Pulmonary embolism during pregnancy in a case of blue rubber bleb nevus syndrome

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To the Editor: Blue rubber bleb nevus syndrome (BRBNS) is a rare systemic vascular disorder characterized by multiple venous malformations involving many organs. BRBNS can occur in various organs, but the most frequently involved organs are the skin and gastrointestinal tract. Gastrointestinal lesions of BRBNS can cause acute or chronic involvement or associated with the acute PE. To avoiding uncontrolled bleeding that would be caused by the hemangiomas in the birth canals, we performed a cesarean section. We found no macroscopically visible hemangioma in the subcutaneous tissue, muscle, or fascia. A hemangioma lesion (3 cm × 4 cm) was present on the parietal peritoneum. The surface of the intestine also had the same lesion [Figure 1D]. The surgery was uneventful.

The women delivered a male newborn weighing 2870 g with an Apgar score of 10 at 1 and 5 min, indicating no abnormality. There was no evidence of BRBNS. The patient accepted routine therapy in the postoperative period, such as antibiotics prophylaxis, oxytocin, and so on. Unfortunately, she suddenly felt dizzy and suffered from severe dyspnea, followed by tachycardia (heart rate: 141 beats/min) and was characterized by declining blood pressure in the second day after the operation. The condition rapidly progressed, leading to cardiac arrest. After active resuscitation and the application of the extracorporeal membrane oxygenation (ECMO) with the help of internal physicians, the patient recovered a normal heart rate, respiration, and blood pressure and was shown to suffer from an extensive PE by CT scan [Figure 1E]. Other therapies included anticoagulants (heparin and warfarin) and antibiotics. The ECMO was applied for 10 days. The patient is now in the process of further recovery.

BRBNS is a rare disease characterized by multiple venous malformations of the skin, gastrointestinal tract, and other organs, mainly leading to recurrent intestinal bleeding and chronic anemia. In this case, involved organs included the skin, gastrointestinal tract, spleen, vulva, and vagina. Because of lacking whole-body assessment through magnetic resonance imaging (MRI) prior to birth, we were unable to determine whether the lungs or brain were involved or associated with the acute PE.

Most BRBNS cases occur sporadically, but this condition can be inherited as an autosomal dominant trait. Recent
analyses identified a locus on chromosome 9 responsible for venous malformations.\(^2\) In our case, the patient had no family history of BRBNS and her baby had no evidence of BRBNS.

Only eight case reports of BRBNS in pregnant women could be found in the current database. Kanai et al.\(^2\) described a pregnant woman with BRBNS suffering from severe gastrointestinal bleeding and accepted endoscopic therapy and partial enterectomy. Terata et al.\(^3\) reported a case during pregnancy complicated by placenta previa. Nirmal et al.\(^4\) documented a patient who was examined for venous malformations in the central nervous system with MRI for regional anesthesia. Tanaka et al.\(^1\) reported a patient with hemangiomas of the vaginal portion of the cervix, which increased in size during pregnancy. The

Figure 1: Representative images of the patient. (A) The cutaneous hemangiomas called “blue rubber bleb” involving in the vulva. (B) “Blue rubber bleb” in the skin of knee. (C) The manifestation of “blue rubber bleb” under the colonoscopy. (D) A hemangioma lesion was present on the surface of the intestine (the arrow). (E) An extensive pulmonary embolism by CT scan. CT: Computed tomography.
patient underwent an elective cesarean section at 36 weeks of gestation. Ochiai et al.\(^5\) reported a familial BRBNS in pregnancy with spinal epidural involvement, in which the patient received a cesarean section under general anesthesia. Bouchghoul and Nizard\(^6\) first reported two consecutive vaginal deliveries with epidural analgesia described for women with BRBNS. Preventive low molecular weight heparin was applied in the postpartum period because of the rising D-dimer. Vaginal delivery with epidural analgesia is believed to be an option for women with BRBNS. The remaining cases were uneventful during pregnancy and all of them underwent cesarean section, to avoid any uncontrolled bleeding resulting from a potential hemangioma in the birth canals.

In our case, the woman received an uneventful cesarean section at 36\(^{+5}\) weeks of gestation. However, she unexpectedly suffered from an extremely severe PE on the second day after the operation. ECMO was needed to address the PE. We believe that the hypercoagulable state the patient was in due to pregnancy may have contributed to the PE. It is suggested that special attention should be paid to pregnant women with BRBNS for thrombosis beyond hemorrhage, similar to the report by Bouchghoul and Nizard.\(^6\) It may be essential to monitor the D-dimer and appropriately utilize preventive low molecular weight heparin in the postpartum period. In addition, we suggest that women with BRBNS should be examined for systemic hemangioma using MRI and gastrointestinal endoscopy before pregnancy. Assessments of anemia and the clotting system are needed. Furthermore, genetic counseling is recommended.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

None.

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