Pediatric Rheumatology

Poster presentation

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Macrophage activation syndrome (MAS) in juvenile systemic lupus erythematosus (JSLE): an underrecognized complication?

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Objective

To define the characteristics of MAS complicating JSLE.

Methods

Patients with JSLE and MAS were collected from: 1) Gaslini Institute of Genoa, Italy; 2) PRINTO and PRCSG investigators; 3) literature. Control groups of JSLE without MAS included 33 patients with active lupus seen at Gaslini Institute (SLE-GI) and 387 patients from a multinational study of damage in JSLE (SLE-MS). Clinical and laboratory features of MAS with (BM+) or without (BM-) bone marrow demonstration of haemophagocytosis were contrasted each other and with those of lupus without MAS.

Results

20 BM+ and 18 BM-patients with JSLE-associated MAS were identified. Comparison of percentage frequency of the main clinical and laboratory features of MAS in patient groups is shown in table 1.

Conclusion

Features of MAS in patients with or without BM haemophagocytosis were comparable, except for a greater frequency of leukopenia in BM+ patients. This suggests that this complication is more common than previously realized. All features but leukopenia and fever discriminated well between MAS and active lupus without MAS.

Table 1: Comparison of percentage frequency of the main clinical and laboratory features of MAS in partient groups. (NA: not available)

	MAS BM+	MAS BM-	SLE-GI	SLE-MS
Fever	95.0	83.3	21.2	64.2
Hepatomegaly	47.4	55.6	12.1	10.4
CNS dysfunction	37.5	28.6	3.0	8.5
Haemorrhages	40.0	33.3	9.1	NA
Leukopenia	90.0	44.4	63.6	NA
Thrombocytopenia	90.0	61.1	18.2	NA
Hypertransaminasemia	80.0	93.8	30.3	NA
Hypertriglyceridemia	75.0	88.2	20.0	NA
Hypofibrinogenemia	37.5	42.9	0	NA
Hyperferiitinemia	92.9	94.4	0	NA