

## LETTER TO THE EDITOR

## Macrophage Activation Syndrome Presenting Unusually with Facial Edema in a Known Case of Mixed Connective Tissue Disease

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Supriya Kushwah, Somashekhar B., Rupali Rokade. Macrophage Activation Syndrome Presenting Unusually with Facial Edema in a Known Case of Mixed Connective Tissue Disease. *Pediatr. Edu. Res.* 2025; 13(2): 51-52.

Macrophage Activation Syndrome (MAS) is an unusual and severe complication in Mixed Connective Tissue Disease (MCTD). It involves multi-organ system and can be fatal if identified late.<sup>1,2</sup>

A 17-year-old female, diagnosed with MCTD one year prior and maintained on hydroxychloroquine, mycophenolate mofetil, and low-dose steroids, presented with 7 days of fever and facial puffiness with vitals stable. On admission child had pallor, skin thickening, digital pitting edema. On admission MAS workup was negative. After 3 days of admission, the patient developed neuropsychiatric symptoms and seizures suggestive of neurolupus and MAS that was confirmed on repeat workup. She was managed with intravenous immunoglobulin (2 g/kg over 5 days), broad-spectrum antibiotics, and pulse methylprednisolone therapy followed by high-dose oral prednisolone. Cyclosporine was added for disease activity control. Child

developed concurrent renal involvement in the form of proteinuria and positive dsDNA antibodies, started on intravenous cyclophosphamide.

This case is notable for MAS at any phase of disease along with disease flare in MCTD, complicated by Central nervous system and renal lupus activity. MAS is a rare but life-threatening hyperinflammatory syndrome, more frequently reported in systemic juvenile idiopathic arthritis than in MCTD. Prompt recognition and aggressive immunosuppression are essential to prevent irreversible organ damage.

Our case underscores the importance of considering MAS in any patient with connective tissue disease presenting with unexplained fever, cytopenias, hyperferritinemia, and organ dysfunction. Early diagnosis and initiation of targeted immunosuppressive therapy can be lifesaving.<sup>3,4</sup> Child is on monthly cyclophosphamide and asymptomatic.

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**Table 1:** Relevant blood investigations during hospital stay

Date	Ferritin (ng/mL)	ESR (mm/hr)	CRP (mg/L)	D-Dimer (mg/L)	Fibrinogen (mg/dL)	LDH (U/L)	Platelets	AST (U/L)	ALT (U/L)	Triglycerides (mg/dL)
21-Apr-25	386	22	9.2	1.65	430	247	Adequate	85	21	198
29-Apr-25	9280*	27	22.1	4.98	252	–	↓ (100k)	661	264	–
02-May-25	4500*	46	<5.0	6.89	244	–	100k	646	261	–
05-May-25	2320	36	<5.0	3.5	198	–	–	545	267	–

**Figure 1:** Patient showing facial puffiness

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