Outcomes of Primary Effusion Lymphoma: Role of Autologous Hematopoietic Cell Transplantation

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Background

- Primary effusion lymphoma (PEL) is a rare non-Hodgkin lymphoma (NHL) commonly seen in the patients with immunodeficiency.
- PEL accounts for approximately 4% of all HIV associated NHL.
- PEL has a predilection for serous body cavities and can form in the pleural space, pericardium, and peritoneum; Extra-cavitary tumors are rare.
- PEL cases must show evidence of infection with human herpes virus 8 (HHV-8); often HIV+, EBV+
- Prognosis is poor and mainstay chemotherapy results in limited disease-free survival.
- Clinical outcome data of autologous stem cell transplant for PEL is limited due to rarity of the diagnosis.
- We describe our experience with PEL patients from two major academic centers, including 4 patients who underwent Autologous Stem Cell Transplant (ASCT).

Methods

- Study population: All patients diagnosed with PEL at the Moffitt Cancer Center and Mayo Clinic between January 1, 1994 and December 31st, 2017.
- PEL diagnosis verified by experienced hematopathologists (P.H.; M.H.) at each institute.
- OS was defined as time from ASCT (day 0) until death from any cause or last follow up date.
- PFS was defined as time from HCT until disease relapse/progression or death or last follow up date.
- Treatment response criteria per Modified Lugano 5-point scale (5PS).

Results

- Nine patients identified; One patient had IRIS after HAART therapy and passed away less than a month after diagnosis
- Four patients received ASCT.
- Median follow-up of alive patients of 25 months (range-8-29).

Table-1: Clinical Characteristics

Characteristic	Number of patients, N=9 (%)
Median patient age (range), years	48 (29-79)
Patient gender	M- 7 (77.8%) F- 2 (22.2%)
Patient Race	Black – 3 (33.3%) White – 6 (66.7%)
Site of Effusion	Pleural – 8 (88.9%) Pericardial – 4 (44.4%) Peritoneal – 2 (22.2%)
Bone Marrow Involvement	No – 8 (88.9%)
Elevated LDH	1 (11.1%)
Lymphadenopathy	7 (77.7%)
HIV	7 (77.7%)
Adjuvant ART	5 (55.6%)
EBV	4 (44.4%)
HHV8	8 (88.9%)
CD30	5 (55.6%)
CD38	3 (33.3%)
HD MTX	1 (11.1%)
ASCT	4 (44.4%)
Lines of therapies before ASCT (range), (n, %)	1- 3 (33.3%) 2- 1 (11.1%) N/A - 4 (44.4%)
Best response to First line	CR1 - 4 (44.4%) PD1 – 2 (22.2%) PR1– 2 (22.2%)
Best response	CR - 1 (11.1%) CR1 - 5 (55.6%) PD1 - 1 (11.1%)
Median Survival from diagnosis, months	23 (0 – 107)
Median PFS from diagnosis after First line	22 (0 – 107)

Table-2: Outcomes of 4 PEL patients s/p ASCT

	Age/Sex	1 st - line	Adjuvant	EBV	Disease status prior to ASCT	Lines of therapy prior to ASCT	HCT- CI score	Conditioning regimen	Day 100 disease status	Follow-up from ASCT (months)	Comments
1	62, M	hyperCV AD	HAART	+	CR1	1	2	BEAM	CR	23	
2	33, M	CHOP	HAART	unk	CR1	2	3	BEAM	PD	10	Relapsed 3 months after ASCT Expired 10 months after ASCT
3	39, M	DA-EPOCH	HAART	+	CR1	1	4	BEAM	CR	9	Relapsed 6 months after ASCT, Expired 9 months after ASCT
4	49, M	hyperCV AD	HAART	+	CR1	1	4	BEAM-R	CR	20	

Conclusions

- A majority of our patients (8, 88.9%) had pleural effusions, associated with poor prognosis in the literature.
- Autologous stem cell transplant along with high dose chemotherapy and HAART therapy also has been used to treat HHV-8 associated PEL
- Our findings revealed ASCT as viable therapy for PEL with a 2-year PFS and OS of 58% and 75%, respectively.
- Unfortunately, the 2-year relapse rate was 50% (19-100).
- Our series showed improved OS of PEL patients compared to previous reports.
- Consolidative ASCT is safe and may be considered for eligible PEL patients after combination chemotherapy.
- Novel treatments are needed to prevent disease relapse after ASCT.

Figure 1: OS of 9 patients

Figure 2: PFS of 8 patients