Cutis marmorata telangiectatica congenita (CMTC) is a rare cutaneous vascular condition found in newborns. Its extraordinary infrequency is reflected in the fact that only 300 cases have been reported worldwide.[8] At birth, CMTC manifests as a pinkish reticulated pattern all over the body mimicking cutis marmorata; however, unlike cutis marmorata, the lesions do not improve with warming.[9] The lesions of CMTC gradually lighten as the patient ages.[8] Limb asymmetry is the most common extravascular complication of CMTC and, similar to SWS, glaucoma also can occur.[10] Cutis marmorata telangiectatica congenita has been known to occur simultaneously with SWS or IH, but the combination of all 3 conditions in our patient is unique. Due to the scarcity of cases, the pathophysiology and treatment is poorly understood, with appropriate monitoring for sequelae recommended.[9]

## Case Report

The patient was born at 39 weeks' gestation following an uncomplicated pregnancy and delivery. She weighed 2950 g, her length was 19 in, and her head circumference was 13.25 in, correlating to the 10th, 50th, and 25th percentiles, respectively. Her Apgar score was 8/9 at 1 and 5 minutes. Her parents were nonconsanguineous and in good health. The patient's family lived in poverty, which led us to conjecture about the role that toxins played in the epigenetics of the patient and her family. It was the mother's third pregnancy; both prior pregnancies resulted in healthy children. The patient was breastfed. No family history of heritable vascular disorders was reported.

On the first day of life during the newborn examination, dark red pigment changes were noticed under the nose and erythematous pigmentation over the whole body was observed (Figure). On examination, 2-toned reticular lesions identified as extensive nevus flammeus were found bilaterally over the distribution of the ophthalmic division of the trigeminal nerve. A separate erythematous plaque over the maxilla also was recognized. The pediatrician suspected SWS and facial IH. The patient was discharged after 3 days with a referral to pediatric dermatology, and appropriate follow-up with a pediatrician was scheduled. The patient returned for these appointments and the significance of SWS was explained to her parents. Consultation with pediatric dermatology at 2 weeks of age confirmed the diagnosis of SWS as well as facial IH.

Upon further follow-up with pediatric dermatology at 2 months of age, the patient received an additional diagnosis of CMTC. These exceedingly rare lesions were located over the back, trunk, arms, and legs. The patient's parents were counseled about the management of these conditions and appropriate follow-up.

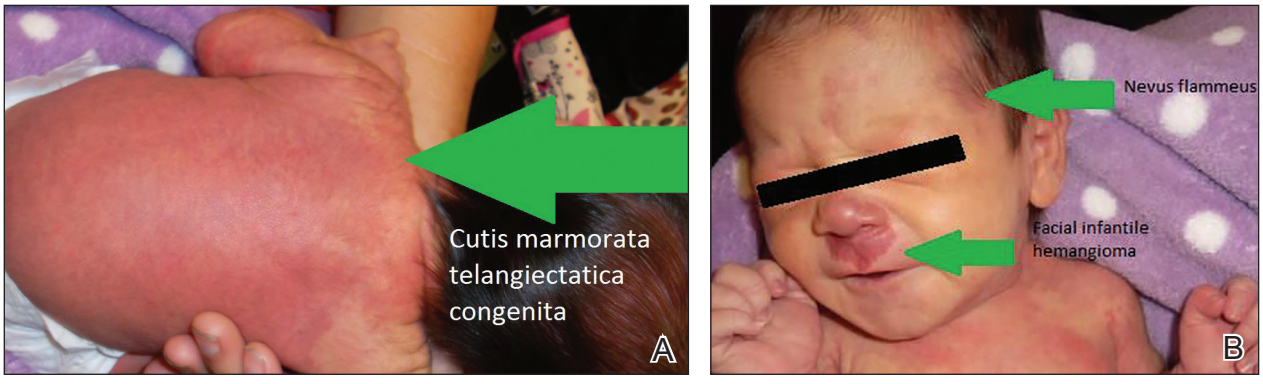
## Comment

This case describes 3 different vascular malformations in the same patient. Cutis marmorata telangiectatica congenita is rare and not described in this patient along with 2 other notable endothelial abnormalities. The clinical interest of this case is heightened by the presence of CMTC.

The causative factor of SWS is a well-documented mutation of the *GNAQ* gene, but there is considerable variability in how it affects the patient. Unlike in SWS, no single factor can be attributed to the development of IH. This case shows that these 3 diseases are not mutually exclusive and can present with unusually severe features when they occur concomitantly. The embryologic basis of SWS traces its roots back to the first trimester during vascular development, where lack of regression and development of the primitive cephalic venous plexus occur.[10] The presence of a large IH on the patient's philtrum that demonstrated markers of pericyte and neural crest cells illustrates that the developmental origins of one neurocutaneous disorder do not necessarily interfere with the development of other cutaneous conditions.[11]

The severity of the SWS in our patient was highlighted by the extensive nevus flammeus. These lesions occurred over the face, trunk, arms, and legs. The port-wine stain






A diffuse purple and pink reticular pattern over the entire back can be observed in a patient with concomitant Sturge-Weber syndrome, facial infantile hemangioma, and cutis marmorata telangiectatica congenita (A). On the face, an infantile hemangioma and distribution of nevus flammeus over the frontal bone denoted Sturge-Weber syndrome (B).