hemato

MDPI

Article

Global Research Trends in Primary Plasma Cell Leukemia: A Bibliometric Analysis †

Öznur Aydın [1,*] and Serkan Güven [2]

[1] Department of Hematology, Faculty of Medicine, Samsun University, Samsun 55080, Türkiye
[2] Department of Hematology, Çanakkale Mehmet Akif Ersoy State Hospital, Çanakkale 17100, Türkiye
* Correspondence: oznur.aydin@samsun.edu.tr
† A preliminary version of this study was presented as a poster (Poster No. LBA-23) at the 23rd International Myeloma Society Annual Meeting on 23 September 2026. The conference abstract was published in Clinical Lymphoma Myeloma and Leukemia, 2026, 26 (Supplement 2), S385 (https://doi.org/10.1016/S2152-2650(26)03949-2).

Highlights

What are the main findings?

- This study provides a comprehensive bibliometric analysis of 210 validated articles on primary plasma cell leukemia published over more than four decades.
- Research output increased overall, with an annual growth rate of 5.95%; however, the leading countries differed according to publication, citation, and authorship indicators.

What are the implications of the main findings?

- The findings identify the leading journals, authors, institutions, countries, and collaboration networks contributing to primary plasma cell leukemia research.
- The relatively limited rate of international co-authorship highlights the need for broader multicenter and international collaboration in this rare hematologic malignancy.

Abstract

Background/Objectives: Primary plasma cell leukemia (pPCL) is a rare and highly aggressive plasma cell malignancy with poor prognosis and limited therapeutic options. This study aimed to characterize global publication and citation trends, leading contributors, and collaboration patterns in pPCL research. **Methods:** Scopus was searched on 8 September 2026 using a phrase-restricted TITLE-ABS-KEY query for the disease name and orthographic variants. English-language records classified as "Article" and published from 1961 through 2026 were eligible. All retrieved records underwent author-led relevance screening and duplicate assessment. Analyses were performed using bibliometrix through the Biblioshiny interface. **Results:** Of 219 records retrieved, nine were excluded because pPCL was mentioned only incidentally or pPCL-specific findings were not separately identifiable; 210 articles published between 1983 and 2026 were included. The articles appeared in 110 sources and involved 1681 authors. The annual publication growth was 5.95%, although 2026 was incomplete, and the mean citation count per article was 30.02. Acta Haematologica was the most productive journal (n = 10), followed by Blood (n = 9) and the Indian Journal of Hematology and Blood Transfusion (n = 8). Among 152 articles with corresponding-author country data, China, the United States, and India contributed the most articles, whereas France, the United States, and Spain had the highest total citation counts. International co-authorship accounted for 10.95% of articles, and the author network contained five collaboration clusters. **Conclusions:** This analysis maps the development of pPCL research and identifies its principal publication sources and contributors.

Check for updates

Academic Editor: Francesco Onida

Received: 14 September 2026
Revised: 29 September 2026
Accepted: 29 September 2026
Published: 1 October 2026

Copyright: © 2026 by the authors. Licensee MDPI, Basel, Switzerland. This article is an open access article distributed under the terms and conditions of the Creative Commons Attribution (CC BY) license.

The limited proportion of internationally co-authored articles supports broader multicenter and international collaboration in this rare disease.

Keywords: primary plasma cell leukemia; bibliometric analysis; research trends; scientific collaboration

1. Introduction

Plasma cell leukemia (PCL) is a rare and highly aggressive plasma cell malignancy characterized by the presence of circulating clonal plasma cells in peripheral blood. It comprises two clinically distinct entities. Primary plasma cell leukemia (pPCL) presents de novo in patients without a previous history of multiple myeloma, whereas secondary plasma cell leukemia (sPCL) represents the leukemic transformation of previously diagnosed multiple myeloma. The present study focuses exclusively on pPCL. Historically, pPCL was defined by the presence of $\geq$20% circulating plasma cells or an absolute circulating plasma cell count exceeding 2×10^9/L. In 2021, the International Myeloma Working Group revised the consensus definition and recommended a lower diagnostic threshold of $\geq$5% circulating plasma cells on peripheral blood smear examination in patients with otherwise symptomatic multiple myeloma [1–3]. This revision facilitates the earlier recognition of patients with highly aggressive disease.

Primary PCL accounts for approximately 1–2% of newly diagnosed multiple myeloma cases [4]. Patients commonly present with a high tumor burden, cytopenias, renal dysfunction, hypercalcemia, and frequent extramedullary involvement. At the molecular level, pPCL is characterized by a high prevalence of high-risk cytogenetic abnormalities, including chromosome 1q gain, translocations involving the immunoglobulin heavy-chain locus, and deletion of chromosome 17p [5]. These genomic features are associated with the aggressive biological behavior of the disease and resistance to conventional therapies. Despite advances in the treatment of plasma cell disorders, including proteasome inhibitors, immunomodulatory drugs, monoclonal antibodies, and autologous stem cell transplantation, the prognosis of pPCL remains poor. Median overall survival historically ranged from less than one year to approximately two years, although improvements have been observed with the introduction of novel therapeutic strategies [6].

Because of the rarity of pPCL, the available literature consists largely of retrospective studies, small cohort analyses, and institutional case series [7,8]. Consequently, the development and overall structure of this research field have not been fully characterized from a bibliometric perspective. To address this gap, the present study aimed to evaluate publication trends, citation impact, leading sources, authors and institutions, country contributions, and international collaboration patterns in global pPCL research using bibliometric methods.

2. Materials and Methods

2.1. Research Design

This study employed a bibliometric approach to evaluate the global scientific literature on primary plasma cell leukemia (pPCL). The analysis was designed to characterize publication trends, leading contributors, institutional productivity, citation impact, and collaboration patterns within the field using established bibliometric methods [9,10].

2.2. Data Source and Search Strategy

Bibliographic records were retrieved from the Scopus database (Elsevier) on 8 September 2026. The following advanced search query was executed in the title, abstract, and indexed keywords (TITLE-ABS-KEY) field:

```
TITLE-ABS-KEY(
  "primary plasma cell leukemia"
  OR "primary plasma cell leukaemia"
  OR "primary plasma-cell leukemia"
  OR "primary plasma-cell leukaemia"
)
AND PUBYEAR > 1960
AND PUBYEAR < 2027
AND (LIMIT-TO(DOCTYPE, "ar"))
AND (LIMIT-TO(LANGUAGE, "English"))
```

This query restricts retrieval to records containing the exact phrase "primary plasma cell leukemia" (or a recognized orthographic variant) in the title, abstract, or author/indexed keywords. The document-type filter DOCTYPE="ar" restricts the search to records classified by Scopus as Articles; the language was restricted to English, and the publication-year window was 1961–2026. The search returned 219 records. Relevance screening and duplicate assessment were performed as described below. This exact-phrase strategy was selected to maximize the specificity of the retrieved corpus. However, potentially relevant records using broader or alternative terminology, such as "plasma cell leukemia" or "de novo plasma cell leukemia", without one of the predefined exact phrases in the title, abstract, or indexed keywords may not have been retrieved. Accordingly, the analyzed corpus represents the literature identified through the predefined search strategy and should not be regarded as an exhaustive collection of all publications potentially relevant to pPCL.

2.3. Inclusion Criteria

Document type was restricted at the database level to records indexed by Scopus as "Article" (DOCTYPE="ar"). This category includes original research articles as well as case reports and small case series; conference papers, reviews, editorials, and letters were excluded by the search filter. Records were therefore described throughout the manuscript as "included articles" rather than "original articles". Eligible records were English-language articles published within the predefined 1961–2026 search window and substantively related to pPCL according to the relevance-screening procedure below.

2.4. Relevance Screening and Duplicate Assessment

Because pPCL is rare and a phrase-restricted search may still retrieve records in which pPCL is only incidental, all 219 retrieved records underwent record-by-record title-and-abstract relevance screening using a pre-specified rule.

Relevance screening: A record was retained when pPCL was a substantive focus or when it reported pPCL-specific results, outcomes, or a separately identifiable pPCL subgroup analysis. Records were excluded when pPCL was mentioned only incidentally within a broader cohort or when pPCL-specific results were not separately analyzable. Nine records were excluded under this rule, yielding 210 articles for bibliometric analysis.

Duplicate assessment: Among the final 210 articles, 186 records contained a DOI and no duplicated DOI was identified. The remaining 24 records lacked a DOI and were additionally checked using bibliographic fields including normalized title, publication year, source, and first-author information. Repeated generic titles were retained only when the

associated year, author, and/or source information showed that they represented distinct publications. No duplicate record was identified in the final analytic dataset.

The screening and duplicate-assessment procedure therefore yielded a final analytic dataset of 210 included articles from 219 records retrieved by the revised Scopus query.

2.5. Data Cleaning, Author Identity Checks, and Institutional Affiliations

Bibliographic data were processed in R using bibliometrix/Biblioshiny, and the standard author-name strings generated by the package were retained for the reported productivity and collaboration outputs. Because abbreviated author labels may represent more than one individual, Scopus Author Identifiers available in the exported author-full-name field were used to verify the leading authors reported in Section 3 and to assess potentially ambiguous high-frequency author labels. The Biblioshiny author counts were not manually replaced by a fully reconstructed identifier-level dataset.

Institutional affiliations were extracted and grouped using bibliometrix applied to the raw Scopus affiliation field. Because affiliation strings are not fully standardized, generic labels may aggregate multiple institutions. Generic affiliation labels were retained in the automated output for transparency but were not interpreted as individual institutions. In particular, the entry "University Hospital" (n = 40) represents a generic Scopus affiliation label and should not be interpreted as a single institution. The leading specifically named institutions are therefore reported separately in Section 3.

Metadata completeness was checked for all fields used in the analyses. Core fields (title, abstract, authors, document type, source, language, publication year, and total citations) were complete across the 210 records. Author affiliation was missing for 0.95% of records and indexed keywords for 5.24%. DOI was unavailable for 24 records (11.4%), and corresponding-author information was unavailable for 58 records (27.6%). Consequently, the corresponding-author country analysis used the 152 records with identifiable corresponding-author country data.

2.6. Data Analysis

Bibliographic records from the final validated dataset were analyzed using RStudio (version 4.6.1) and the bibliometrix package (version 5.0) through the Biblioshiny interface [9]. The analyses included annual scientific production, citation indicators, source and author productivity, institutional and country contributions, and co-authorship/collaboration networks. Selected design-related indexed terms were summarized descriptively to characterize study design-related indexing without imposing a retrospective manual classification of individual articles. Because indexed terms may overlap within the same article, synonymous singular/plural variants were reported separately rather than summed as unique studies unless record-level overlap could be ruled out. Network visualizations were generated using the bibliometrix settings reported in the corresponding figure captions.

Because data collection was performed on 8 September 2026, the publication count for calendar year 2026 (n = 12) represents only the portion of the year indexed in Scopus at the time of the search and was treated as an incomplete annual observation rather than as directly comparable with complete calendar years.

3. Results

Following the search strategy and screening procedure described in Sections 2.2–2.4, the analytic dataset comprised 210 articles published between 1983 and 2026 (Table 1). The final dataset included 110 sources and 1681 authors. The annual publication growth rate was 5.95%, the mean number of citations per publication was 30.02, and the mean number

of citations per publication per year was 2.16. International co-authorship accounted for 10.95% of the dataset. Because data collection was performed on 8 September 2026, the 2026 publication count (n = 12; Figure 1) is incomplete and should not be compared directly with complete calendar years.

Table 1. Main Bibliometric Characteristics of the Study Dataset.

Description	Result
Publication period	1983–2026
Sources (journals)	110
Publications (articles)	210
Annual Growth Rate, %	5.95
Document average age, years	14.7
Average citations per publication	30.02
Average citations per publication per year	2.16
Keywords Plus	2008
Author's Keywords	329
Authors	1681
Authors of single-authored articles	3
Co-authors per publication	10.1
International co-authorship, %	10.95
Document type	Article: 210 (100%)

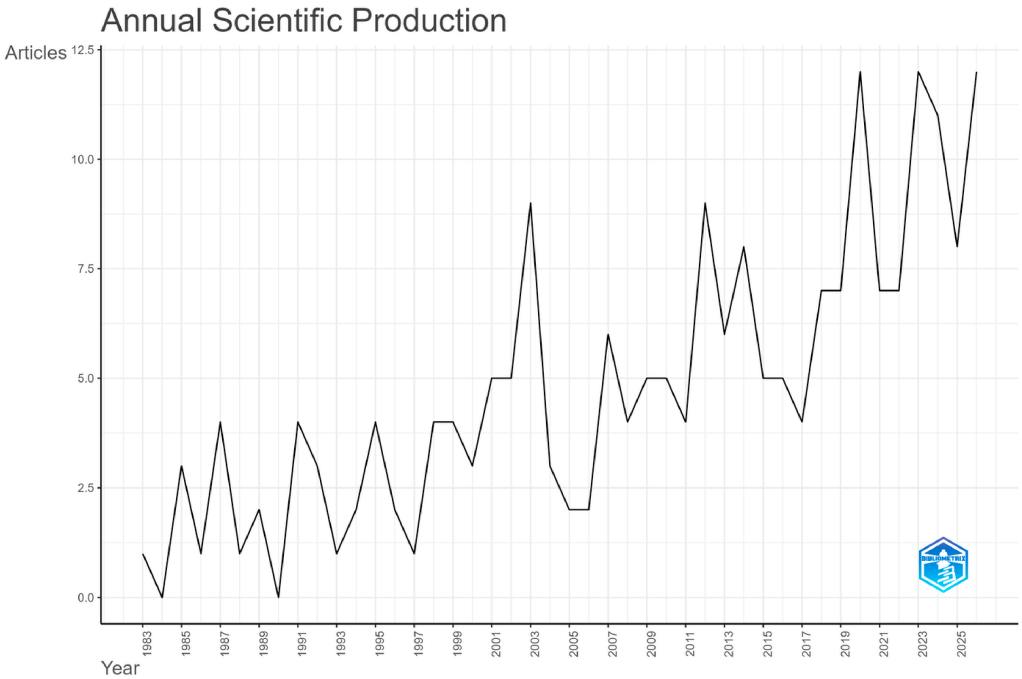


Figure 1. Annual Scientific Production of pPCL Research, 1983–2026. Number of articles published per year on primary plasma cell leukemia in the final validated dataset (n = 210). Because the Scopus search was executed on 8 September 2026, the final data point (2026, n = 12) reflects only the portion of the year captured by the database at the time of the search and does not represent a complete annual total; it should not be compared directly with earlier, complete years.

All 210 included records were classified by Scopus as document type "Article". To provide a descriptive view of study design-related indexing, selected indexed terms were summarized. The term "case report" had a frequency of 104 (49.5% relative to the 210-article corpus), "retrospective study" 43 (20.5%), "retrospective studies" 30 (14.3%), "cohort analysis" 23 (11.0%), and "clinical trial" 16 (7.6%). These are database-indexed terms

rather than independently adjudicated study-design categories; terms may co-occur within the same article, and the singular/plural retrospective-study variants were therefore not summed as a count of unique studies.

4. Discussion

Primary plasma cell leukemia (pPCL) is a rare and highly aggressive plasma cell dyscrasia associated with substantial biological and therapeutic challenges [1]. The present bibliometric analysis maps the global pPCL literature published between 1983 and 2026, paying particular attention to publication and citation patterns, leading contributors, and collaboration networks. By integrating these dimensions, the study provides a framework for understanding the development of the field and identifying areas in which research activity and collaboration remain limited.

The validated corpus indicates that scholarly activity in pPCL has expanded over time, although the overall volume of publications remains limited for the period examined (Figure 1). The final annual observation for 2026 is incomplete and should not be interpreted as evidence of a change in the overall trend. The prominence of the indexed terms "case report" and "retrospective study", relative to "clinical trial", suggests that the field remains largely dependent on descriptive and retrospective evidence. Because these terms may overlap and were not independently adjudicated, they reflect indexing patterns rather than a definitive distribution of study designs. This evidence profile is consistent with the challenges of conducting large prospective studies in a rare and aggressive disease.

The concentration of publications in specialist hematology journals demonstrates that pPCL research remains distributed through relatively focused publication channels (Figure 2). The most relevant authors and institutions are shown in the Figures 3 and 4. The countries leading publication output were not identical to those accumulating the highest citation totals (Figures 5 and 6), indicating that productivity and accumulated scientific influence do not necessarily coincide. Citation-based comparisons should nevertheless be interpreted cautiously because raw totals are influenced by publication volume and article age. Similarly, the institutional analysis identifies centers with sustained activity, but generic affiliation labels such as "University Hospital" limit the interpretation of automated institutional rankings (Figure 4).

International co-authorship involved only 10.95% of the analyzed articles, while the author network showed five relatively distinct collaboration clusters (Figure 7). Together, these findings indicate the presence of established research hubs but limited integration across groups and countries. Broader collaboration between these networks may facilitate the assembly of larger patient cohorts, validation of revised diagnostic criteria, and the development of more informative prospective therapeutic studies.

The increase in pPCL-related publications has occurred alongside the adoption of novel agent-based treatment strategies in plasma cell malignancies. Proteasome inhibitors, immunomodulatory drugs, and monoclonal antibodies have expanded therapeutic options, although pPCL-specific evidence remains largely derived from retrospective studies and small cohorts [3,7]. The growth in publication output therefore indicates increasing clinical interest but not necessarily a corresponding increase in high-level prospective evidence.

Early clinical descriptions established the aggressive presentation of pPCL [11]. Subsequent studies identified frequent high-risk cytogenetic lesions, including deletion 17p, chromosome 1q abnormalities, and complex karyotypes [12–14]. These features may partly explain the poor outcomes observed in pPCL compared with multiple myeloma.

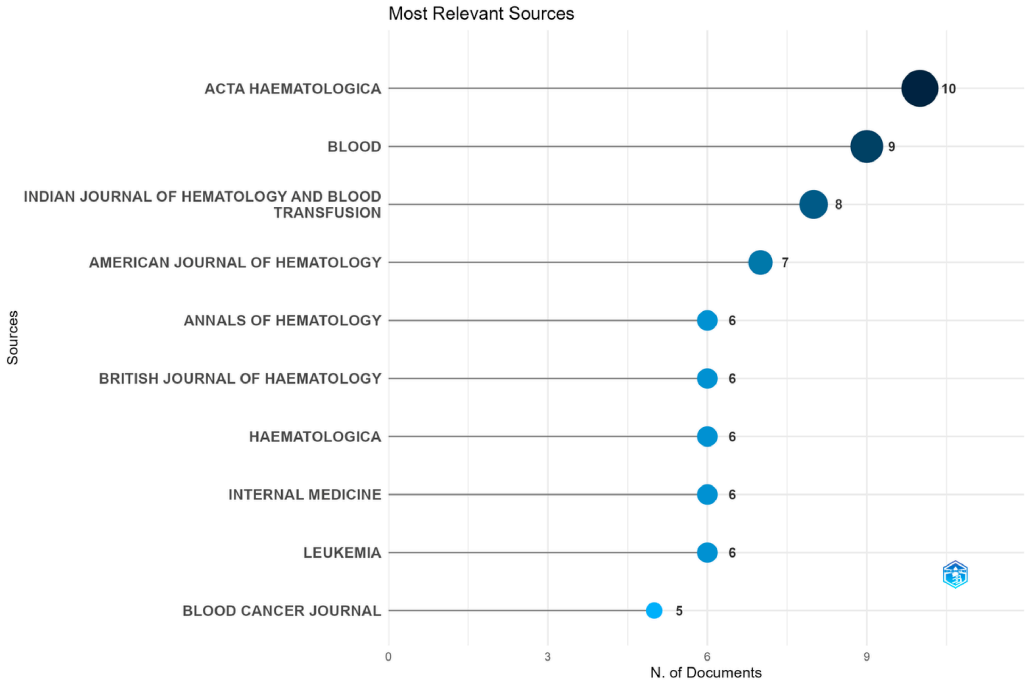


Figure 2. Most Relevant Sources (Journals). The 10 journals with the highest number of published articles on pPCL in the final dataset (n = 210), ranked by absolute article count per journal. Acta Haematologica (n = 10), Blood (n = 9), and the Indian Journal of Hematology and Blood Transfusion (n = 8) were the three most productive sources. The lollipop plot displays article count on the horizontal axis; marker size and shade provide visual emphasis and do not represent additional statistical variables. Article count is distinct from citation impact.

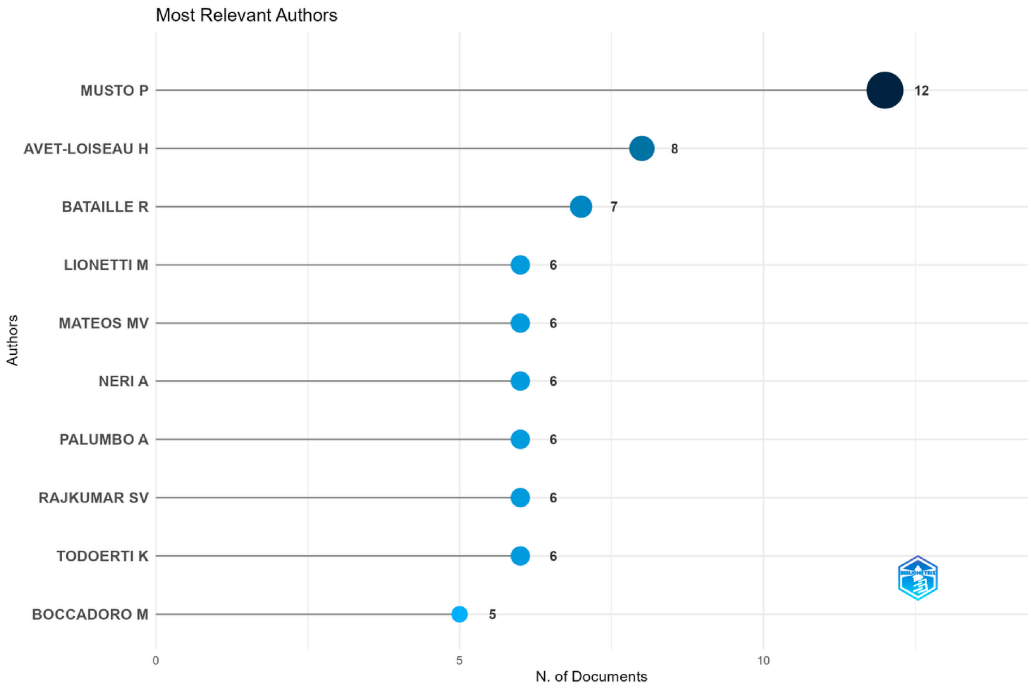


Figure 3. Most Relevant Authors. The 10 most productive authors in the final dataset, ranked by whole-count number of included articles. Musto P. (n = 12), Avet-Loiseau H. (n = 8), and Bataille R. (n = 7) were the most prolific. Potentially ambiguous high-frequency author labels were checked against available Scopus Author Identifiers as described in Section 2.5; the plotted counts remain the standard Biblioshiny name string-based outputs.

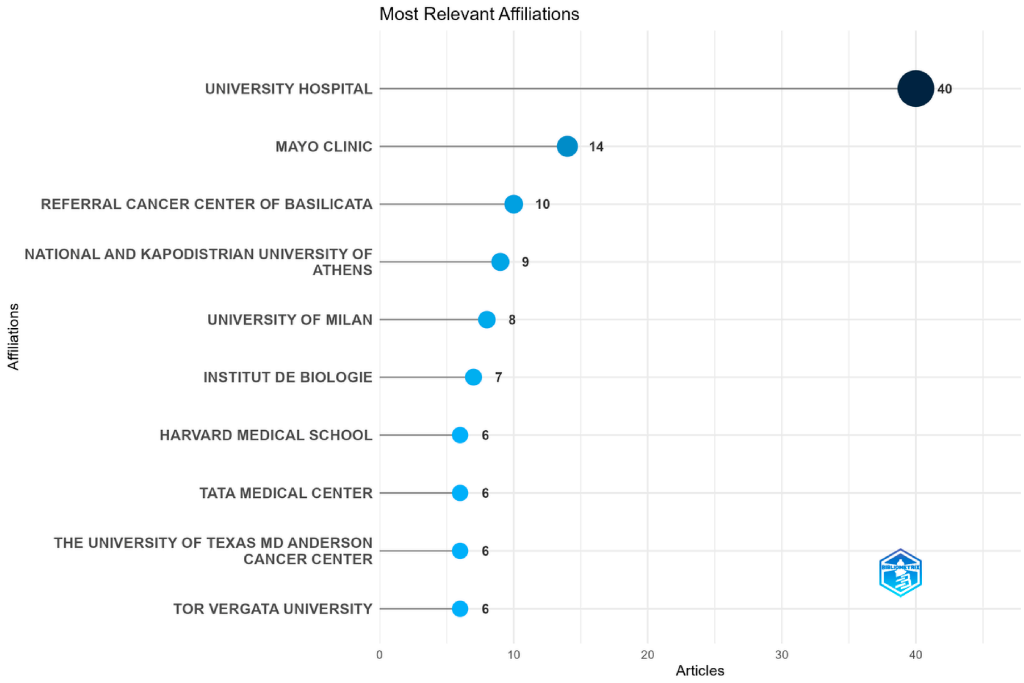


Figure 4. Most Relevant Affiliations. The 10 affiliation labels with the highest number of affiliated articles, based on automated extraction and grouping of the Scopus Affiliations field. The top-ranked entry, "University Hospital" (n = 40), is a generic Scopus affiliation label that aggregates multiple distinct hospitals and should not be interpreted as a single leading institution. Among specifically named institutions, Mayo Clinic (n = 14), the Referral Cancer Center of Basilicata (n = 10), and the National and Kapodistrian University of Athens (n = 9) were the most productive.

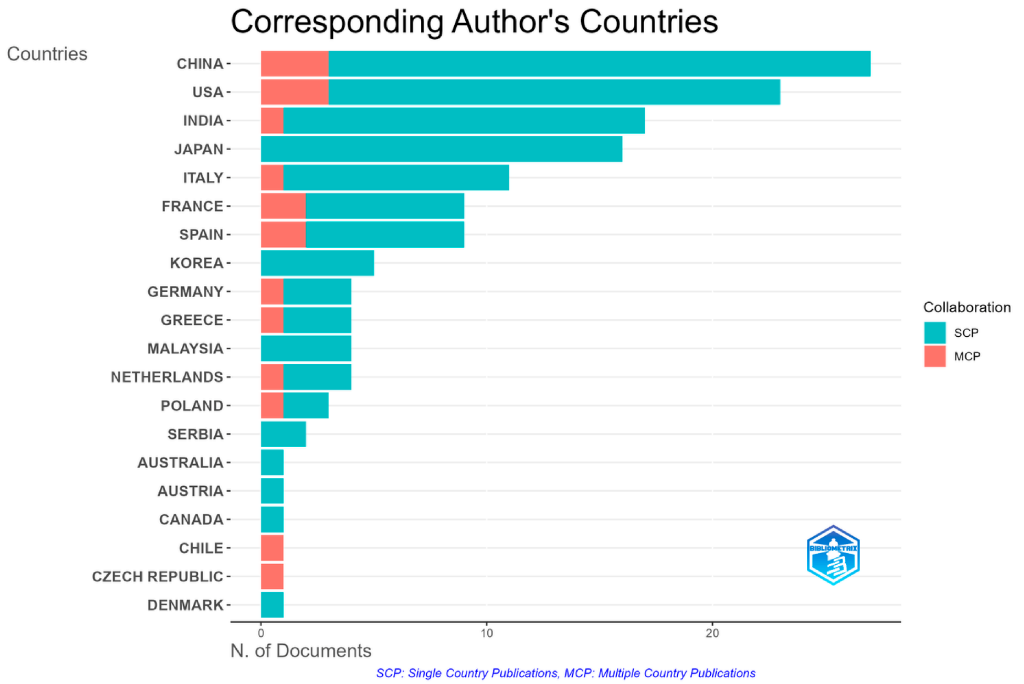


Figure 5. Corresponding Author's Countries: Single- vs. Multiple-Country Publications. Corresponding-author country, ranked by article count, for the 152 of 210 included articles (72.4%) with an identifiable corresponding-author country in the Scopus record; 58 articles (27.6%) lacked this field and are not represented. Publications are classified as single-country publications (SCPs; teal) or multiple-country publications (MCPs; coral). China (n = 27; 24 SCP, 3 MCP), the United States (n = 23; 20 SCPs and 3 MCPs), and India (n = 17; 16 SCPs and 1 MCP) were the three leading corresponding-author countries among records with identifiable data.

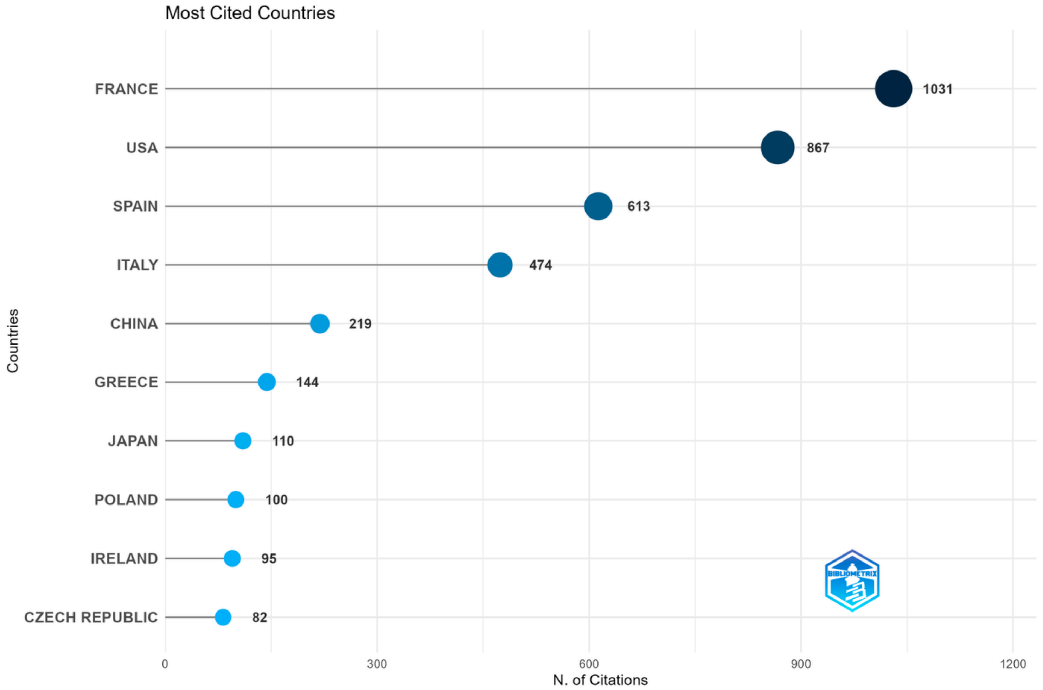


Figure 6. Most Cited Countries. Total citation count received by articles attributed to each corresponding-author country (raw totals, not normalized by article count or publication year). France (1031 total citations; mean 114.6 per article), the United States (867; mean 37.7 per article), and Spain (613; mean 68.1 per article) received the highest total citation counts. Citation totals are influenced by both publication volume and publication age and should be interpreted accordingly.

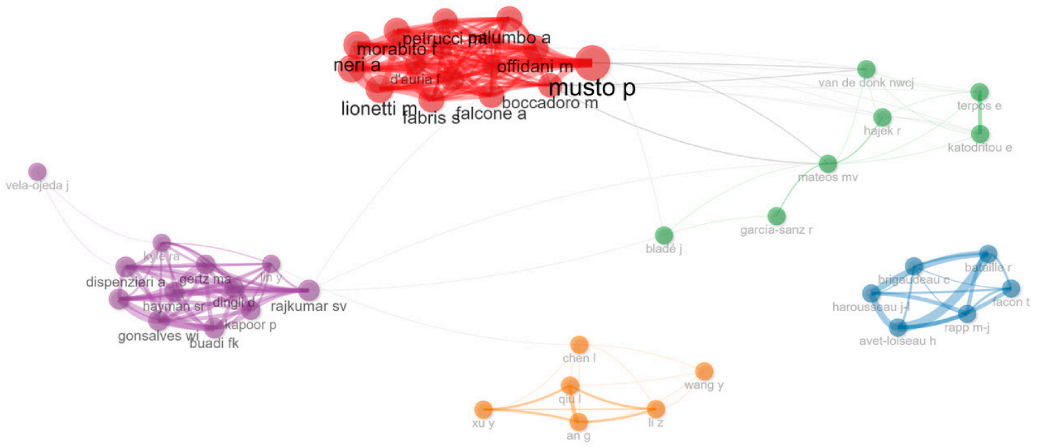


Figure 7. Author Collaboration Network. Co-authorship network extracted from the final dataset using bibliometrix network settings (association strength; Louvain community detection; up to 50 labeled nodes). Nodes represent authors and are sized by number of articles; an edge connects authors who co-authored at least one included article, with edge thickness proportional to the number of co-authored articles. Node and edge colors denote algorithmically identified collaboration clusters and do not represent country or institution. Five main algorithmically identified collaboration clusters were observed.

The increase in pPCL-related publications has occurred alongside the adoption of novel agent-based treatment strategies in plasma cell malignancies. Proteasome inhibitors, immunomodulatory drugs, and monoclonal antibodies have expanded therapeutic options, although pPCL-specific evidence remains largely derived from retrospective studies and small cohorts [3,7]. The growth in publication output therefore indicates increasing clinical interest but not necessarily a corresponding increase in high-level prospective evidence.

Early clinical descriptions established the aggressive presentation of pPCL [11]. Subsequent studies identified frequent high-risk cytogenetic lesions, including deletion 17p, chromosome 1q abnormalities, and complex karyotypes [12–14]. These features may partly explain the poor outcomes observed in pPCL compared with multiple myeloma.

Genomic studies have added to this understanding by identifying increased proliferative activity, genomic instability, and alterations in pathways involving MYC, cell-cycle regulation, and apoptosis [15]. These findings support the view that pPCL has a distinct biological profile rather than simply representing an advanced stage of multiple myeloma.

Compared with multiple myeloma, pPCL more often harbors complex karyotypes and high-risk cytogenetic lesions [16]. Early dissemination into the peripheral blood may reflect a more genomically unstable phenotype [17]. Spatial heterogeneity and the coexistence of different tumor clones may further contribute to disease progression and treatment resistance [18].

Treatment of pPCL has gradually shifted from conventional chemotherapy toward regimens incorporating novel agents. Bortezomib-based combinations are commonly used for induction because of their activity in aggressive plasma cell disorders [19]. Immunomodulatory drugs, including lenalidomide and pomalidomide, may also be used as components of combination regimens [20]. Although these approaches have improved treatment responses, durable disease control remains difficult.

Autologous stem cell transplantation remains an important component of treatment for eligible patients with pPCL. In an EBMT registry analysis of 272 patients undergoing autologous transplantation, the median progression-free survival and overall survival were 14.3 and 25.7 months, respectively, although relapse remained frequent [21]. In a more recent multicenter study of 110 patients with pPCL, treatment with VRd or daratumumab-based quadruplets was associated with a higher complete response rate (41% vs. 17%), longer median progression-free survival (25 vs. 13 months), and longer overall survival (not reached vs. 20 months) compared with standard bortezomib-based combinations or conventional chemotherapy [22]. These results are encouraging, but the retrospective, non-randomized design limits direct comparisons between treatment groups.

BCMA- and GPRC5D-directed immunotherapies are emerging as potential treatment options for PCL. In a multicenter retrospective study of 122 patients with primary or secondary PCL, 27% of whom had pPCL, talquetamab was associated with more favorable survival outcomes than teclistamab or elranatamab, particularly among patients with active PCL [23]. Anti-BCMA CAR T-cell therapy has also shown clinical activity in two patients with relapsed or refractory pPCL included in a phase 1 study [24]. Although these findings are encouraging, they are based on small or heterogeneous patient groups. No randomized head-to-head study has compared bispecific antibodies with CAR T-cell therapy in pPCL; therefore, the available evidence cannot establish the superiority of either approach.

Current pPCL research continues to focus on disease biology, high-risk cytogenetic features, and evolving treatments. Because of its close biological and clinical relationship with multiple myeloma, many therapeutic advances in myeloma have been adapted to pPCL [20,25]. However, the distinct biology and aggressive clinical course of pPCL highlight the need for studies designed specifically for this rare disease [3].

Genomic characterization remains an important direction in pPCL research. Sequencing studies have revealed considerable genomic complexity and heterogeneous mutational patterns, helping to identify potential disease mechanisms and therapeutic targets [26,27]. Integrating molecular findings with clinical data may support improved risk stratification and more individualized treatment approaches, although prospective validation is needed.

Relapsed/refractory extramedullary multiple myeloma represents another aggressive plasma cell phenotype associated with poor outcomes. Although extramedullary multiple myeloma and pPCL are distinct clinical entities, they may share biological features associated

with treatment resistance, including reduced dependence on the bone marrow microenvironment and the presence of high-risk genomic abnormalities [5,26–28]. In the phase 2 RedirecTT-1 study of 90 patients with drug-resistant true extramedullary myeloma, talquetamab plus teclistamab achieved an overall response rate of 79%, including a complete response or better in 54% of patients [29]. Although promising, these findings were obtained in patients with extramedullary myeloma and cannot be directly extrapolated to pPCL. Nevertheless, they provide a rationale for evaluating dual GPRC5D and BCMA targeting in prospective studies specifically involving patients with pPCL.

By combining publication output, citation impact, contributor profiles, and collaboration patterns, this bibliometric analysis shows how pPCL research has developed over time. It also highlights two continuing gaps in the field: limited international co-authorship and the predominance of descriptive and retrospective evidence. These findings may help guide priorities for future multicenter research.

This study has several limitations inherent to bibliometric analyses. First, the dataset was derived from a single database (Scopus), and publications indexed exclusively in other databases may not have been captured. Second, the exact-phrase search strategy prioritized specificity but may have reduced search sensitivity. In particular, older publications or studies reporting separately identifiable pPCL data under broader or alternative terminology may not have been retrieved if the predefined phrases were absent from the title, abstract, and indexed keywords. Therefore, the analyzed corpus represents the literature captured by the predefined search strategy and should not be considered an exhaustive collection of all publications potentially relevant to pPCL. Third, citation counts may favor older publications because they have had more time to accumulate citations. Despite these limitations, the present analysis provides a comprehensive overview of the global research landscape of primary plasma cell leukemia within the validated corpus.

5. Conclusions

This bibliometric analysis provides a structured overview of global pPCL research published between 1983 and 2026. Publication activity increased over time but remained limited, and research output was concentrated in specialist hematology journals. International co-authorship accounted for only 10.95% of the analyzed articles despite the presence of established collaboration clusters. The frequency of indexing terms related to case reports and retrospective studies also suggests that the available literature continues to rely heavily on descriptive and retrospective evidence. Broader multicenter and international collaboration is needed to support larger patient cohorts and more informative prospective studies in this rare disease.

Author Contributions: Conceptualization, Ö.A. and S.G.; Methodology, Ö.A. and S.G.; Investigation, Ö.A. and S.G.; Writing—Original Draft Preparation, Ö.A.; Writing—Review and Editing, Ö.A. and S.G. All authors have read and agreed to the published version of the manuscript.

Funding: This research received no external funding.

Institutional Review Board Statement: Ethical review and approval were waived for this study because it is a bibliometric analysis based exclusively on publicly available published literature and did not involve human participants, animals, or identifiable personal data.

Informed Consent Statement: Not applicable.

Data Availability Statement: The data analyzed in this study were obtained from the Scopus database using the search strategy described in Section 2. Additional information is available from the corresponding author upon reasonable request.

Conflicts of Interest: The authors declare no conflicts of interest.

References

1. Fernández de Larrea, C.; Kyle, R.; Rosiñol, L.; Paiva, B.; Engelhardt, M.; Usmani, S.; Caers, J.; Gonsalves, W.; Schjesvold, F.; Merlini, G.; et al. Primary plasma cell leukemia: Consensus definition by the International Myeloma Working Group according to peripheral blood plasma cell percentage. *Blood Cancer J.* **2021**, *11*, 192. [CrossRef] [PubMed] [PubMed Central]
2. Jung, S.H.; Kim, K.; Yoon, S.E.; Moon, J.H.; Kim, D.; Kim, H.J.; Kim, M.K.; Kim, K.H.; Lee, H.J.; Lee, J.H.; et al. Validation of the revised diagnostic criteria for primary plasma cell leukemia by the Korean Multiple Myeloma Working Party. *Blood Cancer J.* **2022**, *12*, 157. [CrossRef] [PubMed] [PubMed Central]
3. Gong, Z.; Khosla, M.; Vasudevan, S.; Mohan, M. Current Status on Management of Primary Plasma Cell Leukemia. *Curr. Oncol. Rep.* **2024**, *26*, 1104–1112. [CrossRef] [PubMed]
4. Mina, R.; D'Agostino, M.; Cerrato, C.; Gay, F.; Palumbo, A. Plasma cell leukemia: Update on biology and therapy. *Leuk. Lymphoma* **2017**, *58*, 1538–1547. [CrossRef] [PubMed]
5. Hanamura, I.; Karnan, S.; Ota, A.; Takami, A. Primary Plasma Cell Leukemia: Recent Advances in Molecular Understanding and Treatment Approaches. *Int. J. Mol. Sci.* **2025**, *26*, 6166. [CrossRef] [PubMed] [PubMed Central]
6. Musto, P.; Wäsch, R. Plasma cell leukemia: Another piece of the puzzle. *Haematologica* **2023**, *108*, 941–944. [CrossRef] [PubMed] [PubMed Central]
7. Ataş, Ü.; Salim, O.; Iltar, U.; Yücel, O.K.; Küçükdiler Eroğlu, A.H.; Aslan, V.; Yıldırım, M.; Dedeoğlu, Ö.; Seçilmiş, S.; Ulaş, T.; et al. Survival Outcomes of Patients with Primary Plasma Cell Leukemia in the Era of Proteasome Inhibitors and Immunomodulatory Agents: A Real-Life Multicenter Analysis. *Turk. J. Hematol.* **2024**, *41*, 225–235. [CrossRef] [PubMed] [PubMed Central]
8. Gundesen, M.T.; Lund, T.; Moeller, H.E.H.; Abildgaard, N. Plasma Cell Leukemia: Definition, Presentation, and Treatment. *Curr. Oncol. Rep.* **2019**, *21*, 8. [CrossRef] [PubMed] [PubMed Central]
9. Aria, M.; Cuccurullo, C. Bibliometrix: An R-tool for comprehensive science mapping analysis. *J. Informetr.* **2017**, *11*, 959–975. [CrossRef]
10. Donthu, N.; Kumar, S.; Mukherjee, D.; Pandey, N.; Lim, W.M. How to conduct a bibliometric analysis: An overview and guidelines. *J. Bus. Res.* **2021**, *133*, 285–296. [CrossRef]
11. Kyle, R.A.; Maldonado, J.E.; Bayrd, E.D. Plasma cell leukemia. Report on 17 cases. *Arch. Intern. Med.* **1974**, *133*, 813–818. [CrossRef] [PubMed]
12. Tiedemann, R.E.; Gonzalez-Paz, N.; Kyle, R.A.; Santana-Davila, R.; Price-Troska, T.; Van Wier, S.A.; Chng, W.J.; Ketterling, R.P.; Gertz, M.A.; Henderson, K.; et al. Genetic aberrations and survival in plasma cell leukemia. *Leukemia* **2008**, *22*, 1044–1052. [CrossRef] [PubMed] [PubMed Central]
13. Gonsalves, W.I.; Rajkumar, S.V.; Go, R.S.; Dispenzieri, A.; Gupta, V.; Singh, P.P.; Buadi, F.K.; Lacy, M.Q.; Kapoor, P.; Dingli, D.; et al. Trends in survival of patients with primary plasma cell leukemia: A population-based analysis. *Blood* **2014**, *124*, 907–912. [CrossRef] [PubMed] [PubMed Central]
14. Jurczyszyn, A.; Radocha, J.; Davila, J.; Fiala, M.A.; Gozzetti, A.; Grząśko, N.; Robak, P.; Hus, I.; Waszczuk-Gajda, A.; Guzicka-Kazimierczak, R.; et al. Prognostic indicators in primary plasma cell leukaemia: A multicentre retrospective study of 117 patients. *Br. J. Haematol.* **2018**, *180*, 831–839. [CrossRef] [PubMed]
15. Usmani, S.Z.; Nair, B.; Qu, P.; Hansen, E.; Zhang, Q.; Petty, N.; Waheed, S.; Shaughnessy, J.D., Jr.; Alsayed, Y.; Heuck, C.J.; et al. Primary plasma cell leukemia: Clinical and laboratory presentation, gene-expression profiling and clinical outcome with Total Therapy protocols. *Leukemia* **2012**, *26*, 2398–2405. [CrossRef] [PubMed] [PubMed Central]
16. Boyle, E.M.; Davies, F.E.; Leleu, X.; Morgan, G.J. Understanding the multiple biological aspects leading to myeloma. *Haematologica* **2014**, *99*, 605–612. [CrossRef] [PubMed] [PubMed Central]
17. Pawlyn, C.; Morgan, G.J. Evolutionary biology of high-risk multiple myeloma. *Nat. Rev. Cancer* **2017**, *17*, 543–556. [CrossRef] [PubMed]
18. Rasche, L.; Chavan, S.S.; Stephens, O.W.; Patel, P.H.; Tytarenko, R.; Ashby, C.; Bauer, M.; Stein, C.; Deshpande, S.; Wardell, C.; et al. Spatial genomic heterogeneity in multiple myeloma and plasma cell leukemia. *Nat. Commun.* **2017**, *8*, 268. [PubMed]
19. Sonneveld, P.; Broijl, A. Treatment of relapsed and refractory multiple myeloma. *Haematologica* **2016**, *101*, 396–406, Erratum in *Haematologica* **2016**, *101*, 995. https://doi.org/10.3324/haematol.2016.148882. [CrossRef] [PubMed] [PubMed Central]
20. Rajkumar, S.V. Multiple myeloma: 2020 update on diagnosis, risk-stratification and management. *Am. J. Hematol.* **2020**, *95*, 548–567, Erratum in *Am. J. Hematol.* **2020**, *95*, 1444. https://doi.org/10.1002/ajh.25876. [CrossRef] [PubMed]
21. Drake, M.B.; Iacobelli, S.; van Biezen, A.; Morris, C.; Apperley, J.F.; Niederwieser, D.; Björkstrand, B.; Gahrton, G. European Group for Blood and Marrow Transplantation and the European Leukemia Net. Primary plasma cell leukemia and autologous stem cell transplantation. *Haematologica* **2010**, *95*, 804–809. [CrossRef] [PubMed] [PubMed Central]
22. Katodritou, E.; Kastritis, E.; Dalampira, D.; Delimpasi, S.; Spanoudakis, E.; Labropoulou, V.; Ntanasis-Stathopoulos, I.; Gkioka, A.I.; Giannakoulas, N.; Kanellias, N.; et al. Improved survival of patients with primary plasma cell leukemia with VRd or daratumumab-based quadruplets: A multicenter study by the Greek myeloma study group. *Am. J. Hematol.* **2023**, *98*, 730–738. [CrossRef] [PubMed]

23. Gaballa, M.R.; Julian, K.; Afrough, A.; Hansen, D.K.; Goel, U.; de Menezes Silva Corraes, A.; Dima, D.; Rana, M.; Hosoya, H.; Mikkilineni, L.; et al. Real-world outcomes with BCMA- and GPRC5D-targeting bispecific antibodies in plasma cell leukemia. *Blood Adv.* **2026**, *10*, 5938–5950. [CrossRef] [PubMed] [PubMed Central]
24. Li, C.; Cao, W.; Que, Y.; Wang, Q.; Xiao, Y.; Gu, C.; Wang, D.; Wang, J.; Jiang, L.; Xu, H.; et al. A phase I study of anti-BCMA CAR T cell therapy in relapsed/refractory multiple myeloma and plasma cell leukemia. *Clin. Transl. Med.* **2021**, *11*, e346. [CrossRef] [PubMed] [PubMed Central]
25. Kumar, S.K.; Rajkumar, S.V. The multiple myelomas—Current concepts in cytogenetic classification and therapy. *Nat. Rev. Clin. Oncol.* **2018**, *15*, 409–421. [CrossRef] [PubMed]
26. Schinke, C.; Boyle, E.M.; Ashby, C.; Wang, Y.; Lyzogubov, V.; Wardell, C.; Qu, P.; Hoering, A.; Deshpande, S.; Ryan, K.; et al. Genomic analysis of primary plasma cell leukemia reveals complex structural alterations and high-risk mutational patterns. *Blood Cancer J.* **2020**, *10*, 70. [CrossRef] [PubMed] [PubMed Central]
27. Cifola, I.; Lionetti, M.; Pinatel, E.; Todoerti, K.; Mangano, E.; Pietrelli, A.; Fabris, S.; Mosca, L.; Simeon, V.; Petrucci, M.T.; et al. Whole-exome sequencing of primary plasma cell leukemia discloses heterogeneous mutational patterns. *Oncotarget* **2015**, *6*, 17543–17558. [CrossRef] [PubMed] [PubMed Central]
28. Ho, M.; Paruzzo, L.; Minehart, J.; Nabar, N.; Noll, J.H.; Luo, T.; Garfall, A.; Zanwar, S. Extramedullary Multiple Myeloma: Challenges and Opportunities. *Curr. Oncol.* **2025**, *32*, 182. [CrossRef] [PubMed] [PubMed Central]
29. Kumar, S.; Mateos, M.V.; Ye, J.C.; Atrash, S.; Magen, H.; Quach, H.; Chu, M.P.; Trudel, S.; Richter, J.; Rodríguez-Otero, P.; et al. Dual Targeting of Extramedullary Myeloma with Talquetamab and Teclistamab. *N. Engl. J. Med.* **2026**, *394*, 51–61. [CrossRef] [PubMed]

Disclaimer/Publisher's Note: The statements, opinions and data contained in all publications are solely those of the individual author(s) and contributor(s) and not of MDPI and/or the editor(s). MDPI and/or the editor(s) disclaim responsibility for any injury to people or property resulting from any ideas, methods, instructions or products referred to in the content.