soho2023
ELEVENTH ANNUAL MEETING

INTENSIVE CHEMOTHERAPY AND AUTOLOGOUS STEM CELL CONSOLIDATION FOR BING NEEL SYNDROME

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INTRODUCTION

Bing-Neel syndrome (BNS) is a complication of lymphoplasmacytic lymphoma comprising infiltration in the central nervous system (CNS). There is no standard of care; both chemoimmunotherapy (CIT) and BTK inhibitors are effective. Intensive CNS-penetrating CIT is well described in aggressive lymphomas, but there are limited data in BNS and the role of BTKi is uncertain, particularly in the setting of progressive neurological compromise.

AIM

To review the outcomes of those with BNS receiving intensive CIT and autologous stem cell transplant consolidation.

METHOD

Data from patients with BNS referred between 2013-2021 to our Neurohaematology centre were retrospectively reviewed.

PATIENT CHARACTERISTICS AND OUTCOMES

	1	2	3	4	5	6	7
Age at BNS	50	59	62	71	59	44	66
Prior WM therapy	-	-	BR	-	-	RCVP	DRC, Ibrutinib, BR
CNS involvement	CSF PNS	CSF PNS Cranial nerve, brachial/lumbar plexi	CSF Parenchymal Frontal lobe, spinal cord	CSF Parenchymal hydrocephalus, cranial nerves	CSF Parenchymal Cerebellar	CSF PNS Sural nerve	CSF Parenchymal Frontal, parietal ?High grade
Age at ASCT	50	59	62	71	59	44	66
Induction	R-MTX-Arac	R-MTX-Arac	R-MTX-Arac	R-MTX-Arac >2yr remission. Relapse Ibr (PR), RIE	R-MTX-Arac	IDARAM, ESHAP	MATRix
Consolidation	BCNU-thiotepa	BCNU-thiotepa	BCNU-thiotepa	BCNU-thiotepa	BCNU-thiotepa	Cy-TBI	BCNU-thiotepa
Response	Clinical response	Clinical response	Clinical response, CSF relapse (Zanu)	CNS relapse (BR)	Clinical response	Systemic relapse	High grade CNS relapse
Death	-	-	-	Progression	-	-	Progression

BCNU,carmustine; MATRix, MTX-Arac-thiotepa-rituximab; BR, Bendamustine-Rituximab; DRC, Dexamethasone-rituximab-cyclophosphamide; IDARAM, Idarubicin-dexamethasone-cytarabine-methotrexate; ESHAP, Etoposide-methylprednisolone-cytarabine-cisplatin

RESULTS

- Of 43 symptomatic patients, median age at BNS diagnosis was 66 years (range 43-85). All had leptomeningeal disease.
- Seven patients with progressive clinical features had ASCT consolidation (median age 59 years [range 43-68]).
- All patients had 2-4 cycles of intensive CIT (six: R-methotrexate-cytarabine ± thiotepa; one: IDARAM/ ESHAP) due to clinical severity
- In six patients, consolidation ASCT was delivered in first response (median of 11 months; range: 4-22) from BNS diagnosis. 1/7 achieved CR after intensive CIT, however, relapsed after 2 years and commenced ibrutinib with CNS progression, and then proceeded with ifosfamide-based therapy and ASCT.
- 6/7 had BCNU-thiotepa and 1 cyclophosphamide-total body irradiation conditioning. Engraftment occurred at a median of 12 days (range: 9-13) for neutrophils,11 days (range: 10-14), for platelets); hospitalization, 21 days (range: 19-41); critical care admission and 100-day nonrelapse mortality, 0%.
- Median follow-up was 11 months (range: 1-96) from ASCT; 2/7 died with relapsed lymphoma >18 months from ASCT (1/7 high grade transformation); 5/7 patients were alive; 1/7 experienced systemic relapse (post cyclophosphamide-TBI-ASCT) and commenced ibrutinib; 1/7 experienced CNS progression and commenced zanubrutinib.

CONCLUSION

In our selective cohort of relatively young BNS patients with adverse clinical features (extensive CNS burden; high-grade suspicion; BNS post-BTKi/CIT failure) who proceeded with intensive chemotherapy and ASCT consolidation, treatment-related morbidity and nonrelapse mortality were low and clinical response was notable.

CONTACT INFORMATION

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