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Blastic Plasmacytoid Dendritic Cell Neoplasm (BPDCN) :
Rare, aggressive hematologic malignancy, due to proliferation of plasmacytoid dendritic cell precursors CD4+, CD56+, CD123+, **preferentially affects the elderly (1,2)**
Whatever the initial cutaneous presentation, involvement of blood and bone marrow appears rapidly in most patients, associated with poor prognosis.
Acute leukemia-based multi-agent chemotherapy induction followed by allogeneic hematopoietic stem cell transplantation (3): the only treatment to achieve sustained remissions. Recently, CD123-directed cytotoxin tagraxofusp, obtained 90% overall response but at the cost of frequent and sometimes serious toxicity (4).
Most patients are not eligible for these therapies due to their advanced age or co-morbidities and palliative approaches are then proposed (1,2).
14 patients with BPDCN diagnosed in our department between 2003 – 2020 :
6 advanced-age patients (average age of 85; range: 78 to 93), ECOG = 1-3=> all treated with vinca-alkaloids.



Characteristics of the patients, treatment, outcome and survival											
Sex	Age	ECOG	Skin lesions	Skin distribution	Initial bone marrow infiltration	Treatment*	Number of cycles	Initial response	Duration of initial response (months)	Side effects (grade)	Overall survival (months)
F	82	1	Nodules, macules	disseminated	yes	vindesine dexamethasone	6	PR	4	haematological (1) neurological (2)	7
F	87	1	nodule	unique (shoulder)	no	vindesine dexamethasone	12	CR	69	0	82
M	83	1	nodule	unique (trunk)	no	vindesine dexamethasone	8	CR	9	0	11
F	78	3	nodule	unique (thigh)	yes	vincristine dexamethasone	5	PD	NA	0	5
M	89	1	Nodules, macules	disseminated	yes	vincristine then vindesine dexamethasone	5 then 7	CR	2	haematological (1) neurological (2)	15
M	93	2	Nodules, macules	disseminated	no	vincristine dexamethasone	21	PR	23	asthenia (1)	28

vindesine -dexamethasone weekly cycles for 1 month followed by monthly cycles
Average = 10 cycles (range: 5 to 21).

Partial (n=2) or complete response (n=3)
Median duration : 9 months (2 to 69).
Safety:
2 grade 1 haematological toxicity and 2 grade 2 peripheral neurological toxicity
No infectious complication related to chemotherapy.
Median overall survival : 13 months (5-82 months)
All finally died of the disease with a bone marrow infiltration.

These results in advanced age patients compare favorably to intensive chemotherapies, used in younger patients,
Median survival of our patients treated by vinca-alkaloids was greater than expected and published in elderly patients (13 vs 8 months) (1).
Sustained clinical response in the majority of patients, up to 6 years after discontinuation of treatment.

Representation of overall survival (Kaplan Meier) for all patients diagnosed with blastic plasmacytoid dendritic cell neoplasms in our centre (2003-2020)

Red line:

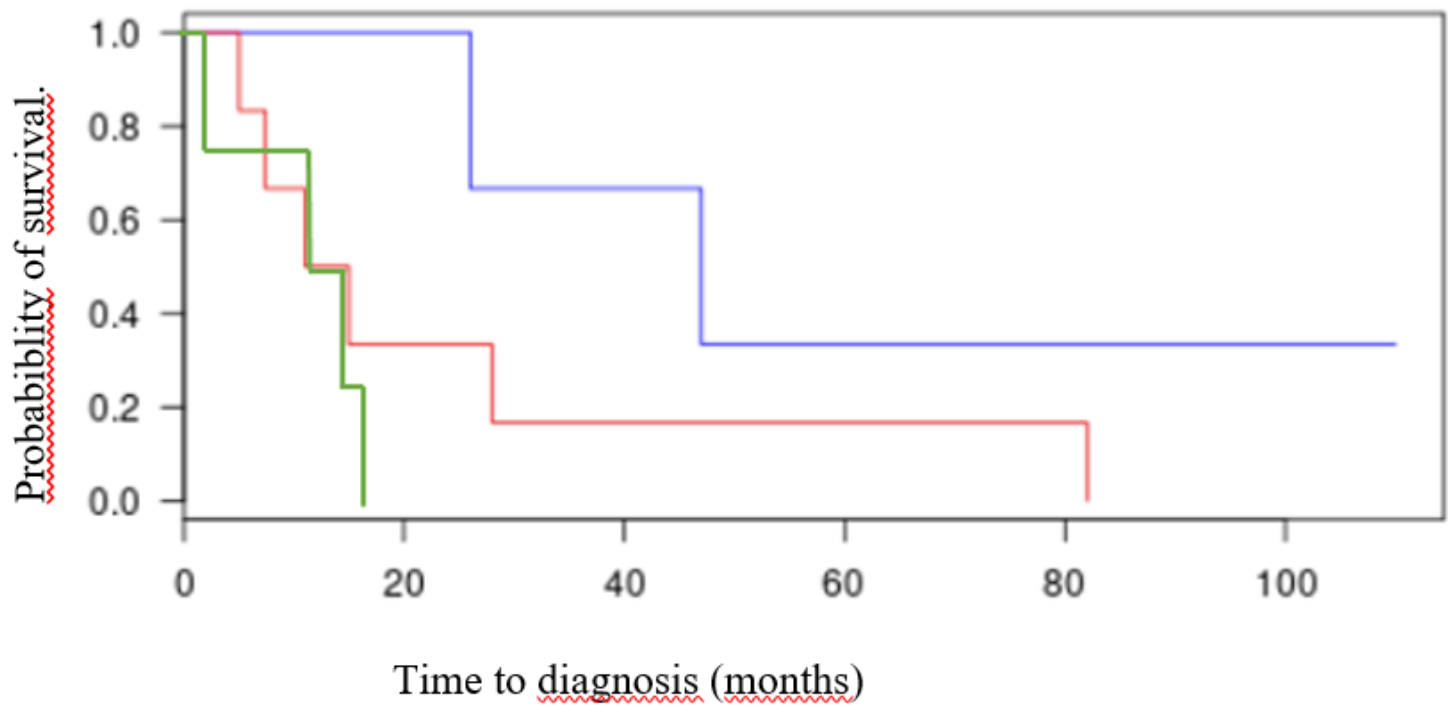
Patients treated with vindesine and dexamethasone (n= 6)
Overall survival: Median: 13 months; mean: 24.7 months (5 to 82)

Blue line:

Patients treated with allogeneic hematopoietic stem cell transplantation (n= 4)
Overall survival: Median 36.5 months; mean 49.25 months (14 to 110)

Green line:

Patients treated with other chemotherapy regimen (n = 4)
Overall survival: Median: 13.5 months; mean 11.6 months (2.5 to 17)



Vinca-alkaloids : an option for BPDCN even in very advanced age patients (prolonging life expectancy with a favorable toxicity profile)

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