

Anesthetic Management of a Pregnant Patient with Phacomatosis Pigmentovascularis Undergoing Cesarean Delivery

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Introduction

- Phacomatosis Pigmentovascularis (PPV) is characterized by vascular malformation and a dermal melanosis with or without extracutaneous systemic involvement. (1)
- The systemic involvement and vascular malformation may lead to seizures, intellectual disabilities, and skin cancer

Case Summary

- A 30 year old primigravida, week 39+2, female, presented to the obstetric anesthesia clinic as a known case of **PPV type Cesio Maromorata Congenita**
- She had bilateral ocular melanosis, **increased intraocular pressure**, history of **deep vein thrombosis**, and **gestational DM**
- Her ECG shower short PR** which was not seen previously in PPV patients
- She underwent a **cesarean section** under intrathecal block with 0.5% bupivacaine, fentanyl, and morphine.
- Postoperative recovery was uneventful and was discharged with no need for further pain management

Discussion

- Phacomatosis Pigmentovascularis poses a possible risk for seizures, arrhythmias, and other cardiovascular abnormalities
- This case has an additional risk of recurrent deep vein thrombosis or pulmonary embolism.
- It’s vital to go for a detailed preoperative assessment including ECG, echo, and lower limb doppler.
- This is the first case of PPV that undergoes neuraxial block delivery to be reported.
- Careful attention to anticoagulation management and thorough monitoring should be done to ensure optimal maternal and fetal outcomes.

Conclusion

- Neuraxial anesthesia can be done safely in pregnant patients with PPV.
- Comorbidities must be ruled out, including Sturge-Weber, and Klippel–Trenaunay, and a full systematic checkup must be performed.

Subtype of disease	Phacomatosis Cesioflammea	Phacomatosis Cesiomarmorata +/- Cesioflammea overlap	Phacomatosis spilorozea	Phacomatosis Melanorozea	Unclassifiable	Total
Ocular melanocytosis	78/115 (68%)	7/16 (44%)	0/2 (0%)	0/1 (0%)	5/15 (33%)	90/149 (60%)
Glaucoma	38/105 (36%)	4/16 (25%)	1/7 (14%)	0/1(0%)	1/12 (8%)	44/141 (31%)
Seizures	26/109 (24%)	0/12 (0%)	1/6 (17%)	0/2 (0%)	1/16 (6%)	28/145 (19%)
Brain Abnormalities ⁺	19/51 (37%)	3/9 (33%)	2/6(33%)	0/1 (0%)	¼ (25%)	25/71 (35%)
Choroidal melanoma	14/80 (18%)	0/15 (0%)	0/5 (0%)	0/1 (0%)	0/9 (0%)	14/110 (13%)
Leg length discrepancy	18/54 (33%)	2/16 (13%)	5/6 (83%)	0/1 (0%)	3/8 (38%)	28/85 (33%)
Limb Hypertrophy	36/52 (69%)	6/17 (35%)	2/5 (40%)	NR	3/9 (33%)	47/83 (57%)

References

- Van Gysel, D et al. “Phakomatosis pigmentovascularis.” Pediatric dermatology vol. 13,1 (1996): 33-5. doi:10.1111/j.1525-1470.1996.tb01184.x
- Hasegawa, Y, and M Yasuhara. “Phakomatosis pigmentovascularis type IVa.” Arch ives of dermatology vol. 121,5 (1985): 651-5.