4CPS-160 Efficacy and safety of anakinra and canakinumab for the treatment of IL-36R antagonist deficiency

L04 - Immunosuppressive agents

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Objetive

Alterations in the interleukin (IL)-1 pathway have been shown to be involved in the pathogenesis of some auto-inflammatory diseases. Deficiency of the IL-36R Antagonist (DITRA) is a recently described hereditary rare disease in which IL-1 antagonists may represent therapeutic alternatives.
Our objective is to summarize the evidence for efficacy and safety of IL-1-targeting drugs in DITRA following the scoping review's methodology.

Materials and methods

A scoping review was conducted following an a *priori* protocol based on the Joanna Briggs Institute Reviewer's Manual and the recently published PRISMA extension for Scoping Review statement. A three-step searching procedure on MEDLINE and EMBASE databases until March 2018 with additional hand searching was performed article selection, and data extraction were carried out by two researchers independently. Evidence on efficacy and safety of therapies for this disease were synthesized.

Results

Nine case reports published between 2011 and 2018 were found. All patients were treated with anakinra at 2–5mg/kg/day or 100 mg/day, and one patient was also treated with canakinumab 3 mg/kg every 8 weeks. The duration of anakinra treatment ranged from 3 days to 12 months. With regard to efficacy of anakinra, time-to-response frequencies were evaluated as immediate (7/9), short term (3/9), and medium-long (2/9). One patient, in whom anakinra had previously failed, received treatment with canakinumab, and this treatment did not prove effective at the initial time or in the short- or long-term analyses. With respect to the safety of anakinra, one case of systemic infection was reported, one of renal and hepatic laboratory abnormalities, rising white blood cell count, deteriorating clinical status with progressive skin pustulation, and pain at the injection site without erythema. No adverse events were reported in the patient who had been treated with canakinumab.

Conclusions

Evidence on the use of anti-IL-1 drugs in DITRA is scarce and based on observational studies in whom anakinra is the most commonly used drug, showing a good immediate response, but decreasing short- and medium/long-term responses. Larger studies with better methodological quality are needed.

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