

Table S1. Literature review of HHV8-negative or type II PEL cases.

Authors (year)	Case No.	Associated Medical Conditions	HIV Positive	HBV Positive	HCV Positive	Fluid Overload	EBV Positive	MYC Rearrangement	Phenotype/ Genotype	Median Survival (months)	1-year Survival Rate
Nador et al. (1996) [1]	4	NA	100% (4/4)	NA	NA	NA	100% (4/4)	100% (4/4)	Indeterminate	6	0% (0/3)
Hermine et al. (1996) [2]	1	NA	0% (0/1)	NA	NA	NA	0% (0/1)	NA	B-cell	NA	NA
Carbone et al. (1996) [3]	2	NA	50% (1/2)	NA	NA	NA	50% (1/2)	NA	B-cell	3	0% (0/2)
Ascoli et al. (1997) [4]	1	LC	0% (0/1)	NA	100% (1/1)	100% (1/1)	NA	100% (1/1)	B-cell	2	0% (0/1)
Carbone et al. (1997) [5]	1	NA	0% (0/1)	NA	NA	NA	100% (1/1)	100% (1/1)	B-cell	0.3	0% (0/1)
Ichinohasama et al. (1998) [6]	1	LC	0% (0/1)	NA	100% (1/1)	100% (1/1)	0% (0/1)	0% (0/1)	B-cell	10	0% (0/1)
Tanaka et al. (2001) [7]	1	No	0% (0/1)	NA	0% (0/1)	NA	0% (0/1)	0% (0/1)	B-cell	3	0% (0/1)
Ashihara et al. (2001) [8]	1	Brain tumor	0% (0/1)	0% (0/1)	0% (0/1)	100% (1/1)	100% (1/1)	NA	B-cell	24	100% (1/1)
Hara et al. (2001) [9]	1	LC	0% (0/1)	0% (0/1)	100% (1/1)	100% (1/1)	NA	NA	B-cell	NA	NA
Ohori et al. (2001) [10]	1	LC; liver transplantation	NA	100% (1/1)	NA	0% (0/1)	100% (1/1)	NA	B-cell	NA	NA
Rodriguez et al. (2001) [11]	1	LC	0% (0/1)	NA	NA	100% (1/1)	100% (1/1)	NA	B-cell	NA	NA
Yamamoto et al. (2001) [12]	1	cerebral aneurysm	0% (0/1)	0% (0/1)	0% (0/1)	0% (0/1)	0% (0/1)	NA	T-cell	5	0% (0/1)
Ohshima et al. (2002) [13]	5	NA	0% (0/5)	NA	NA	NA	20% (1/5)	80% (4/5)	B-cell	NA	NA
Hisamoto et al. (2003) [14]	1	common variable immunodeficiency	0% (0/1)	NA	NA	100% (1/1)	100% (1/1)	NA	B-cell	0.6	0% (0/1)
Chiba et al. (2003) [15]	1	autoimmune hemolytic anemia	0% (0/1)	0% (0/1)	0% (0/1)	0% (0/1)	100% (1/1)	0% (0/1)	B-cell	3	0% (0/1)
Nakamura et al. (2003) [16]	1	left scrotum swelling	0% (0/1)	0% (0/1)	0% (0/1)	0% (0/1)	0% (0/1)	NA	B-cell	NA	NA
Paner et al. (2003) [17]	1	LC	0% (0/1)	NA	100% (1/1)	100% (1/1)	0% (0/1)	NA	B-cell	5	0% (0/1)
Shimazaki et al. (2003) [18]	1	AF	0% (0/1)	0% (0/1)	0% (0/1)	100% (1/1)	0% (0/1)	100% (1/1)	B-cell	5	0% (0/1)
Inoue et al. (2004) [19]	1	HF	0% (0/1)	0% (0/1)	0% (0/1)	100% (1/1)	0% (0/1)	0% (0/1)	B-cell	18	100% (1/1)
Nonami et al. (2004) [20]	1	chylothorax	0% (0/1)	0% (0/1)	100% (1/1)	100% (1/1)	0% (0/1)	100% (1/1)	B-cell	22	100% (1/1)
Takao et al. (2004) [21]	1	LC	0% (0/1)	0% (0/1)	100% (1/1)	100% (1/1)	0% (0/1)	100% (1/1)	B-cell	28	100% (1/1)
Fujiwara et al. (2005) [22]	1	No	0% (0/1)	NA	0% (0/1)	NA	0% (0/1)	0% (0/1)	B-cell	36	100% (1/1)
Jenkins et al. (2005) [23]	1	LC	0% (0/1)	NA	NA	100% (1/1)	NA	NA	Plasmablastic	NA	NA

Matsumoto et al. (2005) [24]	2	No	0% (0/2)	NA	NA	NA	0% (0/2)	NA	B-cell	NA	NA
Venizelos et al. (2005) [25]	1	renal transplantation	0% (0/1)	NA	NA	100% (1/1)	0% (0/1)	NA	T cell	1	0% (0/1)
Youngster et al. (2006) [26]	1	HF	0% (0/1)	0% (0/1)	0% (0/1)	100% (1/1)	0% (0/1)	NA	B-cell	NA	NA
Kobayashi et al. (2007) [27]	2	LC	0% (0/2)	NA	100% (1/1)	100% (1/1)	50% (1/2)	NA	B-cell	22	100% (2/2)
Niino et al. (2008) [28]	1	HF	0% (0/1)	0% (0/1)	0% (0/1)	100% (1/1)	100% (1/1)	NA	B-cell	30	100% (1/1)
Terasaki et al. (2008) [29]	1	No	0% (0/1)	NA	0% (0/1)	0% (0/1)	0% (0/1)	0% (0/1)	B-cell	22	100% (1/1)
Adiguzel et al. (2009) [30]	1	HTN ; CAD	0% (0/1)	NA	0% (0/1)	100% (1/1)	0% (0/1)	NA	Indeterminate	40	100% (1/1)
De Filippi et al. (2009) [31]	1	LC	0% (0/1)	NA	100% (1/1)	0% (0/1)	100% (1/1)	NA	Plasmablastic	9	0% (0/1)
Tsagarakis et al. (2009) [32]	1	AMI	0% (0/1)	100% (1/1)	0% (0/1)	100% (1/1)	100% (1/1)	0% (0/1)	B-cell	NA	NA
Takahashi et al. (2010) [33]	2	NA	0% (0/2)	NA	NA	NA	50% (1/2)	NA	B-cell	12	100% (1/1)
Cooper et al. (2010) [34]	1	HIV immunocompromised	100% (1/1)	0% (0/1)	100% (1/1)	NA	100% (1/1)	100% (1/1)	B-cell	2	0% (0/1)
Kagoya et al. (2011) [35]	1	HF	0% (0/1)	NA	0% (0/1)	100% (1/1)	0% (0/1)	NA	B-cell	3	0% (0/1)
Terasaki et al. (2011) [36]	2	HF	0% (0/2)	NA	0% (0/2)	100% (2/2)	0% (0/2)	50% (1/2)	B-cell	14	100% (2/2)
Wang et al. (2011) [37]	1	HTN	0% (0/1)	0% (0/1)	0% (0/1)	100% (1/1)	0% (0/1)	NA	B-cell	55	100% (1/1)
Sumida et al. (2012) [38]	1	polycystic kidney disease	0% (0/1)	NA	NA	100% (1/1)	NA	NA	B-cell	18	100% (1/1)
Kim et al. (2012) [39]	1	HTN	0% (0/1)	NA	0% (0/1)	0% (0/1)	0% (0/1)	NA	B-cell	18	100% (1/1)
Alexanian et al. (2013) [40]	5	CAD (2), LC (2), HF (1)	0% (0/2)	NA	33% (1/3)	60% (3/5)	20% (1/5)	NA	B-cell	2	0% (0/3)
Wu et al. (2013) [41]	1	No	0% (0/1)	0% (0/1)	0% (0/1)	0% (0/1)	0% (0/1)	0% (0/1)	B-cell	5.5	0% (0/1)
Xiao et al. (2013) [42]	3	LC (1), AF (1), pancreatitis (1)	0% (0/1)	0% (0/1)	0% (0/1)	33% (1/3)	0% (0/3)	100% (1/1)	B-cell 67% (2/3); Indeterminate 33% (1/3)	25.3	66.7% (2/3)
Mohammad et al. (2014) [43]	1	HTN, AF	0% (0/1)	NA	0% (0/1)	0% (0/1)	0% (0/1)	NA	B-cell	14	100% (1/1)
Fan et al. (2014) [44]	1	LC	0% (0/1)	100% (1/1)	0% (0/1)	0% (0/1)	0% (0/1)	NA	B-cell	0.3	0% (0/1)
Choi et al. (2015) [45]	1	HIV immunocompromised	100% (1/1)	0% (0/1)	0% (0/1)	0% (0/1)	100% (1/1)	NA	B-cell	NA	NA

Usmani et al. (2015) [46]	17	HF (7), ESRD (1), Breast ca (2), CAD (1), LBBB (1), HTN (1), COPD (1), PE (1), FL (1), renal disease (1)	0% (0/4)	NA	16.7% (1/6)	47.1% (8/17)	5.9% (1/17)	21.4% (3/14)	B-cell 88% (15/17); Indeterminate 12% (2/17)	47.3	45.5% (5/11)
Shin et al. (2017) [47]	1	Pitting edema	0% (0/1)	0% (0/1)	0% (0/1)	100% (1/1)	0% (0/1)	NA	B-cell	15	100% (1/1)
Chen et al. (2018) [48]	17	Prior cancers (3), CRF (2)	14% (1/7)	0% (0/7)	10% (1/10)	50% (7/14)	13% (2/16)	27% (3/11)	B-cell 77% (13/17); Plasmablastic 18% (3/17); Indeterminate 6% (1/17)	2.7	31% (5/16)
Kaji et al. (2020) [49]	64	Arrhythmia (27), renal dysfunction(11), myocardial infarction (10), hypothyroidism (8), LC (3)	0% (0/58)	0% (0/62)	1.6% (1/61)	20% (12/60)	13.3% (6/45)	19.4% (7/36)	B-cell 100% (64/64)	Median survival time after relapse: 13 months	2-year OS rate: 84.7%
Kim et al. (2020) [50]	1	HTN	0% (0/1)	0% (0/1)	0% (0/1)	0% (0/1)	0% (0/1)	0% (0/1)	B-cell	22	100% (1/1)
Fiori et al. (2021) [51]	2	CML (2)	NA	NA	NA	50% (1/2)	50% (1/2)	50% (1/2)	B-cell 100% (2/2)	16.5	100% (2/2)
Wang RC et al. (2021) [52]	2	No	0% (0/2)	NA	0% (0/2)	100% (2/2)	0% (0/2)	NA	B-cell 100% (2/2)	13	100% (2/2)
Subtotal	167	LC 13% (17/129), renal dysfunction 12% (15/129), HF 10% (13/129), prior cancer/leukemia 6% (8/129), post-transplantation 2% (2/129)	6% (8/130)	3% (3/90)	12% (14/116)	42% (58/139)	21% (30/143)	34% (31/90)	B-cell 92% (154/167); Plasmablastic 3% (5/167); T-cell 1% (2/167); indeterminate 4% (6/167)	–	47% (36/77)

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