Case Report

Spinal Anaesthesia for Cesarean Section in a Patient with Vascular Type Ehlers-Danlos Syndrome

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We report the administration of spinal anaesthesia for cesarean delivery in a parturient with vascular Ehlers-Danlos syndrome. Parturients who genetically inherit this disorder are at risk for significant morbidity and mortality. Risks during pregnancy include premature labor, uterine prolapse, and uterine rupture. Additionally, such laboring parturients are at increased risk of hemodynamic volatility, vascular stress, and severe postpartum hemorrhage. Instrumented delivery and cesarean delivery bring additional risks. Nonpregnancy-related complications include excessive bleeding, intestinal rupture, cardiac valvular dysfunction, and arterial dissection. Despite the complexity of this condition, literature focusing on specific intraoperative anaesthetic management is sparse.

1. Introduction

Ehlers-Danlos syndromes (EDS) comprise connective tissue disorders affecting multiple organ systems to include the integumentary, musculoskeletal, pulmonary, digestive, and cardiovascular systems. These disorders are classified according to the Villefranche Nosology which was originally adopted in 1997 [1]. To date, six underlying types of disease have been identified and are categorised as follows: classical (formerly type I/II), hypermobility (formerly type III), vascular (formerly type IV), kyphoscoliosis (formerly type V1), arthrochalasia (formerly type VIIA/VIIIB), and dermatosparaxis (formerly type VIIIC) [1]. Each type possesses unique clinical manifestations and anaesthetic implications. Peripartum complications associated with vascular EDS include preterm premature rupture of membranes, arterial dissectionrupture (e.g., aorta, iliac, splenic, and coronary), uterine rupture, uterine incision dehiscence, 3rd-/4th-degree lacerations, and postpartum hemorrhage [2]. Peripartum mortality rates are highly variable and range from 4.3% to 25% [2]. Case reports in the literature specifically focusing on the intraoperative anaesthetic management of these patients are sparse [3, 4]. Here, we report the perioperative anaesthetic management of a parturient with vascular EDS. The patient provided written permission for publication of the report.

2. Case Description

The patient was a 23-year-old female (Gravida 1, Para 0) presenting for obstetric care at eight weeks gestational age with genetic documentation of a COL3A1 mutation confirming vascular type EDS. She reported emergency room visits for significant hematomas in addition to current treatment with losartan for a history of cerebral aneurysm. First trimester brain MRI revealed full resolution of the aneurysm. Her second trimester transthoracic echocardiogram was unremarkable. A complete blood count, comprehensive electrolyte panel, and electrocardiogram were also unremarkable. Hemoglobin and hematocrit concentrations were 9.2 g⋅dl−1 and 28.9%. Prothrombin time, partial thromboplastin time, and platelet function analysis were within normal limits. We planned for surgical delivery at 34 + 0 weeks of gestational age. This gestational age represented a balance of fetal risk due to premature delivery and maternal risk due to increasing fetal size and an active second stage of labor. We elected for an
operative hemoglobin and hematocrit concentrations were
cesarean section. Blood loss was 700 ml and the patient's post-
bladder catheterization and subsequent uncomplicated
ultrasound guidance. Spinal level was verified at T6 prior
epihelinrine, $30 \mu g\cdot kg^{-1}$ of clonidine, and $100 \mu g$ of morphine were
injected. She was then placed in the supine position and the
femoral vein was cannulated with a triple lumen catheter under
ultrasound guidance. Spinal level was verified at T6 prior
to bladder catheterization and subsequent uncomplicated cesarean section. Blood loss was 700 ml and the patient's post-
operative hemoglobin and hematocrit concentrations were
7.2 g dl$^{-1}$ and 22.9%. Twenty units oxytocin in 1-liter normal saline was given via IV infusion. Following the procedure, the patient was transferred to the intensive care unit to utilise the combined resources of obstetric and critical care nursing. Hourly neurologic checks were performed to monitor for symptoms of a neuraxial hematoma. Maternal postoperative pain management was dictated by the surgeon and included the use of a patient-controlled analgesic device. Maternal complications were limited to an ileus requiring nasogastric tube placement and a brief extension of her hospital stay. The ileus resolved without complication permitting discharge to home within one week of surgery. During phone follow-up two weeks later, the patient reported satisfaction with her anaesthetic and denied any complaints or concerns.

3. Discussion

Vascular EDS is a single subtype of a heterogeneous collection of connective tissue disorders associated with mutations in the genes that code for the production of collagen. Vascular EDS is associated with mutations in type III procollagen (COL3A1; OMIM #120180). The diagnosis, which carries a significant risk of peripartum morbidity, requires both genetic identification and the presence of at least two of the following major clinical criteria: “(1) arterial, gastrointestinal or uterine fragility or rupture, (2) thin, translucent skin without hyper-elasticity, (3) extensive bruising, or (4) characteristic facial appearance (thin lips and nose, hollow cheeks, large eyes, small chin)” [1]. Vascular EDS represents less than 10% of all EDS patients and carries a relatively low prevalence of 1:100,000–1:200,000 patients [5]. As such, few case reports have addressed the anaesthetic management of parturients with vascular EDS [3, 4]. In this context, we reviewed a publication which reported the management of a vascular EDS patient undergoing elective cesarean section at 36 weeks of gestational age [4]. Coagulation studies, complete blood count, and transthoracic echocardiogram were obtained preoperatively. Vascular access included two large-bore IV catheters, a radial arterial catheter, and an antecubital central venous catheter. Volume preloading consisted of 1-liter crystalloid and 0.5-liter colloid solutions. Tuohy needle-guided spinal anaesthesia was established by subarachnoid injection of 2.8 ml of 0.5% bupivacaine via a 25-gauge Whitacre spinal needle. An epidural catheter was inserted for postoperative analgesia.

Another report described successful continuous epidural anaesthesia with ropivacaine and fingal administered via a 20-gauge polyurethane catheter for labor and forceps delivery in a patient with vascular EDS [3]. In the absence of further case reports specific to the anaesthetic management of vascular type EDS parturients, we referenced an algorithm by Wiesmann et al. suggesting a preoperative approach to the EDS patient [6]. Specifically, we confirmed the genetic subtype, discussed prior EDS complications, and confirmed we had prolonged postoperative care facilities available. We focused extra care on patient positioning, cross-matched the patient for blood products, and administered desmopressin intraoperatively due to the patient's history of recurrent hematomas. We pursued a multidisciplinary approach and were influenced by these preoperative recommendations. We further obtained a first trimester brain MRI to evaluate for cerebral aneurysm and a second trimester transcranial echocardiogram to evaluate for aortic root widening, aortic aneurysm, or valvular pathology. We noted several factors in considering the intraoperative approach to her cesarean delivery. Risks of general anaesthesia include the difficulties of tracheal intubation and the potential complications encountered in vascular EDS patients (i.e., arterial dissection/cerebral hemorrhage during hypertensive response to intubation, possible pneumothorax from positive pressure ventilation, and unstable cervical spine with potential for atlantoaxial subluxation) [3, 7, 8]. Alternatively, neuraxial anaesthesia is potentially less consistently reliable and may increase the risk of epidural hematoma [7]. Though the majority of vascular EDS patients have normal coagulation, there is a tendency toward prolonged bleeding and platelet dysfunction [9]. Furthermore, the vascular fragility combined with the tendency for platelet dysfunction, and in this
The study was carried out at Department of Anesthesiology, Naval Medical Center Portsmouth. The views expressed in the manuscript are those of the authors and do not reflect the official policy or position of the Department of the Navy, Department of Defense, or the Unites States Government. This manuscript was screened for plagiarism and cryptomnesia using Grammarly. LCDR Carness and CAPT Lenart are military service members. This work was prepared as part of their official duties. Title 17 USC 105 provides that “copyright protection under this title is not available for any work of the United States Government.” Title 17 USC 101 defines a United States Government work as a work prepared by a military service member or employee of the United States Government as part of that person’s official duties.

Conflicts of Interest

The authors declare that there are no conflicts of interest and no external funding.

Authors’ Contributions

Jeffrey M. Carness and Mark J. Lenart conducted the manuscript compilation and dissemination.

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