<https://doi.org/10.47660/CBR.2020.17232>

TREATING SYSTEMIC JUVENILE IDIOPATHIC ARTHRITIS WITH ANTI-IL 1: EXPERIENCES FROM A PEDIATRIC RHEUMATOLOGY SERVICE IN BRAZIL

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BACKGROUND

Systemic juvenile idiopathic arthritis (SJIA), a chronic inflammatory disease, presents therapeutic challenges regarding the control of inflammation and harm resulting from associated medications, especially chronic corticosteroid use. Here we describe experiences using Canakinumab, an anti-interleukin 1 (anti-IL1) human monoclonal antibody, in four SJIA patients with intense inflammation — all experienced therapeutic failure in response to typically available medications.

RESULTS

Case 1

A 1-year-old boy was diagnosed with SJIA (fever, rash, splenomegaly, polyarthritis and increased levels of inflammatory markers), with no clinical response to nonhormonal anti-inflammatory (NHA) agents, corticosteroids and methotrexate (MTX). After receiving anti-IL6, tocilizumab (TCZ) and monthly venous immunoglobulin (Ig), he continued to present rash, fever and demonstrated adverse effects associated with steroid dependence (Cushing syndrome, multiple vertebral collapses, growth deficiency, cataract). The patient began Canakinumab therapy after receiving TCZ and Ig for one year without resolution. Two months after anti-IL1 therapy, corticosteroid use was suspended following the observation of normal clinical and laboratory parameters.

Case 2

A 2-year-old girl was diagnosed with SJIA (arthritis, hepatosplenomegaly, rash, fever and polyarthritis), with no response to AINH, corticosteroids and MTX. Upon the introduction of TCZ, she developed an anaphylactic reaction after the second dose. The administration of etanercept did not result in the control of inflammation and evidence of the deleterious effects of corticosteroids (growth deficit, Cushing syndrome, systemic arterial hypertension, obesity) was observed. She presented excellent clinical and laboratory responses to anti-IL1 therapy.

Case 3

An 11-month-old boy diagnosed with SJIA (persistent fever, arthritis, hepatomegaly, rash) did not respond to AINH, corticosteroids, MTX, Ig and cyclosporine and presented an intense inflammatory response (serum ferritin: 7,454.4 ng/mL; Hb: 6.5g), anorexia and difficulty in weight gain. Considering his severe clinical condition and age (< 2 years), anti-IL1 treatment was administered, with notable clinical and laboratory improvement.

Case 4

A patient previously diagnosed with SJIA 13 years prior, beginning at 10 months of age, had taken corticosteroids (chronic use), MTX, cyclosporine, TCZ (evolving to severe sepsis), etanercept and abatacept (partial response). After receiving anti-IL1, an excellent clinical response was observed and corticosteroid use was suspended.

CONCLUSION

The control of inflammation associated with AIJS has improved greatly. Despite the wide use of anti-interleukin 1 therapy in other countries, access remains limited in Brazil, mainly due to high cost. Nonetheless, this medication should be considered under continued difficulty in controlling inflammatory response or in patients who are refractory to other available therapies, thus avoiding the chronic and deleterious use of corticosteroids.