Abstract AB1086 – Figure 1

Note: the number above each bar represents the number of patients at that dose. The ACR Pediatric 30, 50, 70, and 90 responses were defined as an improvement of at least 30% (or 50%, 70%, 90%) respectively from baseline in at least 3 of the 6 signs and symptoms variables, with no more than 1 of the remaining variables worsening by >30%. JIA signs and symptoms variables: physician’s global assessment of disease activity, CHAQ disability index score, CHAQ global assessment of well-being, number of joints with active arthritis, number of joints with limited range of motion, serum CRP or ESR.

Conclusions: Improvement in JIA signs and symptoms occurred at most assessments with limited range of motion, serum CRP or ESR.

Methods:

We present a case of a 5 year old patient with DITRA with prolonged response with tumour necrosis factor alfa inhibition with adalimumab.

We diagnosed plaque psoriasis, with an erythematous scaly dermatitis that appeared throughout the trunk. Treatment with acitretin and cyclosporin were not extended. Following anakinra was added until blood cultures were negative. Although skin lesions improved during the following days, patient was finally discharged, symptoms reappeared when decreasing the steroid dose. Three months later adiponectin and methotrexate were started, allowing the patient to end treatment with corticoids without evidence of activity of the disease.

References:


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AB1087 PROLONGED RESPONSE WITH TUMOUR NECROSIS FACTOR ALFA INHIBITION IN A 5 YEAR OLD BOY WITH SEVERE MANIFESTATIONS OF IL-36 RECEPTOR ANTAGONIST DEFICIENCY (DITRA)

D. Clemente 1, J.C. López Robledillo 1, A. Torrejón 2, A. Hernández 2, E. Villalobos 3.

1 Pediatric Rheumatology Unit; 2 Pediatric Dermatology; 3 Pediatrics, Hospital Niño Jesús, Madrid, Spain

Background: Deficiency of the interleukin (IL)–36 receptor antagonist (DITRA) is an autosomal recessive autoinflammatory syndrome caused by mutations in the IL36RN gene. Clinical manifestations of DITRA include recurrent episodes of generalised skin postulation, fever, systemic inflammation and leukocytosis. An uniformly effective treatment for DITRA has not yet been identified.

Objectives: We present a case of a 5 year old patient with DITRA with prolonged response with tumour necrosis factor alfa inhibition with adalimumab.

Methods: A five-year-old came to our dermatology clinic after worsening of a previous diagnosed plaque psoriasis, with an erythematous scaly dermatitis that extended throughout the trunk. Treatment with acitretin and cyclosporin were not effective and patient developed in few weeks a generalised erythroderma with pustules covering almost every part of his body, including palms and soles. He was admitted for the onset of fever and irritability due to painful rubbing of the skin. Family history of recurrent fevers or psoriasis were not revealed. Parents were not consanguineous.

Complete blood count showed leukocytosis with neutrophilia and thrombocytosis, with an erythrocyte sedimentation rate (ESR) of 6 mm/hr and a C-reactive protein (CRP) of 8.4 mg/dl. Biochemistry panel revealed a mild elevation of liver enzymes without other abnormalities. Anti-nuclear antibody (ANA) and rheumatoid factor were negative with normal serum immunoglobulin and complement. Blood culture grew E. Coli, S. Maltophila and S. epidermidis. Skin biopsy showed acanthosis and papillomatosis with perivascular polymorphous inflammatory cells. Genetic analyses showed a homozygous mutation in the IL36RN gene (pSer131Leu). No mutations were detected in IL1RN and CARD 14 genes.

Results: Treatment was initiated with intravenous methylprednisolone 2 mg/kg/day and subcutaneous anakinra 2 mg/kg/day. Cefotaxime and co-trimoxazole were added until blood cultures were negative. Although skin lesions improved during the following days and patient was finally discharged, symptoms reappeared when decreasing the steroid dose. Three months later adiponectin and methotrexate were started, allowing the patient to end treatment with corticoids without evidence of activity of the disease.

REFERENCES:


Disclosure of Interest: None declared


AB1088 CAPILLARY HEMOSIDERIN DEPOSITS OR EXTRAVASATIONS: A SUBTYPE OF HAEMORRHAGHET THAT ACQUIRES SEPARATE ATTENTION IN QUANTITATIVE ANALYSIS OF NAILFOLD CAPILLAROSCOPY IN CHILDHOOD-ONSET SLE

D. Schonenberg-Meinerma 1, M. van den Berg 2, A. Nassar-Sheikh Rashid 1, M. Boumans 2, M. Cutoio 2, T. Kuijpers 1, V. Smith 4, on behalf of EULAR study group on microcirculation in rheumatic diseases. 1Pediatric Hematology, Immunology, Rheumatology and Infectious diseases, Emma Childrens Hospital, Academical Medical Center (AMC). 2Department of Clinical Immunology and Rheumatology, Academical Medical Center (AMC), Amsterdam, Netherlands. 3Research Laboratory and Academic Unit of Clinical Rheumatology, University of Genova, Genova, Italy. 4Department of Rheumatology, Ghent University Hospital, Ghent, Belgium

Background: Quality of images in nailfold capillaroscopy has improved in the last years by introduction of videocapillaroscopy. Microangiopathy, as observed in capillaroscopy of SLE-patients, 1–2 can now be described by more detailed quantitative analysis. Recently, in a small cohort (n=22) of childhood-onset SLE (cSLE), we described capillary bleeding by two different subtypes: large haemorrhages and small point-shaped haemorrhages with a total count of resp. 0.2/1.5 per