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A MULTINATIONAL STUDY OF THROMBOTIC MICROANGIOPATHY IN MACROPHAGE ACTIVATION SYNDROME: A DREADFUL CONDITION WHICH IS LIKELY UNDER-RECOGNIZED

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Background: Macrophage activation syndrome (MAS) is a severe complication of rheumatologic conditions, mainly systemic juvenile idiopathic arthritis (sJIA), and is classified as a secondary form of hemophagocytic lymphohistiocytosis (HLH). Thrombotic microangiopathy (TMA) is a heterogeneous group of potentially fatal diseases characterized by microangiopathic hemolytic anemia, thrombocytopenia and organ injury. The association between TMA and HLH has been described only in single reports in renal transplant recipients¹ and in two cases of virus-induced HLH². TMA associated with MAS has never been reported so far

Objectives: To present the preliminary data from a multinational cohort of pediatric patients with MAS and TMA

Methods: The clinical charts of patients with MAS were retrospectively reviewed to identify the instances that were associated with TMA. Demographic, clinical and laboratory features at MAS and TMA onset, therapeutic interventions and outcome were collected.

Results: A total of 14 patients, 71.4% females, with MAS and TMA were collected. An underlying rheumatologic disease was reported in 9/14 (8 sJIA or sJIA-like illness and 1 systemic lupus erythematosus). The median age at MAS onset was 8.6 years. In 5 patients MAS and TMA occurred simultaneously, whereas in 4 TMA preceded and in 5 followed MAS. The main features at MAS and TMA onset are presented in Table. Low complement levels and reduced ADAMTS13 activity were observed in 75% and 43% of patients respectively. For MAS management, 85.7% of patients received high-dose corticosteroids and 71.4% cyclosporine; in 5 cases anakinra was added. TMA episodes were treated with plasma-exchange in 57.1% of patients; 7 patients were given biologics (1 rituximab and 6 eculizumab) with good results. Admission to the Intensive Care Unit was required in 85.7% of cases. All patients survived.

Conclusion: The association of MAS and TMA is a life-threatening condition, likely under-recognized. Clues to suspect TMA in a patient with MAS are the increase in LDH and decrease in platelet count out of proportion of other laboratory abnormalities, the drop in haptoglobin level, the finding of schistocytes in blood smear and the new onset of hematuria. Biologics, particularly rituximab and eculizumab, may offer an adjunctive therapeutic option for refractory cases.

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	Features at MAS onset	Features at TMA onset	P
Fever, n (%)	14/14 (100)	10/14 (71.4)	0.0978
Hepatomegaly, n (%)	10/14 (71.4)	7/13 (53.8)	0.5847
Splenomegaly, n (%)	8/13 (61.5)	6/13 (46.2)	0.6940
CNS involvement, n (%)	8/14 (57.1)	12/14 (85.7)	0.2087
Kidney involvement, n (%)	6/14 (42.9)	13/14 (92.9)	0.0128
Median values			
Hemoglobin, g/dl	9.6	8.5	0.7263
Platelet count, x 10 ⁹ /l	117	34	0.0031
Aspartate aminotransferase, U/l	109	159	0.6455
Lactate dehydrogenase, U/l	1,997	3,188	0.2585
Fibrinogen, mg/dl	273	252	0.6599
Ferritin, ng/ml	13,974	11,116	0.5485
BUN, mg/dl	42	86	0.0076
Creatinine, mg/dl	1.3	1.5	0.6031
Hematuria	-	11/14 (71.4)	-
Schistocytes on blood smear	-	8/8 (100)	-
Low haptoglobin levels	-	9/9 (100)	-

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STUDY ON SERUM DNAASE1 ACTIVITY IN PEDIATRIC ONSET SYSTEMIC LUPUS ERYTHEMATOSUS FROM A TERTIARY CARE CENTRE IN NORTH WEST INDIA

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Background: DNAase is an apoptotic endonuclease responsible for degradation of chromatin released by inappropriately cleared dead cells. DNAase1 activity in systemic lupus erythematosus (SLE) patients is lower than that in inactive disease in studies conducted in adult SLE patients from developed country. There is a paucity of data on DNAase1 activity in paediatric SLE from India.

Objectives: To measure the serum level of DNAase1 in pediatric-onset lupus patients and to correlate with the disease activity.

Methods: A cross-sectional observational study was conducted over a period of 1 year. Thirty-three consecutive children with pediatric-onset SLE were enrolled and divided into active and inactive disease activity groups based on Systemic Lupus Erythematosus Disease Activity Index (SLEDAI) score and compared the serum DNAase1 level between the two groups.

Results: Out of 33 children enrolled, 13 (39.3%) had active disease (SLEDAI score ≥ 3) and 20 (60.6%) had inactive disease activity. Mean age at diagnosis was 8.5 years and 10.2 years in active and inactive groups respectively. There is female preponderance (66.7%) in the enrolled patients. Anti nuclear antibody (ANA) was positive in 90.9% of patients. The most common pattern of ANA was diffuse pattern (48.4%). The patients in active disease activity group presented most commonly with nephritis (53.8%), rash (53.8%), arthralgia (38.5%), oral ulcer (30.8%) and central nervous system (CNS) involvement (38.4%) while patients in inactive disease activity group presented with nephritis (35%), arthralgia (35%), rash (25%) and CNS involvement (30%). Class III/IV lupus nephritis was present in 25% in active disease activity group while it was present in 23.1% in inactive disease activity group. Anti-double stranded DNA (anti dsDNA) was elevated in 53.8% in active group and 50% in