which needed urgent intravenous potassium replacement. Exaggeration of hypokalaemia could also be due to dexamethasone therapy for foetal lung maturation.

Patients with lung involvement may be at high risk for general anaesthesia. Involvement of central nervous systems warrants thorough preoperative neurological examination. With neonatal bradycardia, consideration shall be given to reduce the potential effect of general anaesthesia on new-born by using spinal anaesthesia. Similarly, with maternal hypokalaemia and rhabdomyolysis, choice of spinal anaesthesia can be considered to reduce potential neuromuscular weakness from use of neuromuscular blockade with general anaesthesia.

The choice of anaesthetic technique should be decided considering the urgency of the caesarean delivery and balancing both maternal and foetal safety.

Sjogren's syndrome in obstetrics can be challenging. Anaesthesiologists should be aware of the multiple system involvement syndrome and associated difficulties during perioperative care.

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

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Conflicts of interest
There are no conflicts of interest.

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