PEDIATRIC RHEUMATOLOGY

MEETING ABSTRACT Open Access

PW03-006 - IL-1-B inhibition in Schnitzler's syndrome

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Introduction

Schnitzler's syndrome is a chronic disabling autoinflammatory disorder, characterised by chronic urticaria, paraproteinemia and systemic inflammation. The interleukin (IL) 1 receptor antagonist anakinra is a very effective treatment, but requires daily injection and blocks both IL-1α and IL-1β. Canakinumab is a selective human monoclonal anti-IL-1β antibody with a long half-life.

Objectives

We investigated the long-term efficacy and safety of canakinumab in Schnitzler's syndrome.

Methods

In an open-label, single-treatment arm trial, eight patients with Schnitzler's syndrome received monthly injections with 150 mg canakinumab subcutaneously for 6 months, followed by a 3-month observation period. Primary outcome was complete or clinical remission at day 14. Secondary outcome measures included inflammatory markers, quality of life, time to relapse, safety and tolerability.

Results

After stopping anakinra, patients developed moderate to severe clinical symptoms. Canakinumab induced complete or clinical remission at day 14 in all eight patients. Median C-reactive protein concentrations decreased from 169 mg/l at baseline to less than 10 mg/l on day 14 and remained low or undetectable. One patient discontinued participation on day 39 because of return of symptoms while all others remained in complete or clinical remission during the 6-month treatment period. Relapse after last canakinumab dose occurred within 3 months in four patients. For two patients, remission continued several months post-study. Five patients reported at least one adverse event, predominantly mild upper respiratory tract infections. One patient died in a traffic accident.

Conclusion

In this 9-month study, monthly 150 mg canakinumab injection was an effective and well-tolerated treatment for Schnitzler's syndrome. Our data demonstrate that IL-1β plays a pivotal role in this disease.

Disclosure of interest

None declared.

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