Cesarean Delivery in a Parturient with Neurofibromatosis Type 1

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Background

- Neurofibromatosis 1 (NF1) is an autosomal dominant disease characterized by the presence of multiple benign tumor growth on skin and central nervous system
- Pregnancies in NF1 associated with higher incidence of pre-term labor, cesarean delivery, and intrauterine fetal growth restriction
- May be an increase in the number and/or size of dermal neurofibromas and CNS tumors during pregnancy
- Elevated risk of hypertension and preeclampsia spectrum due to renal artery stenosis, aortic coarctation or pheochromocytoma

Patient Demographics

- 21 yo G1P1001 w/ pregnancy complicated by NF1 diagnosed at birth with no history of epilepsy, normal TTE and preeclampsia labs
- Pt was seen in our preop clinic via a video visit. The café au lait spot was noted, and a preop thoracic and lumbar MRI was ordered
- Preop thoracic/lumbar MRI final read stated no abnormal epidural fluid collection or masses within spinal canal with large subcutaneous edema posteriorly in the lumbar region



Figure 1: Neurofibromatosis Type 1 Manifestations



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Case Description

- When presenting for L&D, notable findings included cafe au lait spot extending along the entire lumbar spine as well as extensive edema
- US exam of the back: many circumscribed nodules in subcutaneous tissue
- On review of imaging, large amount of neurofibromas and nodules seen in the subcutaneous and deep subcutaneous region covering the entire lumbar region
- Risk/benefit discussion was had with the patient regarding systemic opioid analgesia, after discussing risks to fetus of opioid medication shortly preceding delivery. Patient desired to proceed with nitrous oxide gas as well as fentanyl PCA.
- Once pain interventions initiated, reevaluated patient. Reported feeling extremely uncomfortable and requesting cesarean delivery. Repeat SVE unchanged at 4/80/-2. Discussed options for pain control during her labor course including nitrous, continued PCA use, and pudendal block
- Given inability to safely perform neuraxial anesthesia, proceeded with low transverse cesarean delivery under GA at 39w3d
- Rapid sequence intubation with 6.5 mm tube successful with a video laryngoscope
- Baby delivered with no complications to mother or baby



Figure 1: Preoperative Lumbar and Thoracic MRI



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Discussion

- Older guidelines for NF1 patients recommended avoiding neuraxial anesthesia unless prior brain and spine imaging was conducted
- Latest guidelines from the American College of Medical Genetics and Genomics states that preanesthetic neuraxial imaging to assess spinal or paraspinal neurofibromas is probably unnecessary before neuraxial anesthesia unless symptoms warrant further investigation
- Always review available as the one performing the procedure
- Ultrasound is a quick, noninvasive way to obtain more diagnostic information of back anatomy/abnormalities
- Regional anesthesia may be a viable option but may be technically difficult if spinal neurofibromas or scoliosis are present
- Raised ICP from cranial tumors and risk of herniation must be ruled out
- Airway management with GA may be complicated by intra-oral pathologies such as tongue or laryngeal neurofibromas, mandibular abnormalities, and cervical involvement



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