Management of a Patient with Moyamoya, Neurofibromatosis Type 1 Requiring Urgent Delivery for Pre-eclampsia with Severe Features

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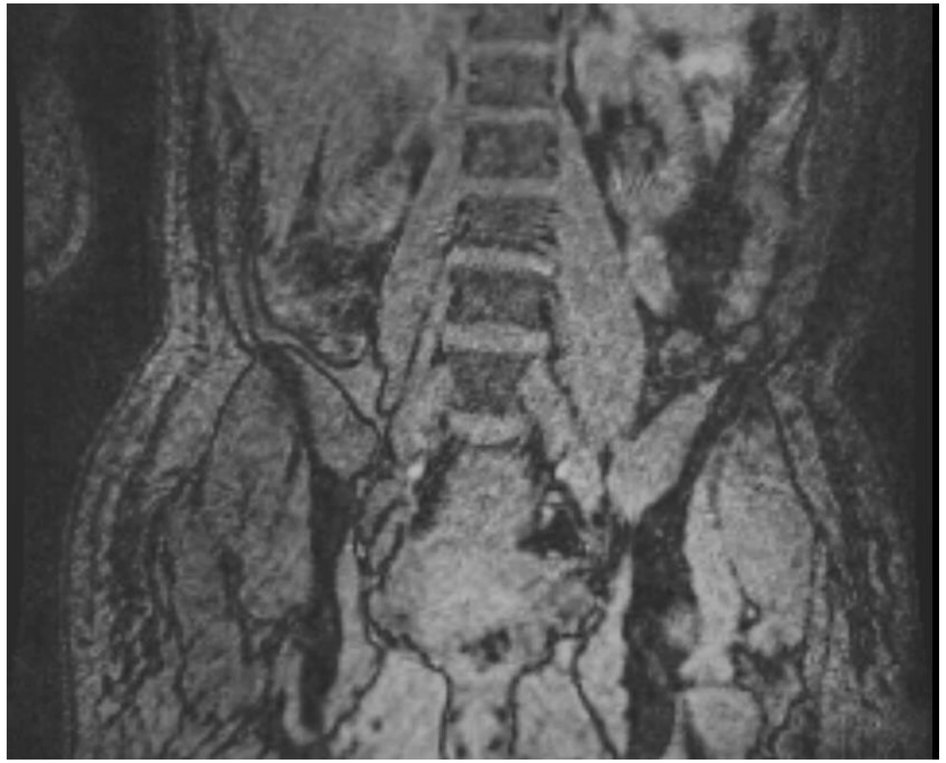
Background

Neurofibromatosis (NF) is a group of genetic disorders characterized by tumors of nerve tissue. NF Type 1 (NF1) is associated with cafe au lait spots, neurofibromas and scoliosis. NF has also been associated with Moyamoya disease (MMD),[1] a rare cerebrovascular disorder in which narrowing of the internal carotid arteries and the formation of fragile collateral vessels predispose the patient to cerebrovascular accidents (CVA). The physiologic changes of pregnancy can increase the number and size of neurofibromas and risks associated with MMD, posing significant challenges for maternal and fetal health.



Case Presentation

We present a 25 year old G1P0 with a history of NF1 and MMD with bilateral encephaloduroarteriosynangiosis (EDAS), seizures and CVA in childhood with residual right upper extremity weakness and numbness requiring cesarean delivery at 27w4d for pre-eclampsia with severe features

1)Maragkos, Georgios A., et al. “Moyamoya Disease in Pregnancy: A Systematic Review - Acta Neurochirurgica.” *SpringerLink*, Springer Vienna, 19 June 2018, link.springer.com/article/10.1007/s00701-018-3597-6.

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Case Presentation

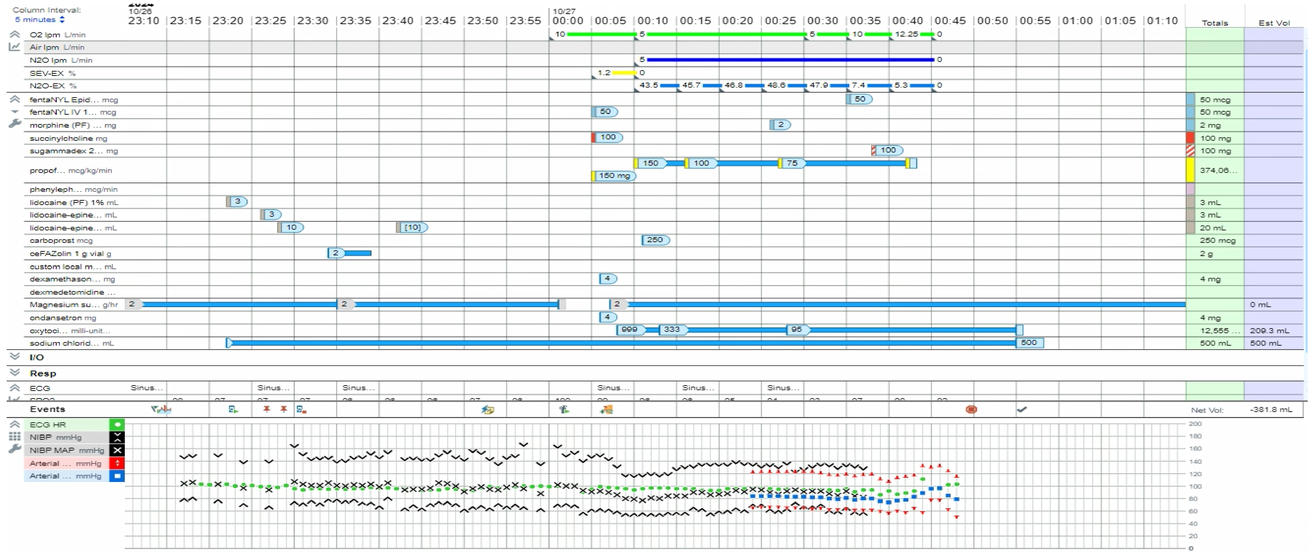
1 Initial Management
- Hypertension → Nifedipine & Labetalol
- Pulmonary Edema → Furosemide (Diuresis)
- Neurology Consulted → MRI Lumbar Spine for planned neuraxial

2 Indications for Urgent Cesarean Delivery
- Progressive Hypertension
- Rising Creatinine
- Worsening Flank Pain

3 Anesthesia & Intraoperative Course
- Planned: Combined Spinal-Epidural → Failed Spread
- Converted to General Anesthesia:
  - RSI: Propofol, Succinylcholine, Fentanyl
  - Maintenance: Propofol Infusion + Nitrous Oxide

4 Hemodynamic Monitoring & Challenges
- Arterial Line ✗ Difficult due to Vasculitis
- Alternative: ClearSite Noninvasive BP Monitor
- Outcome: Stable Hemodynamics (No Vasopressors Needed)

5 Postoperative Recovery
- Uneventful Extubation
- PACU Recovery Without Complications



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Teaching Points

Anesthetic Approach for Moyamoya Disease (MMD):

- Maintain adequate cerebral perfusion. Both general and regional anesthesia have been utilized, though the choice depends on the patient's comorbidities and the case's urgency, as well as team comfortability[1]

Challenges with Epidural or Spinal Anesthesia in NF1 Patients

- Neurofibromatosis type 1 (NF1) can have spinal pathology at baseline and increases the risk of epidural hematoma due to neurofibromas' growth during pregnancy.[2] Preoperative spinal imaging/ early neurology consult allow for risk stratification

Hemodynamic Monitoring

- While invasive blood pressure monitoring is optimal for MMD cases, noninvasive methods like ClearSight may be reasonable alternatives in urgent situations, particularly when vascular disease severity precludes invasive techniques.

Anesthetic Agents

- Propofol and nitrous oxide can be used, balancing cerebral oxygen consumption reduction with cerebral blood flow maintenance.
- Total intravenous anesthesia (TIVA) with propofol and remifentanil or sevoflurane alone are also described options.[3]
- Though nitrous oxide is generally avoided in MMD due to concerns of elevated ICP and cerebral steal, it has demonstrated safe use in certain cases with benefits like rapid titration and stable uterine tone.[4]

1.Maragkos, Georgios A., et al. “Moyamoya Disease in Pregnancy: A Systematic Review - Acta Neurochirurgica.” *SpringerLink*, Springer Vienna, 19 June 2018, link.springer.com/article/10.1007/s00701-018-3597-6.
2.Alves Júnior, Sérgio Ferreira, et al. “Neurofibromatosis Type 1: State-of-The-Art Review with Emphasis on Pulmonary Involvement.” *Respiratory Medicine*, vol. 149, Mar. 2019, pp. 9–15, https://doi.org/10.1016/j.rmed.2019.01.002.
3.Yang, Kevin J., et al. “Update on the Anesthesia Management in Adult Patients with Moyamoya Disease.” *Current Opinion in Anaesthesiology*, vol. 37, no. 5, Lippincott Williams & Wilkins, June 2024, pp. 439–45, https://doi.org/10.1097/aco.0000000000001411. Accessed 22 Jan. 2025.
4.Sharma, V., Prabhakar, H., Girija Prasad Rath, & Bithal, P. K. (2014). Anaesthetic management of patients undergoing surgery for Moyamoya disease – our institutional experience. *Journal of Neuroanaesthesiology and Critical Care, 01*(02), 131–136. https://doi.org/10.4103/2348-0548.130399