



Cesarean Delivery in a Patient with Blue Rubber Bleb Nevus Syndrome

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Introduction

Blue Rubber Bleb Nevus Syndrome (BRBNS) is a rare condition characterized by venous malformations that present in variable locations throughout the body. Symptoms range from asymptomatic or mild to life-threatening bleeding or compression. Few case reports exist describing BRBNS in pregnant patients. Here we present a case of successful anesthetic management of a pregnant woman with BRBNS.

Patient Presentation

- ❖ 21-year-old G1P0 female at 17w5d gestation
- ❖ PMH: BRBNS complicated by chronic anemia requiring multiple blood transfusions and anti-Kell isoimmunization
- ❖ Patient previously evaluated by Dermatology, Hematology, and Gastroenterology services

Case Description

Patient presented at 17w5d gestation for Obstetric consultation. She was initiated on aspirin therapy and advised to follow up with a multidisciplinary team in the outpatient setting.

She re-presented at 26w6d gestation with complaints of dyspnea, fatigue, and dizziness. Lab work revealed Hb of 5.3 g/dL for which she received two units of packed red blood cells and intravenous iron infusion. Patient was evaluated by Obstetric Anesthesia, Otolaryngology, Gastroenterology, Hematology, and Dermatology services during this admission.

Flexible laryngoscopy showed no pharyngeal or laryngeal lesions however, the subglottic airway was not examined.

Case Description (cont'd)

Magnetic Resonance Imaging (MRI) of the spine revealed multiple lesions in the lumbar region, including a large lesion at approximately L4-L5 on the left and two lesions in the gluteal region without obvious connection to the spinal canal or lumbosacral plexus. Based on these results, the patient was to be offered neuraxial anesthesia under ultrasound-guidance during scheduled induction.

The patient later presented at 39w3d with symptoms of COVID-19 infection at which time she underwent induction of labor.

The patient refused neuraxial anesthesia and eventually required urgent cesarean delivery under general anesthesia for non-reassuring fetal heart tones. The patient was intubated via video laryngoscope without incident and delivery was uncomplicated.

The patient required one unit of packed red blood cells on the first postoperative day but otherwise had no significant complications.

Imaging

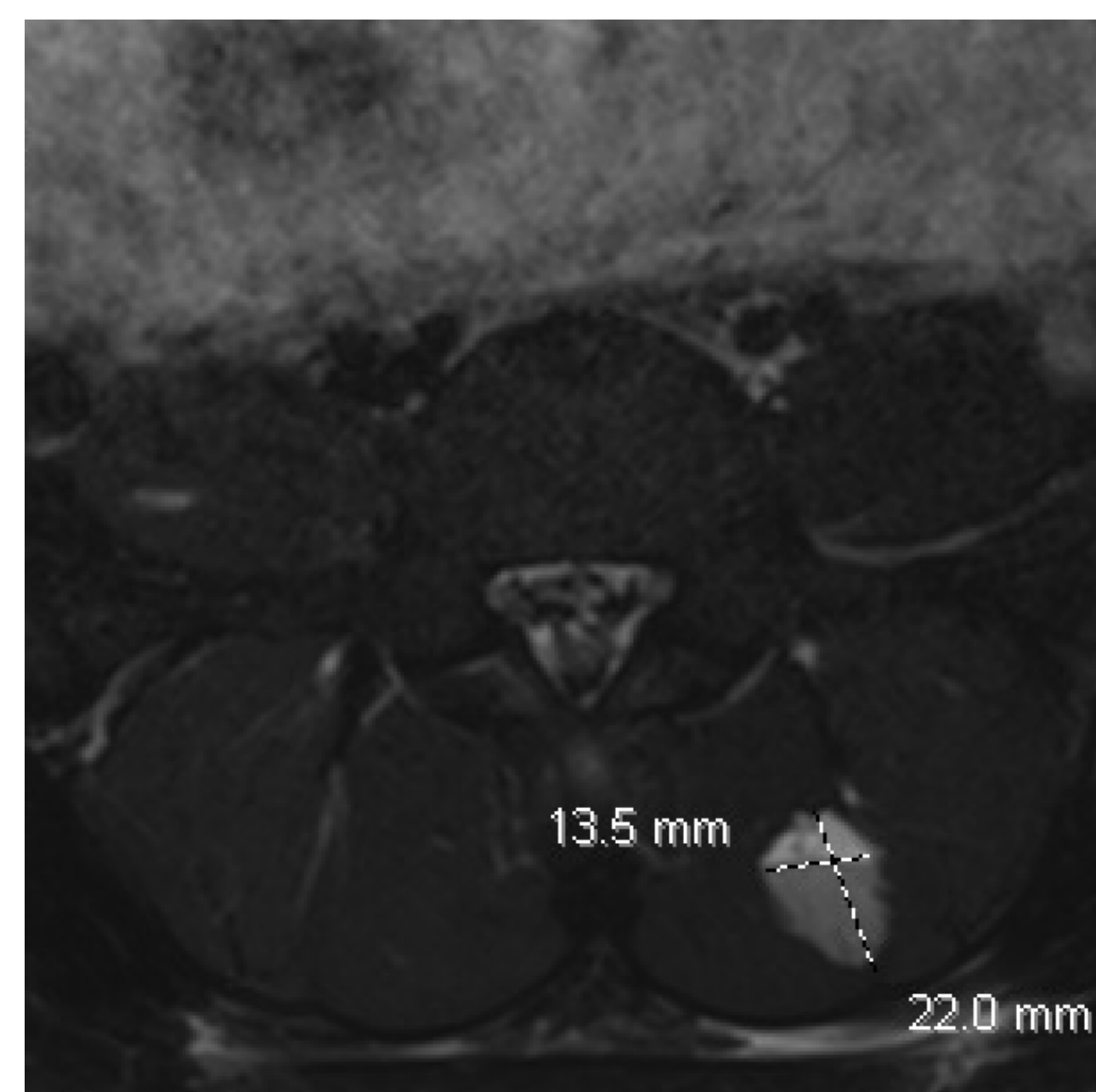


Figure 1. Magnetic resonance imaging of L4-L5 lesion

Discussion

BRBNS is a rare condition of unclear pathogenesis with vascular malformations typically presenting early in life¹.

Lesions are most commonly reported in the cutaneous and gastrointestinal tracts but have been described in nearly all organ systems^{1,2,3}. Symptoms are largely dependent on the location of the venous malformations. In patients with gastrointestinal lesions, chronic iron deficiency anemia requiring iron infusion or blood transfusion is common¹.

Evaluation of the airway is crucial due to potentially catastrophic airway hemorrhage with laryngoscopy and endotracheal tube placement^{2,3}. Similarly, evaluation for neuraxial involvement is essential given the risk of epidural hematoma formation^{2,3}. Recent literature suggests whole-body MRI close to the time of delivery given that the venous malformations may enlarge throughout pregnancy².

Given the rarity of the disease, there is an overall paucity of literature detailing anesthetic management of pregnant women with BRBNS. Available case reports describe success with both neuraxial and general anesthesia, with a multidisciplinary team being vital to their care^{1,2,3}.

Introduction

1. Jin, X. L., Wang, Z. H., Xiao, X. B., Huang, L. S., & Zhao, X. Y. (2014). Blue rubber bleb nevus syndrome: a case report and literature review. *World Journal of Gastroenterology: WJG*, 20(45), 17254.
2. Hult, M., Halldorsdottir, H., Stjernholm, Y. V., Hein, A., & Jörnvall, H. (2021). Blue Rubber Bleb Nevus Syndrome in the Obstetric Patient: A Case Report of Anesthetic Implications and Management. *A&A Practice*, 15(8), e01517.
3. Galey, J., Bharadwaj, S., Crimmins, S., Hong, C. M., & Malinow, A. M. (2016). Anesthetic implications of an obstetric patient with blue rubber bleb nevus syndrome. *A&A Practice*, 6(6), 146-149.