

**Supplementary Table S1.** The classification criteria for MAS in AOSD patients.

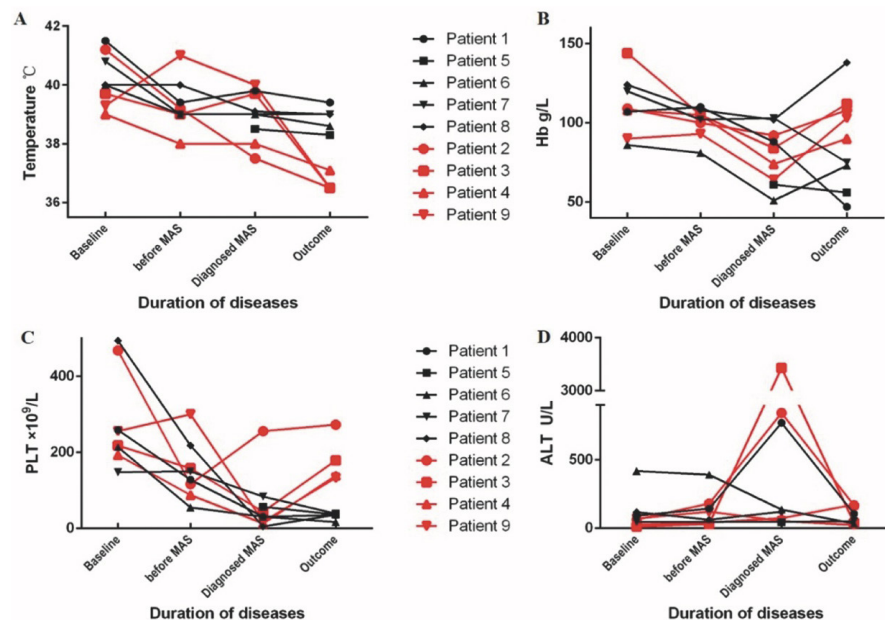
Parameter	Number of points (criteria for scoring)
Clinical features Fever	0 (<38.4), 33 (38.4–39.4), or 49 (>39.4)
Known underlying immunosuppression*	0 (no) or 18 (yes)
Organomegaly	0 (no), 23 (hepatomegaly or splenomegaly), or 38 (hepatomegaly and splenomegaly)
No. of cytopenias**	0 (1 lineage), 24 (2 lineages), or 34 (3 lineages)
Ferritin (ng/mL)	0 (<2,000), 35 (2,000–6,000), or 50 (>6,000)
Triglyceride (mmol/L)	0 (<1.5), 44 (1.5–4), or 64 (>4)
Fibrinogen (g/L)	0 (>2.5) or 30 (≤2.5)
AST (IU/L)	0 (<30) or 19 (≥30)
Haemophagocytosis features on bone marrow aspirate	0 (no) or 35 (yes)

\*Human immunodeficiency virus positive or receiving long-term immunosuppressive therapy (*i.e.*, glucocorticoids, cyclosporine, azathioprine).

\*\*Defined as a haemoglobin level of ≤9.2 g/dL and/or a WBC ≤5,000/mm<sup>3</sup> and/or a PLT count of ≤110,000/mm<sup>3</sup>. AST: aspartate aminotransferase; WBC: white blood cells; PLT: platelet.

**Supplementary Table S2.** Clinical characteristics of AOSD-MAS patients.

Patient	Fever	Peak temperature°C	Rash	Sore throat	Jaundice	Arthritis	Lymphadenophy	Hepatomegaly	Splenomegaly
1	+	41.5	+	-	+	+	-	-	-
2	+	41.2	-	-	-	+	-	-	-
3	+	39.7	+	-	+	-	+	-	-
4	+	39.4	+	-	-	+	-	-	+
5	+	38.5	+	-	-	+	-	-	-
6	+	40.0	-	-	+	-	-	-	+
7	+	40.8	+	+	+	+	+	-	-
8	+	40.0	+	+	-	-	+	-	+
9	+	41.0	+	-	+	-	-	-	-



**Supplementary Fig. S1.** Change of clinical characteristics in AOSD-MAS patients.

1A: Temperature; 1B: Hb; 1C: PLT; 1D: ALT.

**Data availability**

The datasets generated during and/or analysed during the current study are available from the corresponding author on reasonable request.

**Ethical Committee Approval**

The Ethics Committee of the First Hospital of Jilin University approved the research. This work was conducted according to the provisions of the Declaration of Helsinki. Written informed consent was obtained from all patients. The committee’s reference number: 2013-268.