Perioperative management of emergency caesarean section in type I hereditary angioedema

Dear Editor,

Hereditary angioneurotic edema (HAE) is a rare autosomal dominant inherited disorder caused by mutation in the gene encoding C1 esterase inhibitor (C1-INH). In type I HAE, low levels of C1-INH leads to unchecked activation of classical complement pathway leading to overproduction and accumulation of vasoactive peptides resulting in vasodilation, smooth muscle contraction and tissue edema. Though it can affect any site, HAE more commonly causes angioedema of airway, gastrointestinal tract, extremities, face and trunk during an acute attack.[1]

We present a 26-year-old primigravida with 39 weeks gestational period, in active labour with cephalopelvic disproportion scheduled for emergency caesarean section in view of obstetric indication. History revealed that she was diagnosed to have HAE one year ago with initial attacks of angioedema started at 3 years of age. Frequency of attacks were 5-6 episodes per year with two episodes of laryngeal edema. Precipitating factors of angioedema in this patient were pain, trivial trauma and cold weather. The recent C1-INH level was found to be less than 0.028 g/l (normal levels 0.21–0.39 g/l), which was done a year ago and her systemic examination was normal. A multidisciplinary approach was planned to involve the obstetrician, otolaryngologist, neonatologist. Regional anaesthesia with combined spinal epidural block was planned and informed written consent was obtained from the patient. We also obtained consent for the possible publication of the case in future. Anticipating a difficult airway in the event of an acute attack, we kept smaller sized endotracheal tubes, bougie, video laryngoscope and tracheostomy tubes. We transfused two units of FFP preoperatively as a short-term prophylaxis. Combined spinal epidural block was performed in the L2-L3 inter vertebral space and intrathecal injection with 1.8 ml of 0.5% hyperbaric bupivacaine was given. Postoperatively, patient received epidural infusion of 0.125% bupivacaine at 5 ml/hr for 48 hours in intensive care unit. The course of HAE during pregnancy can be unpredictable with either abdominal symptoms or vulval edema as a presenting feature.[2] Though observational studies show that C1-INH concentrates have been successful in preventing acute attacks of HAE, unavailability is the limiting factor.[3] FFP can be used both for prophylaxis and treatment of acute attacks, if the first line drugs such as C1-INH concentrates and Icatibant (bradykinin antagonist) are unavailable.[4] Regional anaesthesia obviates the need for airway manipulation caused by laryngoscopy and intubation, thereby avoiding a localised triggering factor for an acute attack of laryngeal edema in these patients.[5] However, endotracheal intubation should be considered when other determinants such as patient related and surgical factors necessitates. In our patient, an aggressive management of postoperative pain with epidural analgesia helped in preventing an episode of angioedema peri-operatively, since pain is a known precipitating factor.

Surgical pain and airway manipulation are known triggering factors in HAE patients during the perioperative period and hence effective management of pain is vital in preventing an attack of HAE perioperatively. Combined spinal epidural anaesthesia can be an effective tool in the management of caesarean section in HAE patients by mitigating these two known risk factors.

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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Nil.

Conflicts of interest

There are no conflicts of interest.

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