Reviewer 1:
The Authors describe a unique case, the first so far in literature, of BPDCN patient treated with a clofarabine based regimen.

We thank the Reviewer for noticing the novelty of our report.

I understand that this is the first evidence of some degree of efficacy of such regimen in this setting. However, the efficacy was partial, with 6 months of morphological remission (MRD positive).

Again, we thank the Reviewer for her/his positive comments and we agree that, even if the remission was morphologically complete, the MRD remained positive. However, a six-month-long remission and a nine-month-long survival from first treatment is similar to the published results observed in leukemic-phase PBDCN patients treated with anthracyclin-based intensive regimens.

In addition, toxicity was quite severe. As I’m not really surprised that clofa/araC might be somehow effective in a myeloid tumor, I do not see at present a real interest in this case.

The Reviewer is right concerning toxicity but we interpreted the adverse events as being in part amplified by the underlying condition of our patient (recent history of polychemotherapy for lymphoma and diabetes). We also agree that efficacy of clofarabine was previously reported in myeloid malignancies and that its efficacy in PBDCN is not unexpected. However clofarabine was not previously reported in PBDCN patients, which justify to our opinion the interest of our case report.

The collection of a small series of 2-3 cases (being the disease very rare) might be a nice idea.

We certainly agree with the Reviewer on the interest of such a series. However, as mentioned, PBDCN is a very rare disease and when diagnosed, young patients are generally offered AML-like regimens and allotransplant while elderly are treated with less intensive regimens among which clofarabine is generally not considered as an option. Even if the results achieved in one single patient cannot be considered as a definitive proof of efficacy, the absence of valid strategy in this orphan disease prompted us to spread this information among physicians in charge of PBDCN patients.

Reviewer 2:
The authors describe a case of blastic plasmacytoid dendritic cell neoplasms treated palliatively with the combination of clofarabine and cytarabine.

It is an interesting, novel report, however, some (minor) points should be taken into account:

We thank the Reviewer for her/his very positive comments on our work.

The authors claim that this combination is a "suitable option" for patients ineligible for high-dose therapies. However, on the other hand, they state, that "multiple grade IV therapy-related adverse events" had occurred. This is a discrepancy and should be corrected especially an opportunistic infections may also be due to the underlying malignancy.

We agree with the Reviewer that “suitable option” is an overstatement of our findings. We modified “suitable option for patients ineligible for high-dose therapies” by “possible option for patients ineligible for anthracyclin-based regimens”.

Cardiac dysfunction should be explained more in detail (echocardiography etc).
We added details on the anthracyclin-related cardiac dysfunction: “She developed anthracycline-induced severe cardiac dysfunction six months after treatment as diagnosed by echocardiography showing global hypokinesia and decreased right ventricular ejection fraction (20% compared to 60% before chemotherapy).”

The same holds true for "hepatic dysfunction" as it is most likely related to the treatment

Concerning hepatic dysfunction, while chemotherapies given for lymphoma may have had a repercussion in liver function at some degree, all liver biological tests were normal before clofarabine initiation, as were liver morphological features (CT-scan). We therefore attributed liver dysfunction directly to clofarabine administration. To gain in clarity, we modified the text as follows: “and grade IV therapy-related adverse events including clofarabine-related acute hepatitis, palmoplantar erythrodysesthesia and mucositis”.

Reviewer 3:
The case report describes a 55-year-old woman with chemotherapy-related BPDCN, who showed morphological complete remission after clofarabine and cytarabine treatment. However, the patient relapsed and eventually died due to thrombocytopenia-induced intracranial bleeding.

The observation that the patient had a clinical response to clofarabine and cytarabine treatment, where intensive anthracyclin-based regimen and stem cell transplant could not be performed is valid information for clinicians in the field of Hematology.

We thank the Reviewer for these positive comments on our report.

However, there are a few points that need to be addressed by the authors:

1. The abstract should provide information that the PBDCN could be therapy-induced in this patient. Furthermore, the fact that the patient relapsed and eventually died is also valid information for this case report.

The abstract was modified accordingly.

2. In the introduction it is stated that 5q alterations predominate, but also 12p and 13q abnormalities occur frequently and should be mentioned (review of Riaz et al, 2014).

We modified the introduction as: “A range of cytogenetic alterations are reported in PBDCN with a predominance of deletions involving chromosomes 5q (72%), 12p and 13q (64%), 6q (50%), 15q (43%) and 9 (28%)”

3. Dexamethasone dosis is not complete 10 mg?.

We gave dexamethasone 10mg total dose which is now mentioned in the text.

4. The genetic lesions mentioned in the discussion should include IKZF1 and the genes should be written in Italic.

We modified this part as: “IKAROS family of genes (IKZF1, IKZF2, IKZF3), HOXB9, UBE2G2, ZEB2 and TET2 genes.”
5. Reference #14 is not updated correctly (Leukemia 2014).

We thank the Reviewer for noticing this error. Reference #14 is now provided adequately.