

**Table S1. Characterization of the 10 patients**

Age, years/ Sex	Extracutaneous manifestation at first admission										Immunochemistry										IVIG				Extracutaneous manifestation of the last episode			
	HMB	Fever	LPA	SPM	HPM	Cytopaenia	EBER	CD3	CD4	CD8	CD30	CD56	TIA-1	Perforin	Gran-zyme	TCR	Previous therapy	Dose g/kg	Period, years	Other long-term therapies	Follow-up	Fever	LPA	SPM	HPM	Cytopaenia		
1 23/F	Yes	Yes	Yes	No	No	Anaemia	+	+	+	-	-	+	-	+	+	GC	0.4 g x 5 days	2.5	Minocycline Thalidomide	30 months Partial remission	No	Yes	No	No	No			
2 4/M	No	Yes	No	No	No	-	+	+	N/A	-	-	+	-	-	N/A	None	0.4 g x 3 days	Once	N	1 month then lost of follow	-	-	-	-	-			
3 3/F	Yes	Yes	Yes	No	No	Anaemia	+	-	-	N/A	N/A	N/A	N/A	N/A	N/A	GC	0.4 g x 5 days	10.5	N	128 months remission	No	No	No	No	No			
4 6/F	No	Yes	Yes	Yes	Yes	Neutropaenia	+	-	-	-	-	+	+	+	N/A	IFN+GC	0.4 g x 3 days	Once	CTX+VCR+GC	6 months Died	-	-	-	-	-			
5 11/M	Yes	Yes	Yes	No	No	-	+	+	+	-	-	+	-	+	-	IFN	0.4 g x 4 days	0.8	N	10 months Progressing to lymphoma	-	-	-	-	-			
6 3/F	Yes	Yes	Yes	Yes	No	-	+	+	+	-	-	+	N/A	-	+	IFN+GC	0.4 g x 3 days	3.5	IFN	45 months alive Partial remission	Yes	Yes	No	No	-			
7 10/M	Yes	Yes	Yes	Yes	No	-	+	+	N/A	-	-	-	-	+	+	None	0.4 g x 3 days	2	N	26 months alive Partial remission	Yes	Yes	Yes	No	-			
8 4/M	Yes	Yes	Yes	Yes	Yes	-	+	+	+	-	-	+	-	-	N/A	SMILE 2 cycles	0.4 g x 5 days	4.5	N	56 months alive Partial remission	Yes	No	Yes	No	-			
9 3/M	Yes	Yes	Yes	Yes	No	-	+	N/A	+	N/A	-	+	N/A	N/A	+	None	0.4 g x 5 days	1.5	N	18 months alive Partial remission	No	No	No	No	-			
# 18/M	Yes	Yes	Yes	Yes	No	Neutropaenia	+	+	+	-	-	+	N/A	N/A	+	None	0.4 g x 3 days	2	Thalidomide	25 months alive Partial remission	Yes	Yes	Yes	No	No			

LPA: lymphadenopathy; SPM: splenomegaly; HPM: hepatomegaly; M: male; F: female; GC: glucocorticoids; IFN: Interferon; SMILE: steroid (dexamethasone), methotrexate, ifosfamide, L-asparaginase, and etoposide; CTX: cyclophosphamide; VCR: vincristine.