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## Blue rubber bleb nevus syndrome: New case treated with sirolimus

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**Introduction:** Blue Rubber Bled Nevus Syndrome (BRBNS), is a very rare disease characterized by multiple venous malformations in the skin and, less often, in other organ systems. Herein, we report a case with BRBNS in a 22- year old female treated with sirolimus.

**Case Report:** An 22-year-old girl with a 11-year history of skin lesions, fatigue, palor and massive melena. There was the first-degree consanguinity between his parents. Some lesions were at birth and have gradually increased in size and number. She has been hospitalized numerous times for this problem and has undergone many blood transfusions since the age of 6 months. Although the patient had been taking iron supplements, his hemoglobin concentration has been between 3 and 9 g/ dl. The patient has been operated for intussusception. Later in life, the lesions increased in number and size and she presented with multiples cutaneous angiomas over buttocks, tongue and extremities. On physical examination, blood pressure 100/60 mm Hg, weight 45 Kg and no malnutrition signs were present. The lesions were mostly soft, rubbery, compressible, dark blue papules, nodules on the tongue, buttocks, arms, legs and fingers. The patient's abdomen is soft nontender and no hepatosplenomegaly. The patient's hemoglobin level of 9g/dl suggesting iron deficiency anemia (ferritin is 6.1). Platelets count and urinalysis results are normal. Endoscopic examination showed multiple congested, nodular lesions in the duodenum, ileum and colon. As part of the extension assessment, angiography CT scan of thorax, abdomen and pelvis and brain MRI revealed hepatic and intestinal angiomas with the left cerebral vascular hemangioma. BRBNS is diagnosed on the basis of clinical and endoscopic findings. After obtaining the informed consent, oral sirolimus at 1,6 mg /m2/d (2 mg/d) is initiated as an antiangiogenic agent. One month later, the patient hemoglobin concentration increased at 11 g/dl, melena stopped, the size of some lesions decreased but did not disappear. The sirolimus dose was titrated to maintain a plasma concentration between 1-5 ng/ml. We observed no adverse effects or complications, no control endoscopy or abdominal imaging has been performed (no GI bleeding).

**Discussion:** BRBNS is one of the vascular development dysplasia diseases characterized by multiple distinctive cutaneous, gastrointestinal (GI) and many parts of the body's venous malformations. BRBNS is usually reported in adults and most cases are sporadic. Cutaneous lesions are divided into 3 types. The most common complication is severe iron deficiency anemia, due to chronic blood loss; patients often require a prolonged iron supplementation and sometimes regular blood transfusions. Severe GI bleeding is the most difficult complication to manage. No curative therapy is currently available for BRBNS. A few approaches have been used, such as blood transfusion and iron supplementation agent is utilized for the remedy of anemia. Currently, the BRBNS cases with sirolimus treatment around the world are rare and the dose of sirolimus and the period of the treatment are uncertain. we report a case successfully treated with sirolimus as an alternative to blood transfusion in order to save patients life.

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