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DISTURBED GENE EXPRESSION OF TLR NEGATIVE REGULATORS IN XLA PATIENTS

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X-linked Agammaglobulinemia (XLA) is a prototype primary antibody deficiency which is the most common form of primary immunodeficiency diseases. Mutated BTK in these patients affect many immune cells, immunologic responses and molecular interactions. TLRs, in a close interaction with BTK, reported being defected in different subcellular populations of PBMCs of XLA patients. In this concern, we aimed to assess LPS and CpG-A stimulatory action on TLR4 and TLR9 by measuring the activation of some TLR negative regulatory molecules' transcription and cytokine production. Higher transcripts of SOCS1 and RNF216 were found in unstimulated PBMCs of patients. Despite this, interesting patterns of TLR-induced transcription were observed: upregulation of IRAKM and SOCS1 in healthy subjects but downregulation in XLA, lack of RNF216 induction in healthy subjects while downregulation in patients and similar TNFAIP3 downregulation in both XLA and healthy subjects. Further, a lower amount of TNF-α was also produced by XLA patients PBMCs after LPS stimulation by disturbed cytokine production and dysregulated transcription of selected downstream signalling molecules. Our results strengthen the potential TLRs defect pointing out TLR involvement in the pathogenesis of different complications of XLA patients and also the scale of this defectiveness form TLRs expression to downstream signalling and cytokine production.

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