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7.3 Successful use of anakinra, a soluble IL-1 receptor antagonist, in pediatric rheumatic diseases associated macrophage activation syndrome/reactive hemophagocytic lymphohistiocytosis

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Background

Increased interleukin 1 (IL-1) production characterizes macrophage activation syndrome/reactive hemophagocytic lymphohistiocytosis (MAS/rHLH), a potentially lethal complication of pediatric rheumatic diseases. Standard treatment (corticosteroids, cyclosporine, +/- IVIg) is not always effective.

Objective

To test effectiveness of anakinra, a soluble IL-1 receptor antagonist, in pediatric rheumatology patients who failed to respond to standard MAS/rHLH therapy.

Methods

6 pediatric rheumatology patients (3 F:3 M); mean (range) age 8.14 (0.5–13.3) years, with MAS/rHLH were enrolled (SoJIA n = 4; Churg Strauss vasculitis n = 1; and infant onset ANCA +ve pulmonary renal syndrome n = 1). The infant patient was moribund in ICU. Histiocytosis society's 2004 criteria (HLH2004), including a T-cell activation marker, soluble IL-2 receptor (sIL2r), were used to confirm MAS/rHLH. Subcutaneous anakinra (2 mg/kg/day) was added to existing therapy (high dose IV Methylprednisolone n = 5/6; IVIg n = 6/6, and cyclosporine n = 5/6). HLH2004 specified clinical and laboratory data were collected pre, 48 hours, and 2 weeks after initiation of anakinra; including sIL2r in 3/6 patients.

Results

All patients defrevesced within 24 hours of first anakinra dose, and ventilatory and dialysis support was discontinued within 96 hours in the ICU patient. All patients recovered from MAS/rHLH by 2 weeks; 5/6 pts discontinued corticosteroids by 5 weeks. Abnormally elevated baseline sIL2r resolved by 48 hours post first dose of anakinra in all 3 patients tested.

Conclusion

1) Anakinra, in combination with corticosteroids, IVIG, +/- cyclosporine, was effective in controlling MAS/rHLH in all patients, allowing rapid discontinuation of corticosteroids in 5/6 patients.

2) Elevated baseline sIL2r level normalized rapidly following anakinra, suggesting resolution of abnormal T-cell activation.