# 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 bleeding and anemia.[1] Only several case reports of pregnant women with BRBNS could be found in the current database and most of them were complicated by hemorrhage of the gastrointestinal tract. We report a case of BRBNS during pregnancy that suffered from severe pulmonary embolism (PE) and was successfully resuscitated.

The patient had no family history of BRBNS. Since childhood, she manifested recurrent hematochezia and chronic iron-deficiency anemia. She also had multiple cutaneous hemangiomas involving her legs, vulva, and vagina [Figure 1A and 1B]. She was initially diagnosed with BRBNS through endoscopy at 18 years of age [Figure 1C], whereas a computed tomography (CT) scan later suggested the presence of multiple hemangiomas in the gastrointestinal and abdominal veins. The CT scan also revealed the presence of the same lesions in the spleen but not in other visceral organs such as the liver. Unfortunately, the lung and brain were not evaluated. She accepted sirolimus treatment twice in our hospital and had been symptom free for 6 years before the pregnancy. When she was 24 years old, she became pregnant for the first time. At 18 weeks of gestation, she developed colporrhagia, leading to anemia. The patient's hemoglobin level decreased to 65 g/L. She then received a transfusion of concentrated erythrocytes twice, for a total of 800 mL and was given iron agent supplement simultaneously. The intrauterine fetal growth was normal. No other event occurred. At 36 5/7 weeks of gestation, she felt labor pains and was admitted to our hospital on June 8, 2018. Her hemoglobin level increased to 97 g/L. The platelet count and clotting test were normal, but her fibrin level was distinctly higher than before, similar to that in other normal pregnant women. The liver and kidney examinations were normal. 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 beat 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.[1] 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

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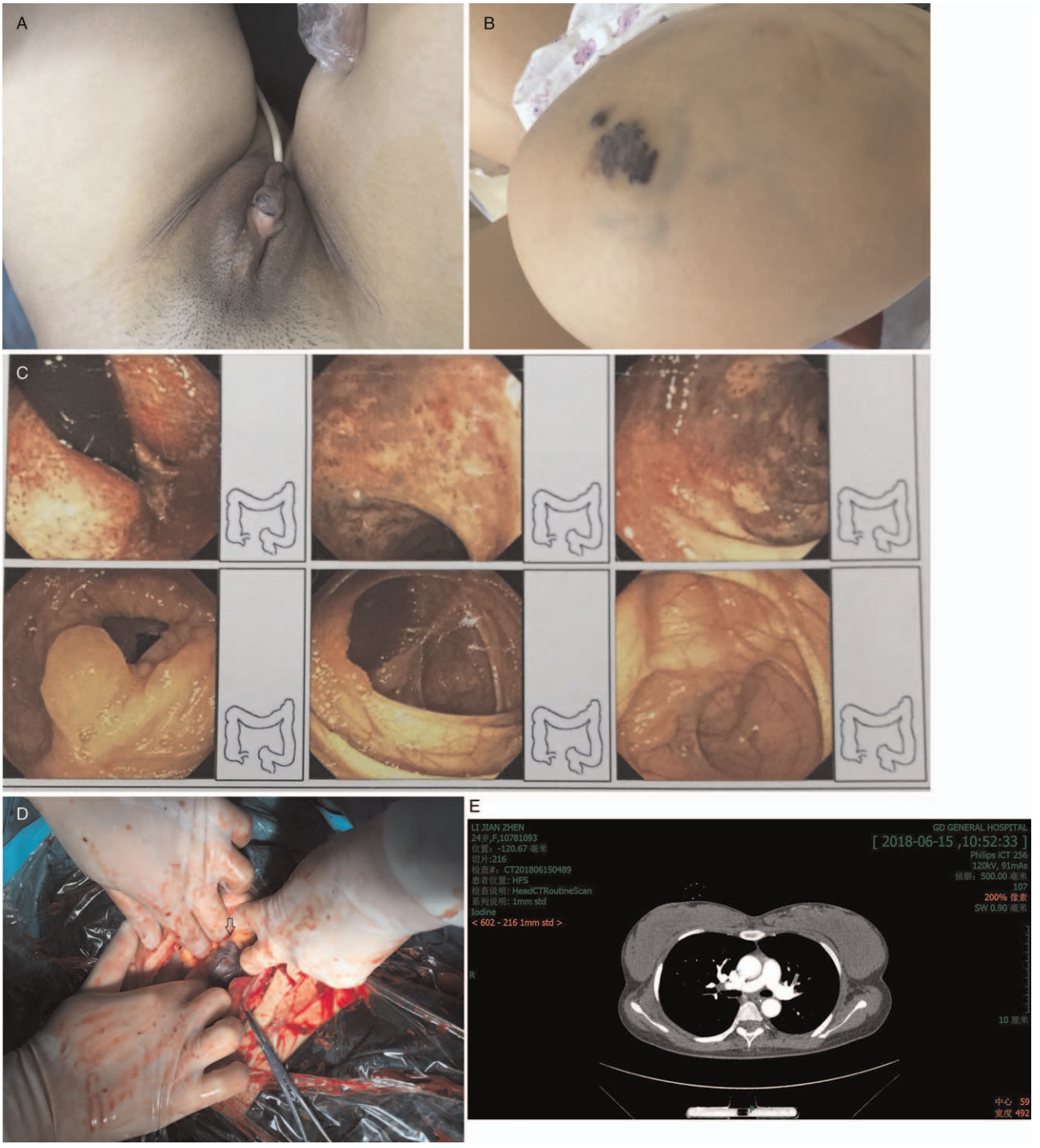


**Figure 1:** Representative images of the patient. (A) The cutaneous hemangiomas called "blue rubber bleb" involving in the vulva. (B) "Blue rubber bleb" in the skin of knee. (C) The manifestation of "blue rubber bleb" under the colonoscopy. (D) A hemangioma lesion was present on the surface of the intestine (the arrow). (E) An extensive pulmonary embolism by CT scan. CT: Computed tomography.

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

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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