

Successful use of spinal anesthesia for an urgent cesarean section in a parturient with a severe Klippel-Trénaunay syndrome

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Klippel-Trénaunay syndrome (KTS) is a congenital malformation with an incidence of 1 out of 27,500 live births described in 1900 including a triad of port-wine stain/capillary vascular malformation, venous malformation/varicose veins, soft tissue, and bony hypertrophy in affected limbs [1]. Although a combined spinal-epidural technique for an elective cesarean section and an epidural analgesia for vaginal delivery have been already presented [2,3], this is the first report that describes spinal anesthesia for an urgent cesarean section in a patient with KTS.

A 23-year-old woman (gravidity: 1, parity: 0) was referred to our Anesthesia Preadmission Clinic at 37 weeks of gestation because of KTS complicating her pregnancy. She weighed 115 kg and was 170 cm in height (body mass index 39.8 kg/m²). She had been diagnosed with KTS at the age of 16 and received several surgical treatments for her right foot varicosities and opioid therapy till the beginning of gestation because of pain in the right leg. She did not have a history of thrombosis or hemorrhage. On hospital admission at 37 weeks of gestation for planned delivery, physical examination showed prominent hypertrophy and multiple varicosities of the right leg (Fig. 1). The circumferences of the thigh, calf, ankle, and knee were 12, 15,

11, and 10 cm, respectively, larger than the left side. Laboratory studies revealed a normal coagulation profile and hemoglobin (Hb) count of 8.5 g/dl. She was scheduled for a magnetic resonance imaging (MRI) scan to determine the existence of arterio-venous malformations (AVM) or hemangiomas in the pelvis, birth canal, spinal cord, bronchial tube, and brain before delivery, but she had not yet undergone MRI. She was admitted to our delivery unit at 38 weeks of gestation for an urgent cesarean section due to abnormal cardiotocography (type 2 urgency according to Lucas' classification). On admission, she was anxious and had breakfast two hours before. On physical examination, her airway revealed a Mallampati Class III and her back had normal anatomy with clearly palpable landmarks with no evidence of port-wine stains. Ultrasound revealed no signs of detectable vascular abnormalities. She refused any attempt



Fig. 1. Detail of the patient's affected leg.

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of awake intubation. MRI of lumbar spine performed six years ago showed no AVM or hemangioma. After discussing rapidly the risks and benefits of general and spinal anesthesia, we decided that spinal anesthesia was safer in this case. However, we informed her the possibility of intraoperative conversion to general anesthesia in case significant hemorrhage occurred during the operation. Two 16-gauge intravenous catheters were placed in the left antecubital fossa bilaterally. Aspiration and antibiotic prophylaxis were performed routinely. In the meantime, an intravenous bolus of 0.9% saline 500 ml was administered before neuraxial anesthesia. Monitors included an electrocardiogram, finger pulse oximetry, and noninvasive blood pressure. In sitting position, spinal anesthesia at L₃₋₄ intervertebral space was performed with 5 µg of sufentanil and 10 mg of hyperbaric bupivacaine. Then, the patient was positioned in the supine position with uterus displacement and her legs were lifted 30 degrees. A T4 anesthetic block was obtained without developing hypotension after neuraxial block. Her systolic blood pressure remained greater than 100 mmHg. A female baby weighing 2,964 g was delivered; her Apgar scores were 8 and 9 at 1 and 5 min, respectively. Abnormal bleeding vessels were noted around the uterine incision during the operation. The estimated blood loss was about 2,000 ml and two units of packed red blood cells were required. Two hours after surgery the Hb count was 8.4 g/dl and coagulation profile was normal. The rest of patient's postoperative course was unremarkable. To prevent thromboembolic disease, low-dose heparin (6,000 U/day) injection was administered for three postoperative days. The patient was discharged with her baby in good health.

A planned anesthesia for a KTS patient is challenging and with due emphasis on airway, hemodynamics, and neuraxial vascular malformation bleeding can result in a favorable outcome. Airway management in such patients may be difficult owing to the soft tissue hypertrophy characteristic of pregnancy and possible underestimated hemangiomas typical of KTS. Hemorrhagic complications may arise as a result of pelvic varicosities being injured by surgery [4]. A whole body MRI is recommended before performing a cesarean section to prevent

hemorrhage complications and the anesthesiologist should pay attention to detect abnormal vessels of bronchial tube and lumbar spine [5]. In this case, an MRI was scheduled, but not executed yet. An MRI of the lumbar spine of six years before was negative for abnormal vessels around the spinal cord. Moreover, considering that an accurate examination of the spine and ultrasound scan excluded detectable central nervous system hemangiomas, we decided for a spinal anesthesia. Two more issues have influenced the choice of neuraxial technique: preoperative fasting recommendations were not respected because of urgency and the Mallampati class 3 airway revealed a probable difficult airway management. Given these concerns and a normal coagulation profile, a spinal anesthesia was executed assuming that a potential risk of difficult airway management and pulmonary aspiration were greater than possible complications correlated to the less probable presence of abnormal vessels near the spine. Another crucial point was the vasodilatation of the right leg varicosities after spinal anesthesia and its potential impact on hemodynamics, which could result in decreased venous return. The lifting of the leg by approximately thirty degrees increased venous return releasing the amount of blood eventually seized in the varicosities. A significant intraoperative blood loss should be expected and at least one unit of packed red blood cells should be readily available to be transfused as has been done in this case. The optimal management of a non-urgent scenario is to perform a neuraxial blockade only after having ascertained a negative spine MRI finding. In case of urgent cesarean section, the risks and benefits of each anesthesia technique must be compared.

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