

ONLINE SUPPLEMENTARY DATA

Supplementary Table 1. Comparison of clinical manifestation in patients with AOSD and patients with MAS. Values are expressed as n (%).

Clinical manifestation	AOSD patients (n=64)			AOSD patients with MAS (n=36)		
	Without MAS (n=28)	With MAS (n=36)	p-value	On admission (n=22)	After admission (n=14)	p-value
Fever [†]	23 (82.1)	28 (77.7)	0.669	17 (77.2)	11 (78.5)	0.928
Joint pain	23 (82.1)	22 (61.1)	0.069	14 (63.6)	8 (57.1)	0.700
Skin rash	12 (42.8)	12 (33.3)	0.438	9 (40.9)	3 (21.4)	0.233
Leukocytosis	25 (89.2)	25 (69.4)	0.058	15 (68.1)	10 (71.4)	0.838
Sore throat	16 (57.1)	19 (52.7)	0.729	11 (50.0)	8 (57.1)	0.679
Lymphadenopathy	18 (64.2)	23 (63.8)	0.974	13 (59.0)	10 (71.4)	0.458
Hepatosplenomegaly	4 (14.2)	8 (22.2)	0.423	4 (18.1)	4 (28.5)	0.471

[†]Fever was defined as a body temperature of over 39°C according to the Yamaguchi criteria for AOSD.

Supplementary Table 2. The use of glucocorticoids and other immunosuppressive agents in patients with adult-onset Still disease (AOSD).

	AOSD patients (n=64)		AOSD patients with MAS (n=36)	
	Without MAS (n=28)	With MAS (n=36)	Without in-hospital mortality (n=24)	With in-hospital mortality (n=12)
Cumulative glucocorticoid dose (mg), median (IQR)	245.0 (95.0-557.5)	2142.5 (320.0-4610.0)**	1055.0 (270-4610.0)	3337.5 (920.0-4667.5)
Glucocorticoid dose/day (mg), median (IQR)	28.6 (20.0-46.0)	86.9 (34.8-287.9)**	60.2 (25.4-209.0)	135.8 (72.7-307.1)
Steroid pulse therapy, n (%)	6 (21.4)	10 (27.7)	7 (29.1)	3 (25.0)
Immunosuppressive Agent, n, (%)				
Methotrexate	6 (21.4)	4 (11.1)	4 (16.6)	0 (0.0)
Cyclosporine	3 (10.7)	13 (36.1)*	8 (33.3)	5 (41.6)
Etoposide	0 (0.0)	6 (16.6)*	2 (8.3)	4 (33.0)
Tocilizumab	1 (3.5)	3 (8.3)	1 (4.1)	2 (16.6)
Anakinra	0 (0.0)	1 (2.7)	0 (0.0)	1 (8.3)
Infliximab	0 (0.0)	1 (2.7)	1 (4.1)	0 (0.0)
Cyclophosphamide	0 (0.0)	1 (2.7)	1 (4.1)	0 (0.0)
IV immunoglobulin	0 (0.0)	5 (13.8)*	1 (4.1)	4 (33.3)*

Values are expressed as the medians (interquartile ranges) or n (%).

IV, intravenous.

*p < 0.05; **p < 0.001 for differences between the two groups.