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DR MARIA TERESA GIURA (Orcid ID : 0000-0001-7228-4477)

DR EMI DIKA (Orcid ID : 0000-0003-3186-2861)

DR SIMONE RIBERO (Orcid ID : 0000-0002-0098-1406)

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Median canaliform nail dystrophy of Heller in a patient with atopic dermatitis: a "miraculous" healing with Dupilumab

M.T. Giura,[1] R. Viola,[2] E. Dika,[3] S. Ribero,[1] M. Ortoncelli[1]

[1] Medical Sciences Department, Dermatologic Clinic, University of Turin, Turin, Italy.

[2] Faculty of Medicine, University of Turin, Italy

[3] Unit of Dermatology, Department of Experimental, Diagnostic and Specialty Medicine, University of Bologna, Bologna, Italy.

Corresponding Author: Simone Ribero, MD, PhD

E-mail: simone.ribero@unito.it

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KEY WORDS: median canaliform nail dystrophy of Heller, nail dystrophy and atopic dermatitis, Dupilumab therapy, pruritus therapy

Median canaliform nail dystrophy of Heller in a patient with atopic dermatitis (AD) can be caused by chronic trauma due to itching. By removing the pruritus, in this case with Dupilumab, given for severe AD, the nail dystrophy was also resolved.

Dear editor,

Median canaliform dystrophy of Heller is a relatively rare nail abnormality characterized by central single longitudinal split with an inverted fir tree pattern, that commonly involves the thumb nails, rarely other finger nails.[1]

The pathogenesis is not entirely clear, the most common hypothesis is repetitive trauma to the proximal nail fold and the underlying nail matrix, also self-induced, or compression by subungual tumors. Familiar cases have been reported. But commonly the cause is idiopathic.[2]

The treatment is mainly focused on avoiding chronic nail trauma, but few cases have reported the efficacy of topical tacrolimus.[3]

A woman in her middle age affected by atopic dermatitis (AD) since childhood came to our observation for severe exacerbation of her AD, uncontrolled with the use of topical and systemic steroids or cyclosporine. In a careful clinical analysis, in addition to the AD manifestations, the presence of median longitudinal dystrophy of the right thumb nail was observed. No other fingernails or toenails were involved. The patient referred its occurrence since a couple of years (Fig 1A) and no one else in the family had this condition.

In the case of our patient the cause of the canaliform dystrophy is probably due to the chronic pruritus related to AD, as also described by Wang C. et al.[4]

Given the severity of the AD, Dupilumab therapy was administered as follows: initial dose of 600 mg followed by 300 mg every two weeks.

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Dupilumab is an IL-4 receptor subunit α (IL-4 Rα) antagonist that inhibits IL-4 and IL-13 signalling and, consequently, the entire Th2 pathway.

These interleukins would appear to be key mediators in the mechanism of chronic pruritus related to AD.

Oetjen et al. have shown that in part chronic pruritus is dependent on neuronal IL-4Rα signaling. The IL-4Rα is expressed directly on sensory neurons both in mice and in the human dorsal root ganglia and the Th2 cytokines (IL-4, IL-13 and IL- 31) directly activate this neurons; blocking IL-4Rα, chronic itching is attenuate in a murine mouse model when the same mechanism is effectively actuated.[5]

After 4 months of Dupilumab treatment was found not only an improvement of the skin manifestation, but also a complete remission of the median canaliform nail dystrophy (Fig 1B). The result obtained is probably due to the efficacy of the drug in improving the itch symptom with consequent cessation of the repeated trauma on the nail by the patient.

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Fig 1: A) Median canaliform dystrophy of Heller in a patient with AD after one month of Dupilumab (fingernails growth rate is about 3 mm/month). B) Remission of the median canaliform dystrophy of Heller after 4 month of Dupilumab

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