

Poster presentation

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Different pattern of synthesis and secretion of IL-1 β in patients with CIAS-1 and TNFRSF1A mutations responding to IL-1 blockade

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Aim

To compare the *in vitro* secretion of IL-1 β in patients carrying CIAS-1 mutations and TRAPS patients, in an effort to understand the mechanism modulating IL-1 β secretion in the different pathologies responding to anti IL-1 treatment.

Methods

Monocytes from 6 CINCA and 4 TRAPS patients selected for treatment with Anakinra were activated with 1 μ g/ml of LPS for 3 hours, at baseline and after 7 days from the beginning of the treatment. For comparison, monocytes from 24 healthy donors were also studied. Intracellular pro-IL-1 β and secreted IL-1 β were analysed by Western blotting and ELISA before and after a short exposure (15 min) to exogenous ATP that accelerates IL-1 β secretion.

Results

In healthy subjects LPS-induced IL-1 β secretion was variable but consistently ≤ 5 ng/ml and it was markedly increased by exposure to exogenous ATP (up to 20 ng/ml). Monocytes from CINCA patients secreted abnormally elevated amounts of IL-1 β after LPS stimulation (up to 40 ng/ml) that were not increased by ATP. Conversely, monocytes from TRAPS patients did not secrete more IL-1 β than healthy controls in response to LPS, but similarly to CINCA patients presented a low response to ATP.

Conclusion

Despite a similar clinical response to anti-IL1 treatment, the pattern of IL-1 β secretion of monocytes from Anakinra-responder TRAPS patients significantly differ from that observed in patients CIAS-1 mutations. This study suggests a different hierarchy in the pathogenic mechanisms leading to the inflammatory response in different diseases responsive to anti-IL-1 treatment.