Blastic plasmacytoid dendritic cell neoplasm: a systematic review and meta-analysis

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Background and Objective

Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a rare, but highly aggressive hematologic malignancy derived from plasmacytoid dendritic cells. Currently, there is no consensus regarding the best therapeutic approach and the diagnosis of BPDCN remains a challenge. We aimed to collect and systematize the currently available evidence about the immunohistochemical (IHC) and flow cytometric (FCM) data in the diagnosis of BPDCN.

Materials & Methods

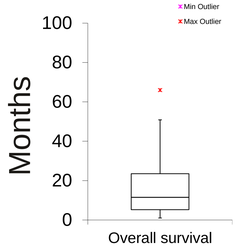
A systematic search was carried out in MEDLINE (via PubMed), Embase, Cochrane Central Register of Controlled Trials, Scopus, Web of Science, China National Knowledge Infrastructure, and Latin American and Caribbean Health Sciences Literature databases for studies published up to 20 April 2021. We selected clinical studies reporting on CD4+/CD56+/CD123+ co-expression in BPDCN patients among other markers. Cell surface markers' expression assessed by either IHC or FCM was collected regarding 106 proteins. The study protocol is currently under evaluation by the PROSPERO International prospective register of systematic reviews' editorial team.

Results

The systematic search and the data collection are ongoing. Out of the 3106 records collected, we identified **171 reported cases** from 83 full-text articles. These individual cases origin from 32 countries (Figure 1).

Figure 1: Geographical distribution of BPDCN cases in this study

The **median age at diagnosis was 62 years** (range: 0—96) and **70.8% of the cases (121/171) affected men**. The median overall survival was provided in 54 cases, which is shown in Figure 2, 33 patients were still followed, and there was no data on OS regarding 44 patients.



Overall survival (month)	
Median	11.5
Minimum	1.0
Maximum	66.0
Q1	5.25
Q3	23.5

Figure 2: Overall survival of the included patients

Figure 3: Ratio of confirmed presence in skin, bone marrow, and lymph nodes

Besides the three mandatory markers, TCL1, CD43, and CD45 were expressed in 92%, 93%, and 95%, respectively. Among the lineage-specific antigens, CD19 (absent in 97%), CD20 (absent in 92%), MPO (absent in 97%), and CD79a (absent in 97%) were almost always absent in the neoplastic cells.

Conclusion

Based on our preliminary findings, a confident immunophenotypic diagnosis can be made when CD4, CD56, CD123, TCL1, CD43, CD45 are present, and other lineage-specific antigens are absent (CD19, CD20, CD3, MPO, and CD79a). This work is continuing in collecting all reported data about the diagnostic algorithm of this disorder which could be used as a template for targeted therapy in BPDCN.

Future perspectives

After completing the selection and data collection process, we intend to provide the clinicians with an updated diagnostic algorithm. To validate our findings, we will re-assess the specific diagnostic markers in biopsies of the unreported BPDCN cases in our institute. We also plan to collect the most characteristic diagnostic markers including the genetic findings of all reported BPDCN cases in the literature.

References

1. https://mapchart.net/world.html Accessed: 2021.08.19.

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