Inadequate spinal anesthesia in a parturient with Marfan's syndrome due to dural ectasia

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Marfan syndrome (MFS) is a rare hereditary connective tissue disorder that affects the cardiovascular, musculoskeletal, and ocular systems. Pregnancy further increases the potential for cardiovascular risks due to increases in blood volume, heart rate and stroke volume. The choice of anesthetic technique is very important in patients with MFS. Dural ectasia (DE) in MFS patients may result in failure of spinal anesthesia. We reported a patient without typical symptoms related to DE who experienced spinal anesthesia failure.

A-29-year-old female (weight, 58 kg; height, 161 cm) with known MFS was admitted for cesarean delivery at 37 + 6 weeks of gestational age. She had valve sparing aortic root replacement surgery due to aortic root aneurysm and was diagnosed with MFS 6 years ago. Preoperative echocardiography revealed a left ventricular ejection fraction of 68% with moderate aortic regurgitation (G2-3), trivial tricuspid regurgitation and intact aortic root graft. Aortic regurgitation had increased since the previous study conducted 6 months earlier. The patient was medicated with atenolol during pregnancy.

Elective cesarean section was scheduled and routine monitoring devices were applied in the operating room. The left radial artery was cannulated for continuous monitoring of arterial pressure. The initial blood pressure was 140/73 mmHg, heart rate was 74 beats /min, and peripheral oxygen saturation was 100% at room air. Combined spinal-epidural (CSE) anesthesia was administered. The epidural space was found using a loss of resistance technique at the first attempt. Clear cerebrospinal fluid (CSF) was obtained on spinal needle insertion. There was no paresthesia. Eight mg of 0.5% hyperbaric bupivacaine and 10 ug

of fentanyl was injected intrathecally. An epidural catheter was inserted without resistance and advanced 5 cm upward. The levels of sensory block were tested by alcohol swabs and pinprick tests. Ten minutes following the intrathecal injection, the patient had only limited lower limb analgesia. The epidural injection was titrated over the next 20 min, and 8 ml of 2% lidocaine and 8 ml of 0.75% ropivacaine were required to achieve T4 sensory block.

The remainder of the procedure was uneventful. Ephedrine 4 mg IV was administered twice to maintain systolic blood pressure above 100 mm Hg and the patient was sedated with midazolam after delivery. The patient's postoperative vital signs were stable with a blood pressure of 111/54 mmHg, a heart rate of 68 beats/min, and an oxygen saturation of 98%. The postoperative pain was managed with patient-controlled epidural analgesia. She had an uneventful postoperative course and was discharged 4 days later.

DE is defined as 1) an enlarged neural canal along the spinal column, usually in the lower lumbar and sacral regions; 2) a thinning of the cortex of the pedicles and lamina of the vertebra; 3) a widening of the neural foramina; or 4) an anterior meningocele [1]. A more recent definition of DE is a widening of the dural sac or spinal nerve root sleeves. The most common clinical symptoms include low backpain, headache, weakness, and loss of sensation above and below the affected limb, bowel and bladder dysfunction, occasional rectal pain and pain in the genital area [2]. The incidence of DE in MFS patients reportedly ranges from 63% to 92% [3].

The associated increase in CSF volume due to DE, and the

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erratic spread of spinal anesthetic is thought to increase rate of spinal anesthesia failure in the MFS parturient [2]. A thorough reevaluation of patient DE symptoms was done before anesthesia. Our patient had no common clinical symptoms of DE, hence we expected success in spinal anesthesia. CSE anesthesia was performed in order to provide postoperative pain control, as well as epidural anesthesia in case of spinal anesthesia failure. Contrary to our expectations, spinal anesthesia failed to produce a block adequate for surgical procedure. Foran et al. [4] characterized DE in MFS patients and reported that such patients are usually asymptomatic. The severity of DE can only be radiologically evaluated by computed tomography or MRI, by assessing dural sac diameter, nerve root sleeve diameter, and lumbar pedicle width [3]. Despite lack of symptoms associated with DE the presence of DE could not be ruled out. The prevalence of DE is high among MFS patients, hence probable DE in the study patient resulted in failure of spinal anesthesia [3].

Baghirzada et al. [5] reported 2 cases of regional anesthesia in parturients with MFS with conflicting spinal anesthesia results. The success of spinal anesthesia differed based on the severity of the DE. Unless a patient undergoes radiologic examination for the presence and severity of DE before surgery, it is not possible to predict the success of spinal anesthesia in MFS patients. Epidural anesthesia provides a gradual titration of local anesthetics that ensures adequate post-operative pain control while minimizing potential hypotension caused by local anestheticinduced sympathectomy. Epidural anesthesia provides more stable hemodynamics in MFS patients with cardiovascular complications.

We recommend the CSE technique or epidural anesthesia for MFS patients regardless of the presence of DE related symptoms, due to its high incidence among MFS patients and often asymptomatic occurrence.

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