Congenital Glaucoma and Cutis marmorata telangiectatica: Report of the Second Case

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To the editor

Described in 1922 [1], cutis marmorata telangiectatica congenita (CMTC) is a very rare disorder. It is characterized by: persistent cutis marmorata, phlebec-tasia and telangiectasias. Ulcers and atrophy are less frequently seen [2]. The disorder is usually present at birth or few months later. It is generally agreed that CMTC improves with age. Up to now, no recognizable pattern of inheritance has been reported. Skin biopsy shows only dilatation of blood vessels with increase in number. In about 50% of patients, it tends to be associated with various congenital anomalies, being hypertrophy or atrophy of an involved extremity the most frequently associated anomaly [3]. Congenital glaucoma has been described in only 1 patient [4]. We had a new case that we consider of interest to communicate.

This Caucasian boy was the first child of a 26-year-old woman. No family history was evident and no consanguinity between parents was related. At birth, the skin surface showed widespread reddish-blue reticular network (fig. 1). The extremities manifested this pattern most severely. Ulcers, phlebeectasies or masses were not observed. The lesions increased with cold and crying and blanched with pressure. Mucous membranes were not involved. Extremities were equal in size and development. Physical examination was otherwise normal with the exception of right ocular glaucoma diagnosed by intraocular pressure measurements. Results of the following tests were normal: Chemistry group, complete blood count, differential count, uri-nalysiss, chest, skull, abdomen, spine and upper and lower limb X-rays were also normal as were brain scan, abdominal echography and EKG. Skin biopsy was not done.

Follow-up examination during 2 years revealed persistence of the reticular pattern, without developing ulcers or atrophy. Development alterations, mental retardation or visual disturbances were not observed. Goniotomy was performed as treatment of the congenital glaucoma.

This is the second report of congenital glaucoma associated with CMTC. In the first report [4], glaucoma was bilateral and other anomalies were present

Fig. 1. Skin surface with a widespread reddish-blue reticular network.

(ductus arteriosus, mental retardation, nevus telangiectaticus). Skin involvement was not very serious, without ulcers or atrophy, and only slight improvement was observed after the patient was 14 months old. Our case showed also minor cutaneous manifestations, without improving after 2 years' follow-up. Glaucoma was unilateral and no other anomalies were detected. Mental retardation, visual disturbances or cardiac anomalies were not observed during evolution. Histologically, CMTC is similar to nevus telangiectaticus. Petrozzi [4] considers his case as an incomplete form of Sturge-Weber disease, supporting the belief that this syndrome, together with Von Hippel-Lindau, Klippel-Trenaunay and CMTC are diseases manifesting telangiectatic nevi associated with other developmental defects, representing defects of the mesodermal system during embryonic life.

Nevertheless, more cases need to be reported before the relationships and differences among them can be understood.

References

1 Von Lohuize C: Cutis marmorata telangiectatica congenita. *Acta Derm Venereol* 1922;3:202-211.

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Powell ST, Su DWP: Cutis marmorata telangiectatica congenita: Report of nine cases and review of the literature. *Cutis* 1984;34:305-312.

Way BH, Herrman J, Gilbert EF, et al: Cutis marmorata telangiectatica congenita. *J Cutan Pathol* 1974;1:10-25.

Petrozzi JW, Rahn EK, Mofenson H, et al: Cutis marmorata telangiectatica congenita. *Arch Dermatol* 1970;101:74-77.

Effect of the Local Application of Ciclosporine A on Chronic Erosive Lichen planus of the Oral Cavity

To the Editor

Chronic erosive lichen is a painful and debilitating disease and its therapeutic management is troublesome. Indeed, topical steroids or retinoic acid are usually inefficient. Beneficial effects of dapsone and griseofulvin have been only occasionally reported [1,2]. Systemic steroids or retinoids are reported by most authors to give satisfying results [3]; however, high doses are often necessary for several months leading to poorly tolerated side effects [4]. For this reason, we have attempted to treat four patients with locally applied ciclosporin A (CsA) in an open trial. The criteria for choosing these patients were as follows: presence of histologically confirmed erosive lichen; lesions covering more than 30% of the oral

Fig. 1. The 2nd patient had an extensive erosive lichen planus covering all the inner cheeks before treatment.

cavity; chronic evolution for more than 6 months; resistance to locally applied treatments; absence of concomitant therapy known to modify the bioavailability of CsA or to exert additive nephrotoxic effects; absence of a previous convulsive crisis; normal creatinemia and liver function; normal blood pressure (systolic < 140 mm Hg, diastolic < 90 mm Hg). The 4 patients thus selected were: (1) a 79-year-old male with coronary insufficiency and, for the past years, an atrophic and erosive lichen affecting 60% of the oral cavity; (2) a 52-year-old female with an erosive lichen affecting 50% of the oral cavity for the past 4 years (fig. 1); (3) a 31-year-old female with an erosive lichen affecting 70% of the oral cavity for the past years; it appeared 3 years after an epidermoid carcinoma on a lichen planus of the tongue, and (4) a 44-year-old female with a lichen affecting more than 40% of the oral cavity which relapsed immediately after three pulse injections of 1 g of methylprednisolone.

For at least 1 month, these patients applied four times a day 25 mg of CsA in an oil solution with their fingers to the lesions and rinsed their mouth 15 min later. This treatment was supervised as follows: blood pressure taken twice weekly; blood creatinine measured on days 0, 15 and 30; differential white blood count, sedimentation rate, dosages of transaminases, alkaline phosphatases and bilirubin, blood and urine electrolytes, and proteinuria measured on days 0 and 30; RIA dosage of CsA blood levels twice weekly, 2 h after local application of CsA.

The effectiveness of this therapy was evaluated after 1 month of treatment and 2 months later. At the end of the treatment period, an improvement of the lichen was observed in all cases with a decrease in the eroded area by more than 80% in 2 patients, 50% in the 3rd and 30% in the 4th.

Two months later, only one erythematous lichen existed in 3 patients and erosion had disappeared (fig. 2). No side effects were noted. A discrete systemic presence of CsA was recorded at the onset of the treatment (serum level 30 and 150 ng/ml, subsequently negative).

We found these initial results worth reporting in spite of the small number of cases. Erosive lichen is a chronic disease which is particularly resistant to treatment. The follow-up for 2 patients now exceeds 1 year.

This efficiency of CsA can be explained by the pathogenesis of lichen planus. A pathologic role of autoreactive cytotoxic cells is suggested by the fact that lichen planus-like skin lesions could be induced by certain autoreactive cytotoxic clone T cells in syngeneic