

Blue Rubber Bleb Nevus Syndrome in Pregnancy

 Department of Anesthesiology
Perioperative, Critical Care
and Pain Medicine

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Background

Presentation

25 yo G1P0 with BRBNS presenting for delivery planning at 31 weeks

Pathophysiology

Rare condition causing venous malformations (VMs) both cutaneously and within visceral organs, most commonly the GI tract¹

Epidemiology

Autosomal dominant condition effecting 1/14,000 births

Imaging findings

MRI head, lumbar, and thoracic spine without evidence of Vascular Malformations

Management

Multidisciplinary approach to with obstetric anesthesia, MFM, OBGYN, and Radiology

Prognosis

Most patients have a good prognosis, although variable given extent of GI and CNS involvement

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Previous BRBNS Complications

- Previous GI VMs requiring multiple bowel resections, partial gastrectomy, and transanal excision of blebs.
- Classic lesions on her arms, shoulder, legs, and lips.
- Routine Fe infusions via portacath for chronic anemia

Delivery Planning

- MRI Head and Spine 4 weeks prior to delivery to evaluate safety of neuraxial
- Patient desired epidural & vaginal delivery
- Previous vaginotomy without blebs, okay for SVD per MFM
- VMs on upper & lower lips making intubation challenging
- Admitted for 38w6d for gestation HTN, later progressing to pre-eclampsia

Labor Admission

- HCT & platelets reassuring at time of admission

Delivery

- Multidisciplinary discussion
- Smooth, atraumatic epidural placement
- SVD with QBL 100

Postpartum Care

- Uncomplicated
- No evidence of postpartum hemorrhage. Hb 11.5g/dL on POD1
- Nifedipine for blood pressure management and 24hrs of mag sulfate postpartum

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Learning Points

Perioperative Evaluation

- CNS imaging as close to delivery as possible for neuraxial anesthesia
- Multidisciplinary management
- Pelvic evaluation if SVD is desired

Management

- Large bore IV access in case of hemorrhage
- Evaluation of CNS imaging and physical exam to assess safety of neuraxial anesthesia
- Active type/cross
- Backup planning is essential in case GA is indicated

Outcomes

- With proper planning and imaging - neuraxial anesthesia can be safely performed

References

