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ATOPIC ECZEMA/DERMATITIS

AARSKOG-SCOTT SYNDROME AND ATOPIC DERMATITIS: A CASUAL ASSOCIATION

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Background: Aarskog-Scott syndrome, also known as Facio-digito-genital Syndrome, is a rare, X-linked disorder, predominantly affecting males, characterized by facial, skeletal and genital anomalies. Its prevalence is not known, but less than 100 cases have been reported in the literature since the first description in 1970. It is caused by mutations in the FGD1 and its typical features are round face, hypertelorism, ptosis, short stature, severly shortened distal extremities, missing teeth, scoliosis and cryptorchidism.

Observation:We report the case of a 36-year-old man affected by Aarskog-Scott syndrome with mental disability and scoliosis, presenting since childhood a severe form of atopic dermatitis with an EASI score (Eczema Area and Severity Index) equal to 36 points. The patient was previously treated with antihistamines, topical corticosteroids and cyclosporine with poor results. Based on these features, we decided to treat the patient with dupilumab, a monoclonal antibody against IL-4.

Key message: In literature, there are not other described cases of Aarskog-Scott syndrome associated with atopic dermatitis.





