SBR 2019
36º CONGRESSO BRASILEIRO DE REUMATOLOGIA
CENTRO DE EVENTOS DO CEARÁ
04 A 07 DE SETEMBRO
FORTALEZA 2019
PROMOÇÃO
70 anos Sociedade Brasileira de Reumatologia
REALIZAÇÃO
SCR SOCIEDADE CEARENSE DE REUMATOLOGIA

THE EFFICACY OF CANAKINUMAB IN THE TREATMENT OF TUMOR NECROSIS FACTOR (TNF) ALPHA RECEPTOR-ASSOCIATED PERIODIC SYNDROME (TRAPS)

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BACKGROUND

TNF alpha receptor-associated periodic syndrome (TRAPS) is a rare autoinflammatory syndrome (AIS), of unknown prevalence and onset of symptoms during early infancy and childhood. It's caused by at least 93 mutations described so far, of the gene TNFRSF1A and its inheritance is autosomal dominant. This nosologic entity presents with longer febrile periods, when compared to other AIS, abdominal pain, myalgia, erythematous/urticarial rash, conjunctivitis, uveitis, headache, arthritis and pleurisy. The diagnosis is clinical and laboratorial, once excluded the autoimmune and infectious diseases. Sequencing the TNFRSF1A gene in DNA genome demonstrates mutations in 32-50% of the cases analyzed. The present study hereby describes a child affected by TRAPS, who presented considerable improvement after using canakinumab, a fully humanized monoclonal antibody of the class IgG1, with strong affinity to IL-1β.

CASE REPORT

KANPS, an 11-year-6-month-old boy, diagnosed with TRAPS in December 2016, after a three months fever, recurring in episodes of 7 to 10 days interspersed with periods no fever of 7 days. During the febrile periods (38,9 to 40º Celsius), the patient referred abdominal pain, recurring diarrhea, arthritis and diffuse urticariform rash. Laboratorial results: Hemoglobin 10.9, Hematocrit 32.2, Leukocytes 19.500, Ferritin 1500, RCP 96 mg/dL and ESR 83 mm. Genetic testing identified a variant of the NOD2 gene, considered as a risk modifier. As the initial diagnostic hypothesis was Familial Mediterranean Fever Syndrome (FMFS), the treatment was initiated wiht colchicine, but was not effective, hence corticosteroid therapy at 2 mg/Kg/day was introduced. The latter medication proved to be more efficient, however during weaning the symptoms worsened. Etanercept was initiated in July 2017 and though the fever subsided, the arthritis in wrists, ankles and knees required high doses of prednisone. In March of 2018, canakinumab was judicially solicited and then released in October of the same year. By the first week of the new treatment there was significant clinical improvement. After one month, the patient returned with laboratory results as follows: Hemoglobin 13.9, Hematocrit 37.2, Leukocytes 6.100, Ferritin 75, RCP 0 and ESR 10mm. The corticosteroid therapy was then suspended. The patient is currently undertaking his fourth bi-monthly canakinumab dose of 150 mg, with sustained clinical and laboratorial remission thus far.

CONCLUSION

The present case report demonstrates the efficacy of canakinumab in treating TRAPS refractory to anti-TNF and corticosteroid medications, while void of the common adverse effects of these substances, therefore improving the patient's and his family's quality of life.