Anesthetic Management of Emergency Cesarean Section in a Patient with Congenital Venous Malformation

Harsha Narkhede, Shrikanta P Oak, Isha Singhal, Neha Apte, Monal Shah

ABSTRACT

Blue rubber bleb nevus syndrome (BRBNS) is characterized by typical bluish, soft, and compressible lesions with rubber-like consistency present since birth. In this syndrome, venous malformation (VM) is always present since birth and is commonly found in the cheek, tongue, lip, and jaw, and sometimes may be present in deep tissue, bone, or muscle. A vascular malformation is known to grow in size throughout the life of an individual with sudden enlargement in size during puberty and pregnancy due to hormonal changes. We report the successful management of a case of congenital VM of the lip and mandible posted for emergency cesarean section under spinal anesthesia.

Keywords: Blue rubber bleb nevus syndrome, Cesarean section, Congenital venous malformation, Pregnancy, Spinal anesthesia.

INTRODUCTION

Venous, lymphatic, and venolymphatic malformations are the most common type of vascular malformations with an overall prevalence of up to 1% in general population. Venous malformation is an abnormality of the larger and deep vessels. It appears dark blue and is very soft to touch and can bleed easily. A VM is always present since birth and is commonly found in the cheek, tongue, lip, and jaw, and sometimes may be present in deep tissue, bone, or muscle. A vascular malformation is known to grow in size throughout the life of an individual with sudden enlargement in size during puberty and pregnancy due to hormonal changes.

Emergency cases presenting with an incidental finding of a superficial VM, complaining with history of sudden enlargement or associated VM of spine and brain, may not be properly evaluated and can be a cause of increased morbidity and mortality. Therefore, a complete and comprehensive understanding is vital for the anesthesia management of such cases. Here, we report the anesthetic challenges involved with the management of a case of congenital VM of the lip and mandible posted for emergency cesarean section.

CASE REPORT

A 30-year-old gravida 4 parity 3 living 1 abortion 2 female first presented at 27 weeks of gestation with lip and mandibular VM and was advised an antenatal visit including neuromedicine, interventional radiologist reference, and magnetic resonance imaging (MRI) of head and spine. Patient was lost to follow-up for a month and directly came to the emergency at 30 weeks of gestation with a complaint of decreased fetal movement since 2 days. Fetal heart sounds were not localized on Doppler and ultrasound was suggestive of intrauterine fetal demise. Patient went into spontaneous labor and obstetricians planned an emergency cesarean section on the same day as she had previous three lower segment cesarean sections.

On examination, general condition and history were not suggestive of bleeding from gastrointestinal tract and consumption coagulopathy. She had bluish VM on the lower jaw and lip with rubber-like consistency on palpation (Figs 1 and 2). Patient's weight was 65 kg, height 153 cm, pulse rate 80 beats per minute, and blood pressure 110/70 mm Hg. The temporomandibular joint was mobile with Mallampati grade II. Hemoglobin was 11 gm/dL, platelet 2 lakhs/cumm, with normal coagulation profile. The patient was counseled for surgery and an informed consent with American Society of Anesthesiologists grade IIE was obtained. Crossed-matched blood and blood products were kept ready. Looking at the nature and vascularity of the lesion, a decision of avoiding any possible manipulation of the airway and lesion was made. Magnetic resonance imaging of head and whole spine could not be performed as patient went into spontaneous labor with intrauterine fetal demise. As there was no history of seizure or neurodeficit and tenderness of spine, combined spinal and epidural anesthesia was planned expecting prolonged duration of surgery because of previous three lower segment cesarean sections.
was atraumatic with clear cerebrospinal fluid on first pass of spinal needle. Subarachnoid block was achieved with 1.5 mL of hyperbaric bupivacaine and epidural catheter was inserted safely. Sensory block of T6 was achieved. After regression of sensory level to T8, epidural was activated with 2 mL of 0.5% xylocard. Oxygen supplementation was done via facemask. Patient was stable hemodynamically throughout perioperative period. Blood pressure at the end of surgery was 120/80 mm Hg. Postoperatively on 2nd day, she was referred to the interventional radiologist and neurologist for further management of the congenital VM like sclerotherapy, laser, etc.

**DISCUSSION**

Cutaneous arteriovenous malformations are present in some syndromes like hereditary hemorrhagic telangiectasia, Sturge–Weber syndrome, von Hippel–Lindau disease, etc. But typical bluish, soft, and compressible lesions with rubber-like consistency are present since birth in BRBNS.4,5 There are approximately 150 cases reported in the world, out of which only 3 cases were present during pregnancy.6 Although these lesions are called as hemangiomas, histologically they are VMs. Most of the cases in BRBNS are sporadic but few familial cases have also been reported.7 In our case, family history was not significant. There is not only a cutaneous manifestation but also systemic involvement, including gastrointestinal system, central nervous system (CNS) with risk of life-threatening hemorrhages. Presenting complaints and symptoms are directly related to the degree and extent of organ system involvement. These patients may have thrombocytopenia, altered coagulation profile, and symptoms like anemia because of intestinal bleed and seizure due to CNS compression.8,9 These lesions are present since birth but may increase in size and frequency with age.10

Diagnosis of BRBNS is based on cutaneous manifestation with or without systemic involvement.11 In BRBNS skin lesions typically appear in trunk (93%), limbs (86%), hip (36%), and face (26%).12 In our case also, cutaneous lesions were similar to lesions of BRBNS. They were painless, bluish, compressible soft lesions over lip and mandible since birth. Such lesions make face mask ventilation difficult as there is a risk of trauma and severe bleeding. Hemangiomas present in cervical area cause restriction of neck movements. Painful oral lesions may cause restriction of mouth opening.

Cutaneous lesions in BRBNS are asymptomatic and many times do not require treatment. But when lesions are present in high-risk areas, such as airway and birth canal, treatment may be required in the form of sclerotherapy or laser. Optimized skin lesions then do not cause any trauma or bleeding in handling the airway during emergency. However, if hemangiomas are present in the birth canal, vaginal delivery is contraindicated due to risk of hemorrhage.

Limitation of our case report was that we could not perform MRI evaluation of patient due to emergency nature of surgery. However, it should be performed close to delivery, since pregnancy may induce new lesions or enlarge existing small ones.13

Considering the emergency nature of surgery; absence of any neurological symptoms, seizures, or gastrointestinal bleed; and the risk of difficult intubation and hemorrhage during airway manipulation, we went ahead with regional anesthesia which would be the safest mode of anesthesia for cesarean section in this patient. Various case reports of difficult intubation were also reported.14 We had kept intubating laryngeal mask airway ready for awake intubation as alternative plan of management in case of emergency as mask ventilation was also difficult.15 In case of Ochiai et al,13 they had to give general

![Fig. 1: Patient with lesion on lip](image1.png)

![Fig. 2: Lesions on lip as well as mandible with difficult mask ventilation](image2.png)
Anesthetic Management of Emergency Cesarean Section

Anesthesia to pregnant patient with BRBNS because of VM with spinal and epidural involvement after MRI. Nirmal et al\textsuperscript{6} managed BRBNS in pregnancy successfully with spinal anesthesia in spite of the presence of small occipital hemangioma.

To summarize, whether regional or general anesthesia is safe in pregnant patients with BRBNS and von Hippel–Lindau disease with systemic VM is still a controversial issue, particularly in the case of nervous system involvement.\textsuperscript{16} It needs more research, but it is wise to be prepared for both. In our view, any pregnant patient with such lesion should undergo detailed examination with at least a MRI of the head and spine to rule out hemangiomas. The MRI evaluation can guide us in planning the mode of anesthesia. Fiberoptic bronchoscopy and video laryngoscopy can help to visualize airway lesions directly.\textsuperscript{17} In all these patients, we should rule out other systemic involvement like that of CNS, gastrointestinal, consumption coagulopathy, etc. So it needs a team approach for correct evaluation, optimization, and management, especially during pregnancy, as pregnancy itself flares up these lesions.

REFERENCES