Severe erythrodermic psoriasis in child twins: from clinical-pathological diagnosis to treatment of choice through genetic analyses: two case reports.


ABSTRACT

BACKGROUND: Pediatric erythroderma is a severe cutaneous disorder, which may pose diagnostic and therapeutic challenges. Psoriasis, ichthyoses, atopy, seborrhoeic dermatitis, pityriasis rubra pilaris, infections, metabolic diseases, drugs reaction, may cause erythroderma. The therapy should be tailored on each aetiology, if possible. The biochemical and metabolic imbalance should be corrected, and particular attention should be paid to the psychosocial behavior often related to this disfiguring disease.

CASE PRESENTATION: Two 3 year-old Caucasian twins have been suffering from an unmanageable erythroderma since the age of 8 months. The diagnosis of psoriasis, already remarkably expressed in the father's family in three cases of fraternal twins, could be enforced for several points. Major histocompatibility complex, class I, Cw*06 was detected in both twins; we found no transglutaminase-1, no corneodesmosin, nor any Interleukin-36 receptor antagonist gene mutations. We performed a cutaneous histology, positive immunostaining for Lympho-epithelial Kazal-type-related inhibitor, dermoscopy and reflectance confocal microscopy. The twins had previously received systemic steroids, short cycles of low-dosage ciclosporine, followed by etanercept at the dosage of 0,8 mg/kg, without reliable results. Cyclosporine was then reconsidered at a dosage of 5 mg/kg/day with close blood monitoring. After three months of treatment, consistent clearing and significant improvement of their social and psychological behaviour were achieved. After over one year of continuous therapy with cyclosporine, the twins have still maintained the result obtained. CONCLUSION: Pediatric erythroderma may pose a great challenge as a potentially life-threatening condition causing extreme distress in children, parents and pediatricians. In young patients it is mandatory to establish correct clinical and instrumental procedures, possibly supplemented by genetic analyses such as those we required, in order to determine an effective and safe therapy in terms of cost-benefit and put patients and family in the best condition to perform common daily activities.