P974 THE CLINICAL CHARACTERISTICS AND PROGNOSIS OF PATIENTS WITH PRIMARY PLASMA CELL LEUKEMIA (PPCL) UNDER THE NEW IWGM DEFINITION CRITERIA

**Topic:** 14. Myeloma and other monoclonal gammopathies - Clinical

Wenqiang Yan1, Jingyu Xu1, Huishou Fan1, Jiahui Liu1, Lingna Li1, Lugui Qiu1, Gang An1

1 State Key Laboratory of Experimental Hematology, National Clinical Research Center for Blood Diseases, Institute of Hematology & Blood Diseases Hospital, Chinese Academy of Medical Sciences & Peking Union Medical College, Tianjin, China

**Background:** Primary plasma cell leukemia has been described as a distinct entity of plasma cell disorders with special features and poor prognosis. In 2021, IMWG proposed that revised the diagnostic criteria of PCL as CPCs ≥ 5% by peripheral blood (PB) smear. As the new diagnostic standard has been established recently, it is of great significance that illustrate the clinical characteristics and outcomes of the new pPCL cohort.

**Aims:** Given the limited data for the rare entity, we performed a large retrospective analysis about the clinical characteristics, survival outcomes, and risk factors for the new-defined pPCL patients treated in our hospital.

**Methods:** We conducted a retrospective analysis of 158 pPCL patients diagnosed from 2000 to 2019 in our hospital. This pPCL cohort was redefined from NDMM patients who appear CPCs ≥ 5% by morphologic evaluation of their PB smear. And we compared them to a control group with 485 NDMM patients. Statistical analyses were performed using the R version 4.1.2.

**Results:** The characteristics and comparisons between pPCL patients and NDMM group are depicted in Table 1. In general, bone marrow suppression and adverse prognostic biomarkers (ie, anemia, thrombocytopenia, elevated LDH, hypodiploidy, and high-risk cytogenetics) were more common in pPCL patients compared with NDMM patients (P<0.05).

For 130 pPCL patients whose treatment and prognosis data were available, with a median follow-up time of 54.7 months, the median PFS and OS were 16.9 months and 30.0 months respectively, both significantly longer than the control NDMM patients (P<0.001). Interestingly, we find the cohort who attended after 2007 got obvious longer PFS than those who received treatment in 2000-2006 (20.6 vs 10.0 months, P=0.017), which showed a similar result in OS (31.6 vs 16.0 months, P=0.034).

A Cox-regression multivariate analysis was performed among the baseline variables. The presence of hypodiploidy and elevated serum LDH were found to be prognostic for worse PFS, whereas age<60 and elevated LDH were the independent predictors for worse OS. As for the cytogenetic aberrations, did not play an important role in the outcome of pPCL patients other than the presence of del(17p) which could impact OS in the univariate analysis.

There are 98 patients who had response data known, with 80(81.6%) patients achieving objective response in the first-line treatment, 56.4%≥VGPR, and 38.9% CR. When stratifying the cohort by the best response, the median PFS for patients achieved: NR, PR, and ≥VGPR were 2.4, 11.2, and 31.0 months, respectively (P<0.001). The OS for patients who had ever achieved NR, PR, and ≥VGPR were 2.4, 24.9, and 62.1 months, respectively (P<0.001). Then, we divided the cohort who had achieved≥PR into early response group (≤2 courses), intermediate response group (3-4 courses), and late response group (≥4 courses). Whereas, there is no statistical difference in the prognosis of 3 groups with diverse remission speed rates (PFS: P=0.35; OS: P=0.77). We also found that achieving deep remission was the best independent favorable predictor for long survival in the multivariate analysis.
Summary/Conclusion: In conclusion, primary plasma cell leukemia (pPCL) defined by the new revised diagnostic criteria (CPCs≥5%), remains an aggressive disease characterized with poor prognosis despite the advancement of treatment regimens. And achieving deep remission (≥VGPR) in the first-line therapy predicts the best prognosis regardless of response rate.

Table 1

| Clinical characteristics (%) | pPCL (n=158) | NDMM (n=485) | P        |
|------------------------------|-------------|--------------|----------|
| Age (median; years)          | 57(22–80)   | 58(29–83)    | 0.230    |
| Sex                          | M:89, F:69  | M:287, F:198 | 0.577    |
| M-component (n=158)          |             |              | NA       |
| IgG                          | 86/156(55.1)| 229/485      |          |
| IgA                          | 30/156(19.2)| 118          |          |
| IgD                          | 4/156(2.6)  | 31           |          |
| Light chain                  | 32/156(20.5)| 95           |          |
| Non-secretory                | 4/156(2.6)  | 8            |          |
| others                       | 0           | 4            |          |
| ISS staging                  |             |              | <0.001   |
| I                            | 10/143(7.0) | 91/485(18.8) |          |
| II                           | 24/143(16.8)| 176/485(36.3)|         |
| III                          | 109/143(76.2)| 218/485(44.9)|         |
| RISS staging                 |             |              | <0.001   |
| I                            | 2/113(1.8)  | 80/485(16.5) |          |
| II                           | 56/113(49.5)| 312/485(64.3)|         |
| III                          | 55/113(48.7)| 93/485(19.2) |          |
| Anemia                       | 134/157(85.4)| 225/485(46.4)| <0.001   |
| Thrombocytopenia             | 91/154(59.1)| 55/482(11.4) | <0.001   |
| Renal dysfunction            | 31/151(20.5)| 56/485(11.5) | 0.007    |
| Elevated LDH                | 48/137(35.0)| 81/485(16.7) | <0.001   |
| Bone disease                 | 92/109(84.4)| 416/485(85.8)| 0.763    |
| Hypodiploidy                 | 22/110(20.0)| 32/467(6.9)  | <0.001   |
| Cytogenetic abnormality      |             |              |          |
| del(17p)                     | 20/118(16.9)| 43/481(8.9)  | 0.013    |
| t (4;14)                     | 15/88(17.0) | 73/470(15.5) | 0.750    |
| t (14;16)                    | 8/87(9.2)   | 11/468(2.4)  | 0.005    |
| t (14;20)                    | 0/77(0)     | 3/441(0.7)   | NA       |
| t (11;14)                    | 25/84(29.8) | 78/462(16.9) | 0.007    |
| 1q21+                        | 55/112(49.1)| 223/476(46.8)| 0.675    |
| 1q21 gain                    | 45/112(40.2)| 143/476(30.0)| 0.043    |
| 1q21 amplification           | 10/112(8.9) | 80/476(16.8) | 0.041    |
| High-risk cytogenetics       | 40/93(43.0) | 119/476(25.0)| 0.001    |

Data are presented as n (%) and median (range) unless otherwise indicated. High-risk cytogenetics: 17p−, t(4;14), t(14;16). Abbreviations: LDH = lactate dehydrogenase.

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