Large Plexiform Neurofibroma Causing Supraglottic and Subglottic Tracheal Stenosis

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Background:

- Neurofibromatosis Type 1 (NF1) presents unique challenges in obstetric anesthesia, particularly concerning the airway, cardiopulmonary, and neurologic manifestations[1]
- Patients face an increased risk of hypertension, including renovascular disease or pheochromocytoma[1]
- Subglottic stenosis can worsen during pregnancy leading to airway compromise[2]
- Two contributing etiologies of subglottic stenosis put this patient at elevated risk for worsening stenosis and airway complications

1. Hirsch NP, Murphy A, Radcliffe JJ. Neurofibromatosis: clinical presentations and anaesthetic implications. Br J Anaesth. 2001;86(4):555-564. doi:10.1093/bja/86.4.555.
2. Kuczkowski KM, Benumof JL. Subglottic tracheal stenosis in pregnancy: anaesthetic implications. Anaesth Intensive Care. 2003;31(5):576-577. doi:10.1177/0310057X0303100514.

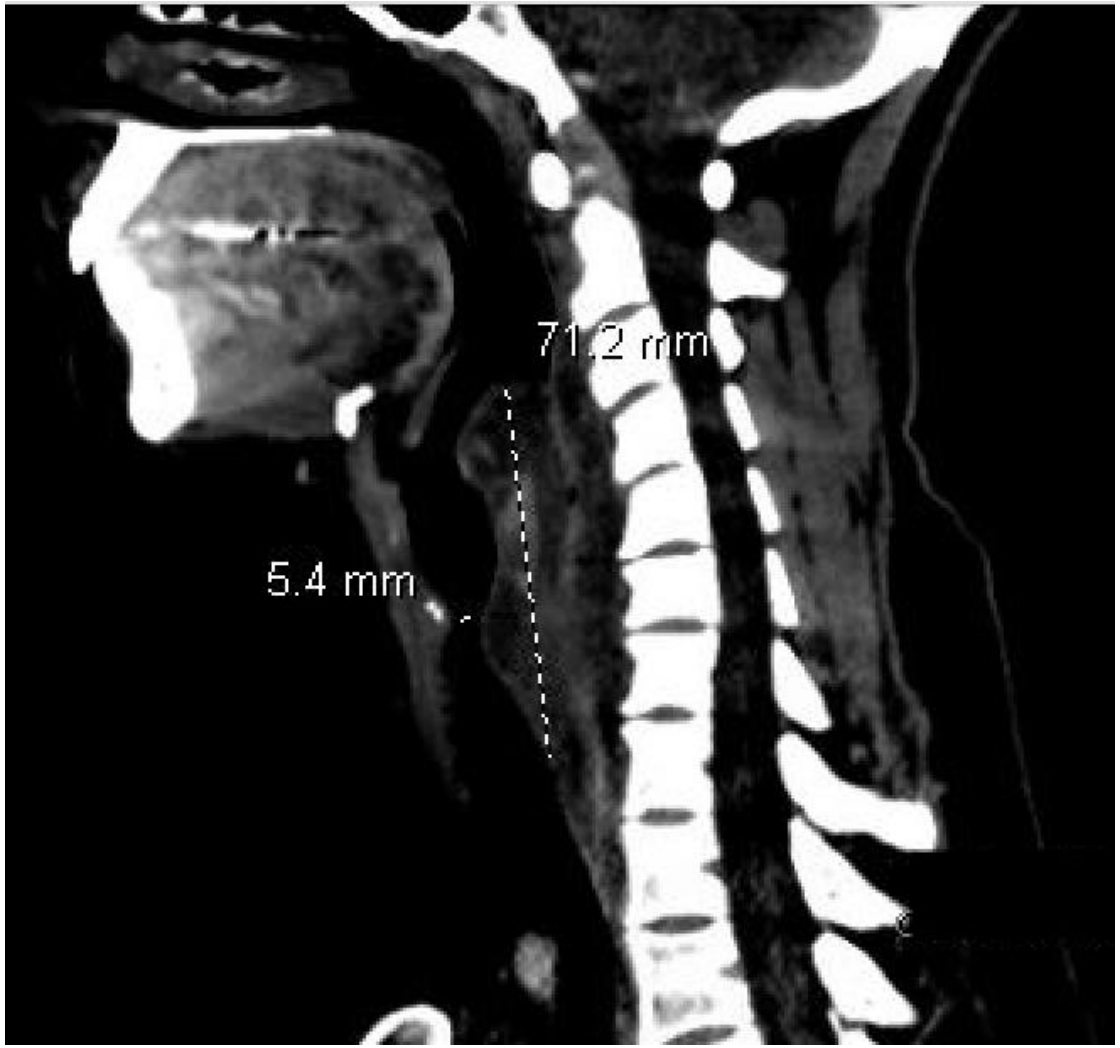
Case Progression

Preoperative

- 31-year-old G2P1001 with a history of a difficult airway due to a large plexiform neurofibroma presented for repeat Cesarean delivery (CD)
- History of awake tracheostomy and partial resection of NF
- Decannulated one year later
- Subsequently required a balloon dilation, LASER, and debridement of the neurofibroma
- Inspiratory stridor on exam, but unchanged from baseline and no worsening throughout pregnancy
- ENT evaluation including fiberoptic nasal laryngoscopy which visualized right sided NF with patent glottis
- A multidisciplinary team, including obstetrics, ENT, and OB anesthesia, planned for a repeat CD under neuraxial in the main operating rooms to ensure ENT availability for airway backup.

Delivery

- Combined spinal epidural with 1mL of 0.75% hyperbaric bupivacaine, fentanyl and morphine
- A T4 level was obtained after 15mL of 2% epidural lidocaine
- Intraoperative and postoperative course were uncomplicated



Teaching Points

- NF1 can involve multiple organ systems, necessitating careful obstetric anesthetic evaluation.
- Patients face an increased risk of hypertension, including renovascular disease or pheochromocytoma.[1]
- Neurofibromas affecting the airway may cause glottic obstruction or subglottic tracheal stenosis, necessitating thorough preoperative airway evaluation.
- Pregnancy can both worsen subglottic stenosis and increase neurofibroma size putting a patient with both pathologies at high risk for airway compromise.[2]
- Spinal involvement, though typically extradural, may alter neuraxial anesthesia spread or increase the risk of bleeding into the epidural space.[1]
- These considerations underscore the importance of multidisciplinary planning for safe delivery in the setting of NF1 and subglottic stenosis

1. Hirsch NP, Murphy A, Radcliffe JJ. Neurofibromatosis: clinical presentations and anaesthetic implications. Br J Anaesth. 2001;86(4):555-564. doi:10.1093/bja/86.4.555.
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