



Third time lucky: A case report of relapsing Blastic plasmacytoid dendritic cell Neoplasm



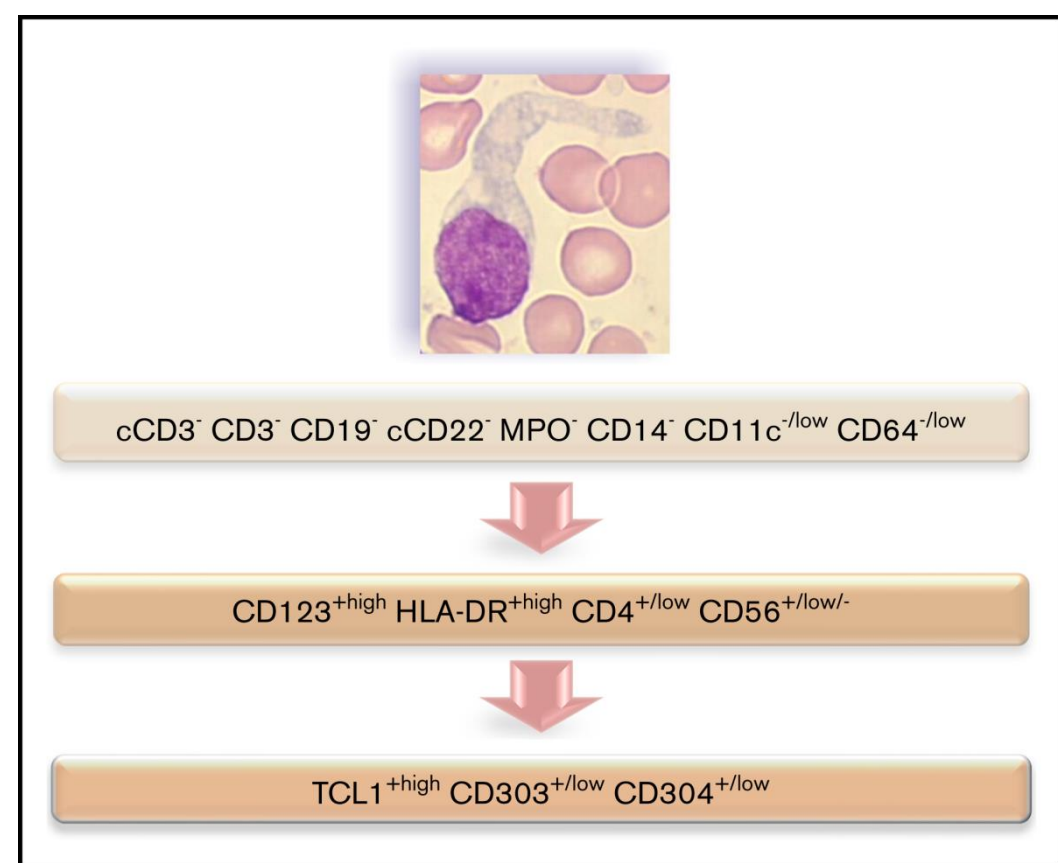
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Abstract

Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a rare and aggressive leukemia for which there is currently no consensus regarding the best therapeutic approach. The disease is now clearly categorized in the new World Health Organization (WHO) classification of hematological malignancies and distinguished from acute myeloid leukemia (AML). BPDCN is debatable in its diagnosis with no standard consensus on management making it a challenge to any physician.

Background

Most patients respond to various chemotherapy regimens, but relapses are almost inevitable, with a median overall survival (OS) of ~1 year. Better results are reported in the literature with leukemia-type regimens based on AML or acute lymphoblastic leukemia (ALL) chemotherapies. Nevertheless, only patients who undergo hematopoietic stem cell transplantation (HCT) seem to have a prolonged survival, especially after allogeneic HCT. Also the introduction of Tagraxofusp has



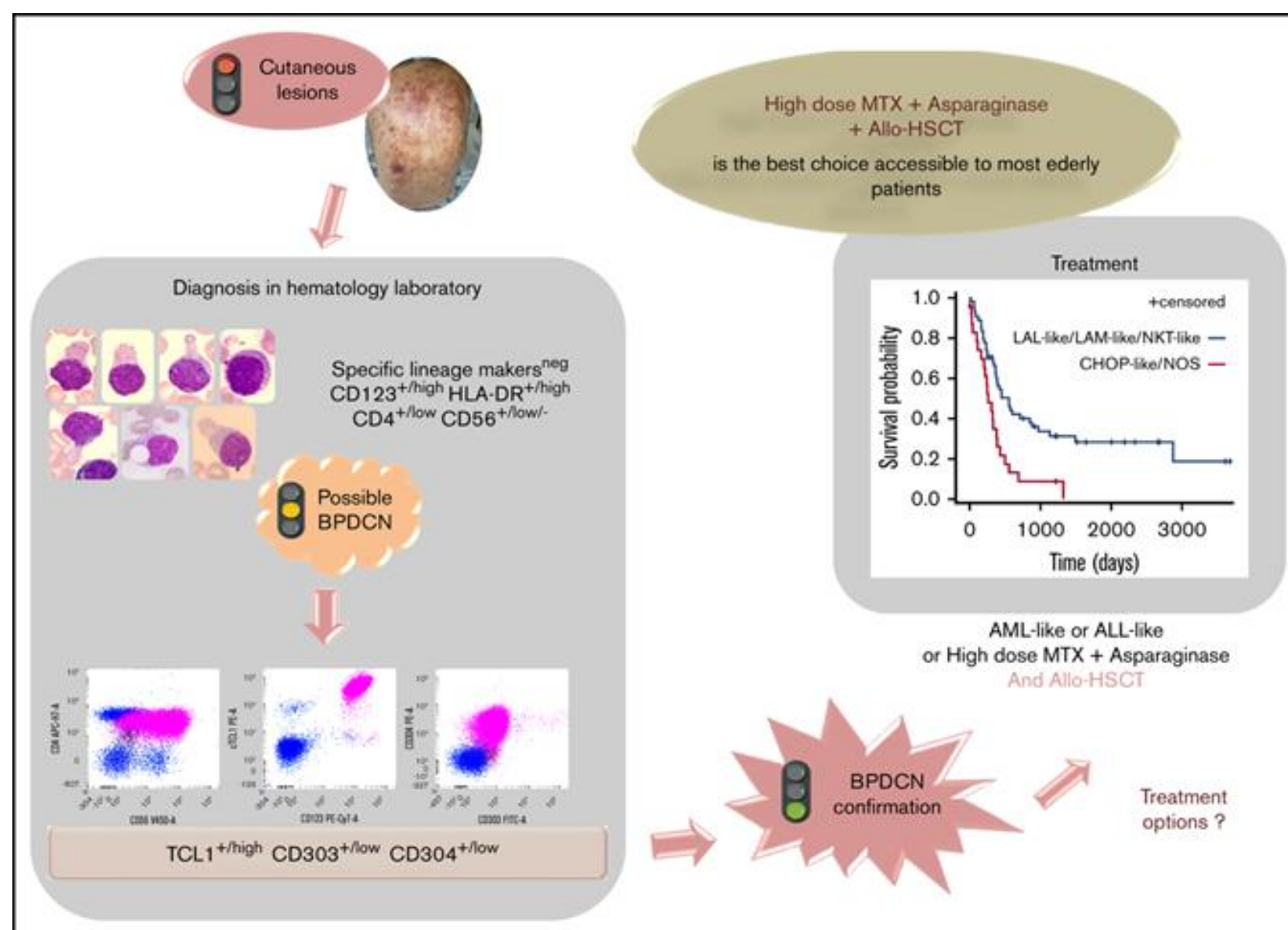
added great momentum and more treatment options for BPDCN patients either as first line or salvage treatment

Diagnosis of BPDCN1

Objective

We aim to illustrate the management of such a rare disorder like BPDCN in developing characters with coping with the changes in care especially in relapsed setting.

We focus on the management of BPDCN in developing countries where there is limited resources and scarce treatment options mainly related to conventional chemotherapy and radiotherapy with few patients having the possibility of undergoing autologous stem cell transplant.



Management of BPDCN1

Materials & Methods

DESIGN: This is a descriptive case report of a BPDCN from November 2017 till May 2021.

SETTING: Our facility is an academic referral center.

PATIENTS OR OTHER PARTICIPANTS: Our patient is a married 37 year old fisherman living in a fishing village in a nearby costal district, moderate cigarette smoker with no previous medical history presented to us in September 2017 with history of having a scalp lesion for three months with toxic symptoms in the form of night sweats and fever, He did a biopsy from the scalp lesion showing rounded cell tumor +ve LCA , -ve CK and HMB 45 and CBC showing mild leucopenia with BMA showing hypercellular BM 58% blasts suggestive of AML M5 for IPT and BMB hypercellular BM suggestive of acute leukemia. On examination he had bilateral bulky Cervical LNs about 3*3 cm and bilateral parotid swellings with spleen being not palpable. We repeated BMA being hypercellular 0 blasts and was confirmed by BMB and revision of the scalp lesion biopsy IHC: positive for LCA, CD4, CD56, S100. A case of BPDCN.

Summary

INTERVENTIONS: Patient started HyperCVAD protocol January 2018 with good response and disappearance of the scalp lesions. He was referred for HLA matching but his siblings refused so he continued up to eight HyperCVAD courses and was prepared for the ninth on October 2018 where he lost follow up and came five months later with a recurrent swelling in the left forearm showing recurrence after medullary clearance, He started HAM protocol June 2019 and received 2nd followed by four cycles of HDA protocol last January 2020 after which PET/CT showing negative study. On April 2020 he had a recurrent forearm lesion which showing recurrence, His PET CT was negative and received steroid with local irradiation.

Discussion

MAIN OUTCOMES MEASURES: OS of 44 months.

RESULTS: DFS of 13, 9 and 9 months (31 months total).

CONCLUSIONS: Treating BPDCN depending on chemotherapy only has higher incidence of relapse.

References

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