

Supplementary Table S1. Clinical features of EBV-positive patients analysed in this study.

Case	Sex	Age at onset of MAS, year	Underlying rheumatic disease	Duration of underlying disease	The time of EBV infection	EBV-DNA (serum)	EBV-DNA (PBMC)	EBV-infected lymphocyte subsets	Initial treatment	Initial treatment efficacy	Refractoriness or relapse	Salvage treatment and efficacy	HSCT	EBV-DNA at the end of treatment	Outcome at last follow-up
1	Female	48	CTD	5 months	During MAS diagnosis	-	6.5E+05	ND	MP, CsA	PR	Relapse	DEP, PR	No	Negative	PR
2	Female	28	AOSD	8 months	During MAS diagnosis	1.9E+03	1.0E+04	B	MP, IVIG	NR	Refractoriness	R+DEP, PR	No	Negative	CR
3	Female	23	AOSD	2 months	During MAS diagnosis	<5.0E+02	8.8E+02	B	Ru, DEP	PR	None	-	No	Negative	CR
4	Male	64	AAV	8 years	During MAS diagnosis	<5.0E+02	1.3E+04	B	MP	PR	Relapse	R+MP+Ru, relapse after PR	No	Negative	Death due to gastrointestinal bleeding causing by MAS progression
5	Female	39	CTD	5 months	During MAS diagnosis	<5.0E+02	2.1E+03	B	MP	CR	Relapse	Ru, MP	No	Negative	CR
6	Female	62	AOSD	1 months	During MAS diagnosis	-	1.4E+04	B	HLH-94	PR	Refractoriness	R+DEP, PR	No	Negative	CR
7	Female	31	AOSD	9 years	After MAS treatment	<5.0E+02	2.8E+04	B	Dex, CsA	PR	Relapse	DEP, PR	No	Negative	Death from infection
8	Female	54	AOSD	3 months	During MAS diagnosis	-	1.1E+03	B	MP, CsA	CR	None	-	No	Negative	CR
9	Female	46	CTD	2 years	During MAS diagnosis	<5.0E+02	5.7E+04	NK and/or T	HLH-94	NR	Refractoriness	DEP, PR	No	Positive	Death from worsening of HLH
10	Female	74	RA	12 years	During MAS diagnosis	<5.0E+02	1.3E+03	NK and/or T	MP, IVIG	PR	Relapse	-	No	Positive	Death from worsening of HLH
11	Female	26	AOSD	3 months	After MAS treatment	<5.0E+02	3.5E+04	ND	HLH-94	NR	Refractoriness	-	No	Positive	Death from worsening of MAS
12	Male	30	AOSD	5 years	During MAS diagnosis	1.8E+05	1.3E+05	NK and/or T	Ru, L, DEP	NR	Refractoriness	-	No	Positive	Death from worsening of HLH
13	Male	35	AAV	6 months	During MAS diagnosis	<5.0E+02	4.7E+03	NK and/or T	DEP	PR	Relapse	-	Yes	Positive	Death from relapse after HSCT
14	Female	50	AOSD	2 months	After MAS treatment	<5.0E+02	7.3E+03	B	MP, CsA	NR	Refractoriness	HLH-94, PR	No	Negative	CR
15	Female	79	SLE	4 months	After MAS treatment	6.5E+02	<5.0E+02	ND	HLH94	NR	Refractoriness	-	No	Positive	Death from worsening of MAS
16	Female	15	SLE	1 months	During MAS diagnosis	-	6.6E+02	B	MP, CsA	CR	Relapse	Clinical trial	No	Negative	CR
17	Female	44	SLE	8 years	During MAS diagnosis	3.2E+03	4.6E+04	ND	MP	NR	Refractoriness	DEP, CR	No	Negative	CR
18	Female	47	AOSD	8 months	After MAS treatment	-	1.1E+03	ND	HLH-94	NR	Refractoriness	DEP, PR		Negative	CR
19	Male	53	CTD	8 months	During MAS diagnosis	<5.0E+02	1.9E+03	B	MP	PR	Relapse	R+DEP, relapse after PR	No	Positive	Death from worsening of MAS
20	Female	46	AOSD	3 months	After MAS treatment	-	6.2E+02	ND	MP	NR	Refractoriness	DEP, relapse after PR	No	Positive	Death from worsening of MAS
21	Female	29	CTD	2 years	During MAS diagnosis	<5.0E+02	5.2E+03	B	MP	CR	-	-	-	-	Loss to follow-up

AOSD: adult-onset Still's disease; CTD: connective tissue disease; SLE: systemic lupus erythematosus; RA: rheumatoid arthritis; AAV: ANCA-associated vasculitis; MP: methylprednisolone; DEX: dexamethasone; IVIG: intravenous immunoglobulin; L: PEG-asparaginase; Ru: ruxolitinib; R: rituximab; DEP: doxorubicin-etoposide-methylprednisolone regimen; ND: no done; CR: complete response; PR: partial response; NR: no response; HSCT: haematopoietic stem cell transplantation.

Supplementary Table S2. Clinical characteristics of MAS patients with simultaneous detection of EBV infection and rheumatic disease during aetiological screening.

Case	Sex	Age at onset of MAS, year	Underlying rheumatic disease	EBVDNA (serum)	EBVDNA (PBM)	EBV-infected lymphocyte subsets	Initial treatment	Initial treatment efficacy	Refractory or Relapse	Salvage treatment and efficacy	EBVDNA at the end of treatment	HSCT	Outcome at last follow-up
1	Female	22	SLE	<5.0E+02	2.1E+03	B	MP	PR	No	-	Y	No	CR
2	Female	54	AOSD	-	1.1E+03	B	DEX	PR	No	-	Y	No	CR
3	Male	15	SLE	-	3.2E+04	B	DEP	PR	No	-	Y	No	CR
4	Female	69	CTD	<5.0E+02	2.0E+03	B	MP	NR	Refractoriness	DEP, PR	Y	No	PR
5	Female	19	CTD	<5.0E+02	2.9E+03	B	DEX	NR	Refractoriness	R+DEP, CR	Y	No	CR
6	Female	65	CTD	3.2E+04	1.4E+06	NK and/or T	PD-1 monoclonal antibody	NR	Refractoriness	PD-1 monoclonal antibody, Ru	N	No	PR
7	Female	30	CTD	1.4E+05	2.4E+05	NK and/or T	MP	NR	Refractoriness	LDEP, PR	N	Yes	Death due to HLH Relapse after HSCT

AOSD: adult-onset Still's disease; CTD: connective tissue disease; SLE: systemic lupus erythematosus; MP: methylprednisolone; DEX: dexamethason; DEP: doxorubicin-etoposide-methylprednisolone regimen; L: PEG- asparaginase; R: rituximab; CR: complete response; PR: partial response; NR: no response; HSCT: haematopoietic stem cell transplantation.