Anesthetic management for cesarean section and tubal ligation in a patient with Marfan syndrome, multiple sclerosis, and multiple postdural puncture headaches

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ABSTRACT

We report a 29-year-old woman with Marfan syndrome, multiple sclerosis, and multiple postdural puncture headaches who presented for a scheduled repeat cesarean delivery with bilateral tubal ligation at 37 weeks gestation. During an outpatient preoperative visit, a general anesthetic plan was ultimately selected through a shared decision-making process. The patient had an uneventful general anesthetic that included a rapid sequence induction with direct laryngoscopy. Neonatal Apgar scores were 8 at 1 minute and 9 at 5 minutes. Prior to emergence, fentanyl, acetaminophen, and ketorolac were administered intravenously and a transversus abdominus plane block was performed. On the first postoperative day, the patient expressed satisfaction with the anesthetic plan and, in particular, the avoidance of a neuraxial technique and postdural puncture headache. The patient was discharged on the second postoperative day with no apparent sequelae. A neuraxial anesthetic technique is usually preferred in patients undergoing cesarean delivery, and it is safe to perform this technique in patients with either Marfan syndrome or multiple sclerosis. We formulated an anesthetic plan that honored our patient's autonomy and produced a good maternal and neonatal outcome.

KEYWORDS Anesthesia; cesarean section; Marfan syndrome; multiple sclerosis; postdural puncture headache; transversus abdominus plane block

Patients presenting for cesarean section occasionally have multiple comorbidities that have anesthetic implications. Marfan syndrome is a connective tissue disorder often accompanied by aortic root dilation and dural ectasia.¹ Multiple sclerosis is a demyelinating disease that has an unpredictable response to neuraxial anesthetic techniques.² Postdural puncture headaches are usually caused by unintentional puncture of the dura with a large bore needle and are treated with injection of autologous blood into the epidural space.³ Anesthesiologists in the USA tend to avoid general anesthesia for cesarean delivery in an attempt to mitigate maternal morbidity and mortality.⁴ We report a 29-year-old woman with a history of Marfan syndrome, multiple sclerosis, and postdural puncture headaches who had a planned general anesthetic for her scheduled repeat cesarean delivery with bilateral tubal ligation and a multimodal postoperative analgesia plan that included a transversus abdominus plane block prior to emergence.

CASE DESCRIPTION

A 29-year-old woman Gravida 2, Para 1 with Marfan syndrome, multiple sclerosis, and multiple postdural puncture headaches scheduled for repeat cesarean delivery and bilateral tubal ligation presented initially for outpatient preoperative consultation. Her previous delivery was at a US military hospital in Germany, where she apparently had an unintentional dural puncture with a Touhy needle and subsequently underwent cesarean delivery. She had a difficult postoperative course due to a postdural puncture headache and eventually received an epidural blood patch, which resolved her symptoms. Upon return to the USA, she received a lumbar puncture for evaluation of multiple sclerosis and again developed a postdural puncture headache that was treated with an epidural blood patch. She was 178 cm tall and weighed 88 kg; a head and neck exam revealed a Mallampati Class II airway and a thyromental distance of three fingerbreadths. The most recent echocardiogram from

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2 months prior revealed a mildly dilated aortic root at 4.1 cm. There was no lumbar spine magnetic resonance imaging.

A shared decision-making process with the patient was utilized to determine the anesthetic plan. The patient prioritized avoiding the discomfort of a neuraxial technique and the potential of a postdural puncture headache over recollection of her delivery. General anesthesia was selected as the primary anesthetic plan for cesarean delivery with bilateral tubal ligation along with a multimodal postoperative analgesic plan that included a transversus abdominus plane block.

On the day of surgery, the patient had a rapid sequence induction performed under direct laryngoscopy with lidocaine, propofol, and succinylcholine. Delivery was uneventful and the neonate had Apgar scores of 8 at 1 minute and 9 at 5 minutes. Prior to emergence, the patient received fentanyl, acetaminophen, and ketorolac intravenously along with a transversus abdominus plane (TAP) block performed under ultrasound, where 40 mL of 0.25% bupivacaine was administered. In the immediate postoperative period, the patient reported her pain as 10/10. She was given patient-controlled analgesia with hydromorphone and her next documented pain score was 2/10 approximately 6 hours after her procedure. On the first postoperative day, the patient expressed satisfaction with the avoidance of neuraxial anesthesia and possible sequelae.

DISCUSSION

Patients with Marfan syndrome who have an aortic diameter >4 cm are at higher risk for aortic dissection.⁵ Our patient had an aortic diameter of 4.1 cm, was managed with beta-blockers, and was advised by her cardiologist not to become pregnant again. Epidural anesthesia has been recommended for patients with Marfan syndrome undergoing cesarean delivery due to its propensity to avoid large hemodynamic shifts.⁶ Patients with Marfan syndrome also have a predisposition for dural ectasia, which increases the risk of unintentional dural puncture.⁷

The evidence regarding the safety of neuraxial anesthetic techniques in patients with multiple sclerosis, particularly subarachnoid blocks, is mixed.⁸ However, a survey among anesthesiologists in the United Kingdom found that 98% would perform a neuraxial technique for cesarean delivery.⁹

Closed claims data from the 1980s indicated that maternal morbidity and mortality were significantly higher in patients who had cesarean sections with general anesthesia compared to patients who had a neuraxial technique.¹⁰ In response, obstetric anesthesiologists changed their practice to decrease the number of cesarean sections performed under general anesthesia.¹¹

The patient had a pain score of 10/10 in the immediate postoperative period that could be attributed to the TAP block not yet reaching full efficacy. We used bupivacaine, which is commonly employed for these blocks.^{12,13} Perhaps performing the TAP block preoperatively would have allowed more time for the block to set up, producing a better pain score.

Shared decision making has been proposed as the preferred form of decision making in patient-centered care.¹⁴ Our patient had a strong preference to avoid the discomfort of neuraxial placement and potential morbidity from a postdural puncture headache. A neuraxial anesthetic technique is usually preferred in patients undergoing cesarean delivery and it is safe to perform this technique in patients with either Marfan syndrome or multiple sclerosis. Through shared decision making we formulated an anesthetic plan that honored our patient's autonomy and produced a good maternal and neonatal outcome.

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1. Judge DP, Dietz HC. Marfan's syndrome. *Lancet*. 2005;366(9501):1965–1976.
2. Compston A, Coles A. Multiple sclerosis. *Lancet*. 2008;372(9648):1502–1517. doi:10.1016/S0140-6736(08)61620-7.
3. Gaiser RR. Postdural puncture headache: an evidence-based approach. *Anesthesiol Clin*. 2017;35(1):157–167. doi:10.1016/j.ancclin.2016.09.013.
4. American Society of Anesthesiologists task force on obstetric anesthesia, Society for Obstetric Anesthesia and Perinatology. Practice guidelines for obstetric anesthesia. *Anesthesiology*. 2016;124(2):270–300.
5. Hiratzka LF, Bakris GL, Beckman JA, et al. 2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM guidelines for the diagnosis and management of patients with thoracic aortic disease. *J Am Coll Cardiol*. 2013;62(11):1039–1040.
6. Goland S, Elkayam U. Pregnancy and Marfan syndrome. *Ann Cardiothorac Surg*. 2017;6(6):642–653. doi:10.21037/acs.2017.10.07.
7. Vacula BB, Gray C, Hofkamp MP, et al. Epidural analgesia complicated by dural ectasia in the Marfan syndrome. *Proc (Bayl Univ Med Cent)*. 2016;29(4):385–386. doi:10.1080/08998280.2016.11929477.
8. Makris A, Piperopoulos A, Karmanioliou I. Multiple sclerosis: basic knowledge and new insights in perioperative management. *J Anesth*. 2014;28(2):267–278. doi:10.1007/s00540-013-1697-2.
9. Drake E, Drake M, Bird J, et al. Obstetric regional blocks for women with multiple sclerosis: a survey of UK experience. *Int J Obstet Anesth*. 2006;15(2):115–123. doi:10.1016/j.ijoa.2005.10.010.
10. Hawkins JL, Koonin LM, Palmer SK, et al. Anesthesia-related deaths during obstetric delivery in the United States, 1979–1990. *Anesthesiology*. 1997;86(2):277–284. doi:10.1097/00000542-199702000-00002.
11. Tsen LC, Pitner R, Camann WR. General anesthesia for cesarean section at a tertiary care hospital 1990–1995: indications and implications. *Int J Obstet Anesth*. 1998;7(3):147–152.
12. Tawfik MM, Mohamed YM, Elbadrawi RE, et al. Transversus abdominus plane block versus wound infiltration for analgesia after cesarean delivery: a randomized controlled trial. *Anesth Analg*. 2017;124(4):1291–1297. doi:10.1213/ANE.0000000000001724.
13. John R, Ranjan RV, Ramachandran TR, et al. Analgesic efficacy of transverse abdominal plane block after elective cesarean delivery—bupivacaine with fentanyl versus bupivacaine alone: a randomized, double-blind controlled clinical trial. *Anesth Essays Res*. 2017;11(1):181–184. doi:10.4103/0259-1162.186864.
14. Sandman L, Munthe C. Shared decision-making and patient autonomy. *Theor Med Bioeth*. 2009;30(4):289–310.