**Fluorescein Angiogram Findings in a Case of Cutis Marmorata Telangiectatica Congenita**

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**ABSTRACT:** Cutis marmorata telangiectatica congenita is a well-characterized cutaneous vascular disorder with variable and rare ocular involvement. It has been reported in association with glaucoma, bilateral congenital retinal detachments, bilateral tractional retinal detachments secondary to proliferative vitreoretinopathy, and retinoblastoma. This case demonstrates novel findings of bilateral peripheral retinal vascular abnormalities and retinal nonperfusion on fluorescein angiography without retinal detachment that have not previously been described in cutis marmorata telangiectatica congenita. Laser photocoagulation was applied to areas of retinal nonperfusion with stability in the retinal pathology at follow-up examination 3 months later.


**INTRODUCTION**

Cutis marmorata telangiectatica congenita is a rare cutaneous vascular disorder with variable phenotypic features that may include ophthalmic manifestations. There is no known etiology or genetic origin. Diagnosis is made on clinical grounds; although no formal criteria exist, major and minor criteria for diagnosis have been proposed.¹ Patients with cutis marmorata telangiectatica congenita (CMTC) invariably demonstrate cutaneous reticulate erythema that does not respond to local warming. There are many reported associations, including syndactyly, nevus flammeus, focal cutaneous atrophy, and macrocephaly, among others.¹,² Ocular findings are infrequent, with glaucoma being the most common.¹ In this report we present a rare case of an infant with CMTC and bilateral peripheral retinal vasculopathy and retinal nonperfusion.

**CASE REPORT**

An 11-month-old female infant with a history of CMTC was referred for retinal exam under anesthesia. The patient was born at 36 weeks of gestation and had no history of prior ocular abnormalities. Anterior segment examination was unremarkable. Dilated fundus exam revealed abnormal peripheral retinal vasculature without retinal detachment that have not previously been described in cutis marmorata telangiectatica congenita. Laser photocoagulation was applied to areas of retinal nonperfusion with stability in the retinal pathology at follow-up examination 3 months later. Fluorescein angiography was performed using a RetCam system (Clarity Medical Systems, Pleasanton, CA) and demonstrated microaneurysms, telangiectatic vessels, and an abnormal branching pattern with anastamotic vessels (Figures 1 and 2). Both eyes had significant areas of bilateral nonperfusion. There was no exudate or retinal detachment on initial examination; however, the significant areas of bilateral nonperfusion were believed to confer an increased risk of exudation, neovascularization, and retinal detachment. Informed consent was obtained and bilateral laser photocoagulation was applied to the areas of nonperfusion. Follow-up examination with fluorescein angiogram 3 months later demonstrated adequate laser without neovascularization, exudate, or leakage (Figures 3 and 4). No progression of the areas of nonperfusion occurred. No additional treatment was recommended at that time.

**DISCUSSION**

CMTC may include phenotypic features such as patchy cutaneous telangiectasias, dental abnormalities, and developmental delays.¹,³ Ophthalmic mani-
Festations are rare; there are no pathognomonic ocular findings. Multiple reports have identified an association between CMTC and congenital glaucoma.4-9 Most reports of glaucoma associated with CMTC are diagnosed in infancy, although there is a report of a patient with CMTC diagnosed with glaucoma at the age of 9 years.10

Shields et al described a case of CMTC with bilateral congenital retinal detachment and secondary neovascular glaucoma.11 In 1997, Pendergast et al reported a case of CMTC associated with bilateral exudative vitreoretinopathy with bilateral tractional retinal detachment in a 3-year-old girl.12 Others have also reported an association with retinal detachment.13 Schwartz et al reported an unusual case of CMTC with macrocephaly in which the patient developed retinoblastoma; the authors postulated that macrocephaly-CMTC confers an increased risk of neoplasm.14

Various other systemic and ocular diseases can present with similar funduscopic and angiographic findings, including incontinentia pigmenti, familial exudative vitreoretinopathy, Norrie’s disease, retinopathy of prematurity, and Coats’ disease. In this case, diagnosis was made based on the presence of classic skin lesions.1

To our knowledge, this is the first case report of CMTC with bilateral retinal vascular abnormalities and retinal nonperfusion without associated retinal detachment. The natural history of this entity is unknown, and it is unclear how laser photocoagulation applied to the areas of retinal nonperfusion impacts the course of this disease. It is reasonable to consider
deferring laser treatment in favor of observation if circumstances allow for serial examination.

REFERENCES