GeneReview Title: Lipoid Proteinosis – Management, Treatments of Unproven Efficacy

Authors: Vahidnezhad H, Youssefian L, Uitto J

Posted: January 2016

Dermabrasion may improve the appearance of the skin, chemical skin peeling, and blepharoplasty [Bannerot et al 1998].

Carbon dioxide laser surgery has been successful in treating thickened vocal cords and beaded eyelid deposits in some studies [Rosenthal et al 1997, Kroukamp et al 2007, Calıskan et al 2015].

D-penicillamine. In a 13-year-old girl treated with 600 mg/day of D-penicillamine for two years, reported decreased itching, less hoarseness, and softening of the cutaneous lesions [Kaya et al 2002].

Dimethyl sulfoxide (DMSO). Results for use of DMSO are contradictory. A 41-year-old man reported successful treatment of skin lesions, hoarseness and abnormal esophageal function using oral DMSO60 mg/kg/day for three years. In contrast, subsequent reports failed to reproduce the results in three patients treated for an average of three years [Wong et al 1998, Ozkaya-Bayazit et al 1997].

Corticosteroids. Topical corticosteroids in a 4-year-old girl with eroded lesions resulted in significant healing of skin lesions and prevented new lesion formation [Kaya et al 2003].

Etretinate. A 30-year old woman showed significant improvement in lesions on the knees, elbows, and palms at a dose of 1.0 mg/kg daily for the first two months and 0.75 mg/kg daily for the next two months [Gruber et al 1996].

Acitretin. 0.5 mg/kg/day in a 3-year-old with erosive and vesiculobullous lesions improved hoarseness in the sixth month of treatment, but did not change cutaneous lesions significantly [Toosi & Ehsani 2009].

A 21-year-old treated with acitretin 0.5 mg/kg/day for six months showed improved hoarseness but no change in cutaneous lesions [Gündüz et al 2012].

Two patients showed some regression and softening of skin lesions with acitretin; however, no histopathologic change in PAS-positive deposition could be detected [Akoglu et al 2011].

In ten patients treated with acitretin 0.5 mg/kg/day for six months, the following results were noted: cutaneous papules and plaques disappeared (7), hoarseness receded (7), vesiculobullous lesions were reduced (3), and the frequency of oral ulcers decreased (3), palmoplantar hyperkeratosis disappeared completely (1) [Dertlioğlu et al 2014].

References

Akoglu G, Karaduman A, Ergin S, Erkin G, Gokoz O, Unal OF, Hamada T. Clinical and histopathological response to acitretin therapy in lipoid proteinosis. J Dermatolog Treat. 2011 Jun;22(3):178-83.

Bannerot H, Aubin F, Tropet Y, Najean D, Blanc D. [Lipoid proteinosis: importance of dermabrasion. Apropos of a case]. Ann Chir Plast Esthet. 1998;43:78-81.

Çalıskan E, Açıkgöz G, Tunca M, Koç E, Arca E, Akar A. Treatment of lipoid proteinosis with ablative Er:YAG laser resurfacing. Dermatol Ther. 2015;28:291-5.

Dertlioğlu SB, Çalık M, Çiçek D. Demographic, clinical, and radiologic signs and treatment responses of lipoid proteinosis patients: a 10-case series from Sanlıurfa. Int J Dermatol. 2014;53:516-23.

Gruber F, Manestar D, Stasic A, Grgurevic Z. Treatment of lipoid proteinosis with etretinate. Acta Derm Venereol. 1996;76:154-5.

Gündüz O, Sahiner N, Atasoy P, Senyücel C. Acitretin treatment for lipoid proteinosis. Case Rep Dermatol Med. 2012;2012:324506.

Kaya TI, Kokturk A, Tursen U, Ikizoglu G, Polat A. D-penicillamine treatment for lipoid proteinosis. Pediatr Dermatol. 2002;19:359-62.

Kroukamp G, Lehmann K. Treatment of laryngeal lipoid proteinosis using CO2 laser. S Afr Med J. 2007;97:90-2.

Ozkaya-Bayazit E, Ozarmağan G, Baykal C, Uluğ T. [Oral DMSO therapy in 3 patients with lipoidproteinosis. Results of long-term therapy. Hautarzt. 1997;48:477-81.

Rosenthal G, Lifshitz T, Monos T, Kachco L, Argov S. Carbon dioxide laser treatment for lipoid proteinosis (Urbach-Wiethe syndrome) involving the eyelids. Br J Ophthalmol. 1997;81:253.

Toosi S, Ehsani AH. Treatment of lipoid proteinosis with acitretin: a case report. J Eur Acad Dermatol Venereol. 2009;23:482-3

Wong CK, Lin CS.Remarkable response of lipoid proteinosis to oral dimethyl sulphoxide. Br J Dermatol. 1988;119:541-4.