



ORAL PRESENTATION

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Interferon gamma (IFN γ) drives disease in the TLR9-mediated cytokine storm syndrome in mice

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The Cytokine Storm Syndrome (CSS) is characterized by an overwhelming activation of immune cells observed in life-threatening disorders such as familial hemophagocytic lymphohistiocytosis (fHLH) and secondary (s) HLH/macrophage activation syndrome (MAS) as well as during serious infection. However, it is not known if the CSS can be attributed to a single cytokine. Increased blood levels of interferon gamma (IFN γ) in HLH and sHLH/MAS patients potentially indicate a central role for this cytokine in the CSS. Using a mouse model that mimics an infection-driven CSS (i.e., CpG-ODN), our study showed that total IFN γ levels originating within organs are 500 to 2,000-fold higher than those measured in peripheral blood as CSS develops. Ablation of IFN γ activity in tissues led to the amelioration of the plethora of associated CSS clinical and laboratory parameters. Furthermore, the IFN γ signature gene products, CXCL9 and CXCL10, correlated with disease severity in the mouse model of CSS and patients with sHLH. Thus, anti-IFN γ targeted therapy should control diseases associated with the cytokine storm and we propose the use of CXCL9 or CXCL10 as a means to monitor total IFN γ activity in patients.

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