PSORIASIS

FIRST USE OF BIOLOGIC TREATMENT FOR PSORIASIS IN A PATIENT WITH COHEN SYNDROME

Ac Fougerousse [1]

Hia Begin, Dermatology, Saint Mandé, France [1]

Managing psoriasis in patient at risk of infection is challenging. Cohen syndrome is autosomic recessive and has been linked to mutations of the VPS13B gene. This entity associate neutropenia (responsible for chronic infections with gingivostomatitis), mental retardation and a dysmorphic syndrome that includes microcephaly, facial abnormalities, myopia, pigmentary retinitis, trunk obesity, and ligament hyperlaxity. We report the first case of a patient with Cohen syndrome and psoriasis that was treated with etanecept.
A 35 years old women with Cohen syndrome consulted for moderate plaque psoriasis resistant to topical therapy (PASI 10, BSA 15%, DLQI non measurable) with severe pruritus. She had no history of chronic infection. Phototherapy was technically impossible due to mental retardation, methotrexate and cyclosporin were contraindicated by neutropenia. Apremilast was initiated. After 3 weeks the patient developed comportemental modifications, anorexia, nausea and biologic hepatitis (ASAT 100 UI/L, ALAT 330UI/L), leading to apremilast discontinuation. A treatment with etanercept (50 mg twice a week for 3 months) was initiated after pneumococcal immunization and exclusion of tuberculosis, under clinical and biological control. After 3 months of treatment, no adverse event was reported. Neutropenia was fluctuant between 0.3G/L and 0.5 G/L, without fever or infection; hepatic transaminases were normal. PASI was 3.9, BSA 3% and there was a major effect on pruritus. Treatment was continued at regimen of 50mg of etanercept weekly.
This is, to our knowledge, the first use of biologic for psoriasis in patient with Cohen syndrome. No adverse event (especially infectious) was reported after induction phase of etanercept treatment. Further monitoring is needed for this patient at risk of infection.