

Blue Rubber Bleb Nevus Syndrome in the Obstetric Patient: A Case Report of Anesthetic Implications and Management

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Blue rubber bleb nevus syndrome (BRBNS) is a rare systemic syndrome characterized by venous malformations usually found in the skin and visceral organs. To date, 11 case reports describing BRBNS during pregnancy have been published. To our knowledge, this is the first report describing intracranial, airway, epidural, and birth canal involvement of venous malformations in the same parturient. Key lessons learned include clinical presentation, workup, team management, and care of obstetric patients with this disorder. (A&A Practice. 2021;15:e01517.)

GLOSSARY

BRBNS = blue rubber bleb nevus syndrome; **MRI** = magnetic resonance imaging

Blue rubber bleb nevus syndrome (BRBNS) is a rare systemic syndrome characterized by venous malformations.¹ The etiology remains unknown and the majority of cases occur sporadically as a new mutation, although a few autosomal dominant cases have been reported.^{1,2} Histopathological findings are often consistent with cavernous hemangiomas of varying size that may increase over time.^{1,3,4} The most common locations of venous malformations are the skin and the gastrointestinal tract with subsequent bleeding and iron deficiency anemia. However, venous malformations can also be present in the central nervous system, spine, upper airway, soft tissue, and other internal organs. A majority of patients (87%) have vascular malformations present at multiple sites.¹ Possible complications include massive bleeding, cerebral hemorrhage, thrombocytopenia, and consumptive coagulopathy.^{1,3,5} Approximately 200 cases of BRBNS have been described in the literature,^{1,5} of which 11 cases are reported

in pregnant women.²⁻¹² To our knowledge, this is the first report of BRBNS describing intracranial, airway, epidural, and birth canal involvement in the same parturient.

This case report was approved by the Swedish Ethical Review Authority and written informed patient consent was obtained.

CASE DESCRIPTION

A 37-year-old primiparous woman with a body mass index of 27 kg/m², who was diagnosed with BRBNS in childhood, was referred to our hospital at 36 weeks of gestation. She was otherwise healthy with asymptomatic blue rubber bleb nevus located on her skin, but investigation for further organ involvement had not been performed. Before referral to a tertiary care center, she was told no further testing or imaging was necessary in preparation for delivery and had planned for a vaginal delivery with epidural analgesia. However, an obstetric anesthesiologist at the referring hospital ordered a magnetic resonance imaging (MRI) before delivery which revealed multiple venous malformations involving the soft tissues of the neck, airway, and birth canal. Intracranial and epidural malformations were also found. The venous malformations in the soft tissues of the neck were in close proximity to large veins and involved the base of the tongue. The intracranial malformations were located in the right occipital and temporal lobes and left side of the pons. The epidural involvement was located at L2-L3 and L1-L2, the latter compressing the dural sac and dislocating it ventrally (Figure 1).

Clinical examination showed that the venous malformations on her neck distended during Valsalva maneuver. A fiberoptic laryngoscopy was performed to further evaluate the upper airway. This procedure was challenging because of the patient's anxiety and fear of the hospital environment, but revealed large venous malformations in the epiglottic vallecula (Figure 2). Despite this, the patient had not experienced any voice change during pregnancy.

After a multidisciplinary conference, a cesarean delivery under general anesthesia was planned at 37 gestational

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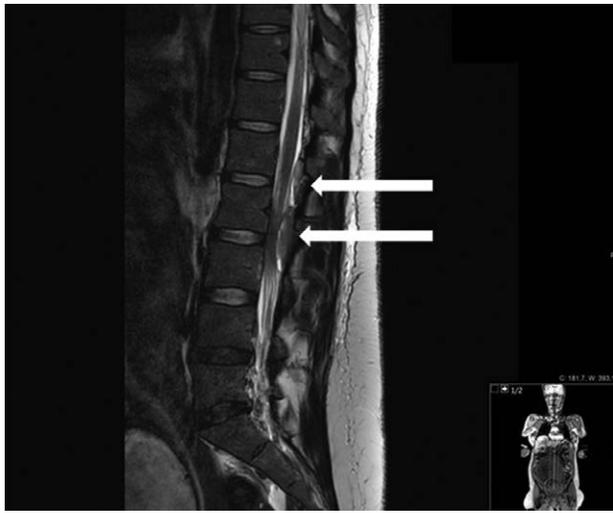


Figure 1. MRI performed at 35 + 3 weeks of gestation revealing prominent venous malformations in the epidural space (arrows). MRI indicates magnetic resonance imaging.

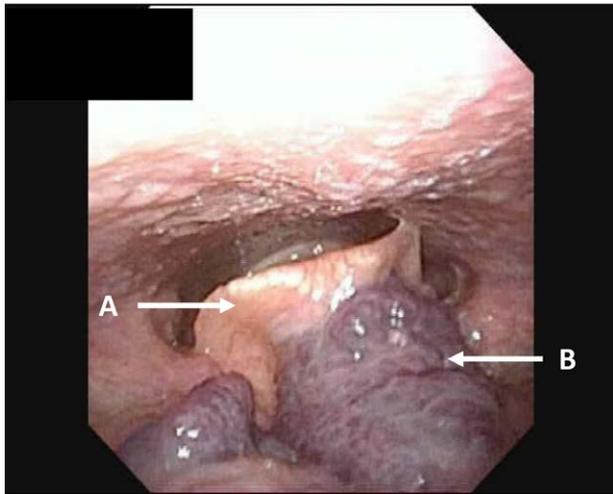


Figure 2. Preoperative fiberoptic laryngoscopy showing the epiglottis (A) and the large venous malformation (B).

weeks. It was performed in a hybrid operating room where an interventional radiologist, a vascular surgeon, and an otolaryngologist were present during the operation. Additional neurosurgery capabilities were available at the hospital.

Before induction of anesthesia, an arterial line was placed in the right radial artery and an otolaryngologist marked the incision site for an emergency cricothyrotomy. The inguinal area was sterilized and draped, in case surgical access or endovascular intervention was needed. General anesthesia was achieved with rapid sequence induction using thiopental (500mg), remifentanyl (0.7 µg/kg/min), and succinylcholine (100mg). The patient was intubated using a C-MAC video laryngoscope (Karl Storz) with curved blade such that the vascular malformations in the epiglottic vallecula were visualized (Figure 3) (Supplemental Digital Content, Video 1, <http://links.lww.com/AACR/A447>). Anesthesia was maintained with sevoflurane and remifentanyl infusion (0.3–0.5 µg/kg/min). A male newborn with birth weight of 2048g and an Apgar score 10 at 5 minutes was delivered 4 minutes after skin

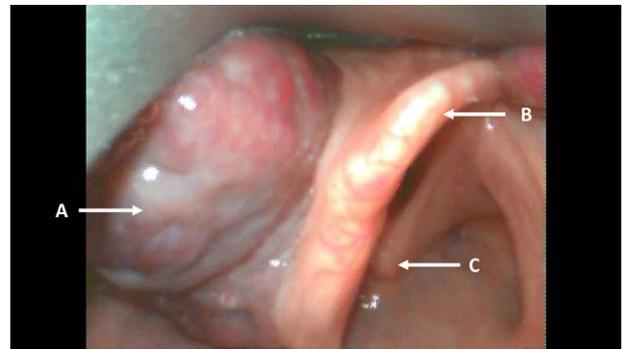


Figure 3. Laryngoscopic view with C-MAC video laryngoscope during intubation. Arrows pointing out the venous malformation (A), epiglottis (B), and posterior cartilage (C).

incision. Perioperative blood loss was 300 mL, and the postoperative care was uncomplicated. No vascular malformations were found in the neonate.

DISCUSSION

The anesthetic management of an obstetric patient with BRBNS is not well described in the medical literature. To our knowledge, this is the first report describing multiple organ involvement with intracranial, airway, epidural, and birth canal venous malformations in the same parturient.

To guide the decision on the mode of delivery, the extent of venous malformation involvement must be fully evaluated. A whole-body MRI is a key part of the workup before delivery. Additionally, since BRBNS malformations have been reported to expand during pregnancy, particularly in the third trimester, the timing of the MRI is of importance.^{4,13} In the presented case, the MRI showed venous malformations in the birth canal. This finding is generally considered a contraindication for vaginal delivery,^{2,4,7,10,11} as rupture of paravaginal malformations could cause uncontrollable bleeding. Thus, in the presented case, we chose to deliver by an elective cesarean delivery.

The findings of the predelivery MRI will also influence the anesthetic management. In the presented case, neuraxial blockade was considered contraindicated due to the epidural involvement of the venous malformations with the unacceptable risk of causing an iatrogenic epidural hemorrhage as well as unpredictable efficacy of administered local anesthetic. We therefore opted to perform the cesarean delivery under general anesthesia. However, the airway management was a concern as venous malformations in the upper airway constitute a potential risk for iatrogenic bleeding during airway manipulation. Furthermore, the patient's voice was unchanged during pregnancy, indicating that the absence of self-perceived voice changes does not rule out clinically relevant airway involvement. Therefore, thorough evaluation of the airway including pre anesthetic fiberoptic laryngoscopy is recommended on broad indications.

As the preoperative airway examination had been a challenge for the patient, it was obvious that she would not cope with an awake fiberoptic intubation. Therefore, we planned for intubation using video laryngoscopy, with an otolaryngologist prepared to perform an emergency cricothyrotomy in case of a difficult intubation. We considered this to be safer than a fiberoptic intubation with heavy sedation in a parturient at term.

Irrespectively of mode of delivery and anesthetic technique, necessary peripartum monitoring must be carefully evaluated. An arterial line was placed for precise blood pressure monitoring during induction due to the intracranial involvement as well hemodynamic monitoring in case of profuse perioperative hemorrhage. In addition, an interventional radiologist was standby to perform endovascular embolization or placement of a balloon occlusion catheter in the aorta.

In conclusion, the presented case illustrates the importance of a whole-body MRI in late pregnancy to evaluate the localization and extent of vascular malformations in a parturient with BRBNS. Pending birth canal involvement, an optimal mode of delivery can be chosen. The anesthetic management must be individualized and planned according to the examination findings with special consideration of potentially difficult airway or neuroaxial involvement. ■■

DISCLOSURES

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