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Single-center real-world data on the efficacy, safety, and current availability of pegylated interferon-α in hematologic neoplasms

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Abstract

Aims: Pegylated interferon-α (PEG-IFN-α) offers improved pharmacokinetics compared with conventional interferon-α (IFN-α), yet real-world data on its clinical activity and safety across heterogeneous hematologic neoplasms are limited. This study aimed to evaluate the real-world efficacy, durability of response, and toxicity of PEG-IFN-α in patients with diverse hematologic neoplasms.

Methods: This retrospective study evaluated the efficacy, duration of response, and toxicity of PEG-IFN-α using patient medical records and hospital electronic registries. Thirty patients were included: polycythemia vera (PV, n=12), essential thrombocytosis (ET, n=6), chronic myeloid leukemia (n=2), primary myelofibrosis (n=1), systemic mastocytosis (SM,n=3), hypereosinophilic syndrome (HES, n=1), Erdheim–Chester disease (ECD, n=4), and lymphomatoid granulomatosis (LYG, n=1).

Results: PEG-IFN-α was initiated due to resistance to prior therapies in 12 patients (40%), intolerance or toxicity in 10 patients (33.3%), and as first-line treatment in 8 patients (26.7%). Among PV patients, a complete response was achieved in 41.6% and a partial response in 50%. In ET patients, 83.3% achieved a complete response, while 16.7% showed no response. All patients with SM demonstrated clinical improvement when PEG-IFN-α was used as first-line therapy. In ECD patients, follow-up PET imaging showed stable disease in two patients, partial response in one, and no response in one. Partial responses were also observed in patients with HES and LYG. Treatment-related toxicity occurred in 8 patients (26.6%) and led to treatment discontinuation in 6 patients (20%) (including cytopenias, influenza-like symptoms, and elevated liver enzymes).

Conclusion: In this real-world cohort, PEG-IFN-α showed encouraging activity across several hematologic neoplasms, with toxicity and discontinuation rates in line with previously published series of conventional interferon-α; however, its clinical use remains limited by regulatory and access constraints.

Keywords: pegylated interferon-α, myeloproliferative disorders, Erdheim–Chester disease, lymphomatoid granulomatosis

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Introduction

Interferon-α (IFN-α) is an immunomodulatory agent with well-established antitumor activity and a long history of use across a broad spectrum of hematologic neoplasms. By targeting abnormal hematopoietic clones, IFN-α has demonstrated clinical efficacy in disorders including chronic myeloid leukemia (CML), classical myeloproliferative neoplasms, systemic mastocytosis (SM), hypereosinophilic syndrome (HES), selected histiocytic disorders, and rare lymphoproliferative diseases. However, the widespread clinical use of conventional IFN-α has been limited by its unfavorable toxicity profile and inconvenient dosing schedules, resulting in substantial treatment discontinuation.

Pegylated interferon-α (PEG-IFN-α), developed through covalent conjugation with polyethylene glycol, exhibits improved pharmacokinetic properties, including a prolonged serum half-life, reduced clearance, and more stable drug exposure, thereby enabling once-weekly administration and improved tolerability. In chronic-phase CML, prospective trials have primarily explored PEG-IFN-α in combination with tyrosine kinase inhibitors (TKIs), with several randomized, phase III studies demonstrating deeper molecular responses than TKI monotherapy in selected patient populations [1].

Beyond CML, PEG-IFN-α has an established evidence base in classical myeloproliferative neoplasms — particularly polycythemia vera (PV) and essential thrombocytosis (ET)—where prospective studies and long-term follow-up cohorts have reported durable hematologic control and, in some series, molecular responses [2]. In contrast, evidence supporting PEG-IFN-α in non-myeloproliferative hematologic neoplasms and rare entities remains fragmented, largely derived from small observational cohorts or disease-specific case series. In SM, interferon-based therapy has historically served as a cytoreductive option, although its role has diminished with the advent of KIT-targeted agents [3]. Similarly, IFN-α demonstrated early clinical activity in Erdheim–Chester disease (ECD), but contemporary management increasingly favors molecularly targeted therapies when actionable alterations are present [4,5]. In lymphomatoid granulomatosis (LYG), IFN-α has been used as an immunomodulatory approach in selected patients, although data on PEG-IFN-α remain limited [6,7].

In this context, real-world data addressing treatment indications, response patterns, tolerability, and discontinuation rates of PEG-IFN-α across heterogeneous and rare hematologic conditions are particularly informative. Moreover, the evolving regulatory landscape has created a paradox in which interferon-based therapies with long-standing biological and clinical activity may face restricted accessibility in routine practice, while closely related formulations receive indication-specific approvals. This discrepancy further underscores the importance of real-world evidence beyond formal licensing frameworks.

Therefore, this study aimed to evaluate real-world treatment indications, clinical responses, toxicity profiles, and discontinuation rates of PEG-IFN-α in a heterogeneous cohort of patients with hematologic neoplasms and rare entities treated at a tertiary hematology center, and to contextualize these findings within the existing literature, particularly beyond classical myeloproliferative neoplasms.

METHODS

Patients and Methods

Study Design and Patient Selection

This retrospective, single-center study was conducted at Hacettepe University Faculty of Medicine, Department of Hematology. Patients with a diagnosis of hematologic neoplasia who received PEG-IFN-α between January 2014 and August 2016 were identified by screening institutional drug approval records. At the time of treatment, PEG-IFN-α had no established reimbursement indication for hematologic neoplasms in the Turkish Social Security Institution Health Implementation Notification (SUT). Therefore, all patients included in this study received PEG-IFN-α via individual off-label drug-use approvals obtained in accordance with national reimbursement regulations and institutional authorization procedures.

Only patients who had received formal institutional approval and were followed at our center were included in the analysis. Clinical and diagnostic data were obtained from the hospital electronic medical record system and patient files.

Data Collection

The following data were recorded: demographic characteristics, date of diagnosis, previous treatments and treatment responses, indication for initiation of PEG-IFN-α, treatment start date and duration, response to PEG-IFN-α, occurrence of treatment-related toxicity, last follow-up date, and survival status when applicable.

Response and Toxicity Assessment

Treatment responses were evaluated using disease-specific standardized criteria:

European LeukemiaNet (ELN) criteria for CML [8]

International Working Group–Myeloproliferative Neoplasms Research and Treatment (IWG-MRT) criteria for PV and ET [9]

European Myelofibrosis Network (EUMNET) criteria for primary myelofibrosis [10]

Nordic proposal criteria for hypereosinophilic syndrome [11]

Modified consensus criteria for systemic mastocytosis [12]

Cheson criteria for lymphomatoid granulomatosis [13]

Positron emission tomography (PET) imaging for response assessment in ECD [14]

Treatment-related adverse events were classified according to the Common Terminology Criteria for Adverse Events (CTCAE), version 4.0 [15].

Statistical Analysis

Statistical analyses were performed using IBM SPSS Statistics for Windows, version 22.0 (IBM Corp., Chicago, IL, USA). Given the retrospective and descriptive nature of the study and the limited sample size, analyses were primarily descriptive. Categorical variables were summarized as counts and percentages, whereas continuous variables were reported as median and range. No formal hypothesis testing or inferential statistical comparisons were conducted.

RESULTS

Patient Characteristics

A total of 30 patients were included (female/male: 17/13), and the median age at treatment initiation was 48 years (range, 23–73). Comorbid conditions were present in 70% of patients (n=21), most commonly hypertension (33.3%, n=10) and a history of thrombosis (33.3%, n=10). At the last follow-up assessment, 86.7% of the cohort (n=26) were alive.

The diagnostic distribution included polycythemia vera (PV, n=12), essential thrombocytosis (ET, n=6), chronic myeloid leukemia (CML, n=2), primary myelofibrosis (PMF, n=1), systemic mastocytosis (SM, n=3), hypereosinophilic syndrome (HES, n=1), Erdheim–Chester disease (ECD, n=4), and lymphomatoid granulomatosis (LYG, n=1).

A prior history of arterial or venous thrombosis was documented in 33.3% of patients (n=10). No arterial or venous thrombotic events occurred during PEG-IFN-α treatment.

Treatment Indications

Overall, PEG-IFN-α was initiated due to resistance to previous therapies in 40% of patients (n=12) and due to intolerance or toxicity related to prior treatments in 33.3% (n=10). In the remaining 26.7% of patients (n=8), PEG-IFN-α was administered as first-line therapy.

All patients with PV, ET, CML, and PMF had previously received standard treatments and were switched to PEG-IFN-α due to treatment resistance or intolerance.

Treatment Response in Classical Myeloproliferative Neoplasms

Among patients with PV, the best response to PEG-IFN-α was complete response in 41.7% (n=5), partial response in 50% (n=6), and no response in 8.3% (n=1). The median time to best response was 4.1 months (range, 0.5–21). None of the PV patients required phlebotomy during treatment. Treatment discontinuation due to toxicity occurred in 50% of PV patients (n=6).

In ET patients, complete response was achieved in 83.3% (n=5), while 16.7% (n=1) showed no response. The median time to best response was 7.3 months (range, 5.1–12.8). No major bleeding or thrombotic events were observed during treatment. Mean platelet counts decreased from 1174.2×10^9/L at baseline to 428.5×10^9/L at 6 months and 405.3×10^9/L at 12 months.

In the two patients with CML, PEG-IFN-α was administered in combination with tyrosine kinase inhibitors due to suboptimal response. One patient achieved a major molecular response, while the other achieved a hematologic response.

The single patient with PMF achieved a partial response, with a median time to best response of 8.1 months.

Baseline patient characteristics, treatment details, response outcomes, and toxicity profiles for patients with PV, ET, CML, and SM are summarized in Table 1.

Treatment Response in Rare Hematologic and Histiocytic Disorders

All three patients with systemic mastocytosis received PEG-IFN-α as first-line therapy and demonstrated clinical improvement. Symptom relief, particularly resolution of pruritus and skin manifestations, was observed at a median of 2 months (range, 1–3).

The patient with hypereosinophilic syndrome achieved a partial response after 13.3 months of treatment.

Among patients with ECD, PET-based response assessment showed stable disease in two patients (50%), partial response in one patient (25%), and no response in one patient (25%). Three patients had diabetes insipidus at baseline without additional pituitary hormone deficiencies, and desmopressin requirements remained unchanged during PEG-IFN-α therapy. One patient developed nephrotic syndrome due to renal involvement of ECD and subsequently died during follow-up.

The patient with lymphomatoid granulomatosis received PEG-IFN-α as first-line therapy and achieved a partial response, which was followed by durable remission, leading to treatment discontinuation.

Patient characteristics, treatment details, clinical responses, and toxicity profiles of patients with Erdheim–Chester disease, primary myelofibrosis,

Table 1. Patient characteristics, treatment details, responses, and toxicity in PV, ET, CML, and SM

Characteristics	PV (n=12)	ET (n=6)	CML (n=2)*	SM (n=3)
Age, median (range)	47 (27–65)	30 (24–63)	52 (48–56)	47 (29–48)
Gender (F/M)	7 / 5	5 / 1	1 / 1	1 / 2
Previous treatments, median (range)	2 (1–3)	2 (1–3)	3	None
Indication for PEG-IFN-α	Resistance to HU/IFN: 6; Intolerance/toxicity: 6	Resistance to HU/anagrelide: 3; Intolerance/toxicity: 3	TKI suboptimal response: 1; Failure: 1	First-line
Duration of PEG-IFN-α, months (median, range)	15.9 (1–69.6)	21.5 (3–61.1)	31.5 (11.07–52.1)	12.1 (9.3–14.8)
Best response	CR: 5; PR: 6; NR: 1	CR: 5; NR: 1	MMR: 1; HR: 1	CI: 3
Time to best response, months (median, range)	4.1 (0.5–21)	7.3 (5.1–12.8)	2.3	Not applicable (subjective data)
Toxicity	Hair loss: 1; Nephropathy: 2; Myelosuppression: 1; Skin rash: 1; Flu-like symptoms: 1	Autoimmune thyroiditis: 1; Autoimmune hepatitis: 1	None	None
Reason for discontinuation	NR: 1; Toxicity: 6; Sustained remission: 1	NR/progression: 2; Sustained remission: 2	Sustained remission: 1	Sustained improvement: 2

* PEG-IFN-α was used in combination with TKI. CI, clinical improvement; CR, complete response; HR, hematologic response; HU, hydroxyurea; IFN, interferon; MMR, major molecular response; NR, no response; PR, partial response; TKI, tyrosine kinase inhibitor.

Table 2. Patient characteristics, treatment details, responses, and toxicity in ECD, PMF, HES, and LYG

Characteristics	ECD (n=4)	PMF (n=1)	HES (n=1)	LYG (n=1)
Age, median (range)	46.5 (23–73)	63	29	68
Gender (F/M)	2 / 2	1 / 0	0 / 1	0 / 1
Previous treatments	Cladribine in 1 patient; none in others	Hydroxyurea	Steroids	None
Indication for PEG-IFN-α	Non-response: 1; First-line: 3	Intolerance to hydroxyurea	First-line	First-line
Duration of PEG-IFN-α, months (median, range)	6.6 (2.53–28.6)	16.6	15	13.3
Best response	SD: 2; PR: 1; NR: 1	PR: 1	PR: 1	CR: 1
Time to best response, months (median, range)	6.6 (5.3–10.9)	8.1	13.3	12
Toxicity	None	None	None	None
Reason for discontinuation	Non-response: 1	–	Sustained remission: 1	Sustained remission: 1

* PEG-IFN-α was partially used in combination with chemotherapy. CR, complete response; HU, hydroxyurea; NR, no response; PR, partial response; SD, stable disease.

hypereosinophilic syndrome, and lymphomatoid granulomatosis are summarized in Table 2.

Toxicity and Treatment Discontinuation

Overall, treatment-related toxicity was observed in 26.6% of patients (n=8), resulting in permanent discontinuation of PEG-IFN-α in 20% (n=6). Reported adverse events included alopecia, skin rash, myelosuppression, nephropathy with nephrotic-range proteinuria, autoimmune thyroiditis, autoimmune hepatitis, and flu-like symptoms. No toxicity was observed in patients with SM, HES, ECD, or LYG.

DISCUSSION

In this retrospective, single-center study, we evaluated the real-world use of PEG-IFN-α across a heterogeneous cohort of patients with myeloproliferative neoplasms and rare hematologic disorders. Our findings suggest that PEG-IFN-α may demonstrate encouraging real-world activity in selected patients, particularly in PV and ET, although treatment discontinuation due to toxicity remains a significant limitation in routine clinical practice.

In PV and ET, our response rates (PV: CR 41.7%, PR 50%; ET: CR 83.3%) are broadly consistent with prior prospective studies reporting high hematologic response rates with pegylated interferons [2,16]. Quintás-Cardama et al. reported overall response rates exceeding 80% in both PV and ET, with higher complete response rates than those observed in our PV cohort; however, frequent partial responses in our patients resulted in a comparable overall disease control rate. In contrast, the high complete response rate observed in our ET patients closely mirrors that reported in prior prospective studies of PEG-IFN-α in ET [2]. More recent phase III trials further support the sustained disease-controlling potential of interferon-based therapies in PV. In the PROUD-PV and CONTINUATION-PV trials, ropeginterferon alfa-2b demonstrated increasing hematologic response rates over time and superior responses compared with hydroxyurea at longer follow-up, despite not meeting non-inferiority criteria at 12 months [17]. Although differences in interferon formulation and study design limit direct comparison, these data are consistent with the durable response patterns observed in our real-world cohort.

Similarly, the phase III trial by Mascarenhas et al. comparing PEG-IFN-α with hydroxyurea in high-risk PV and ET showed no significant differences in complete response or disease progression, while PEG-IFN-α was associated with greater molecular responses but a higher frequency of grade ≥3 adverse events [18]. Consistent with these findings, toxicity-related

treatment discontinuation occurred in a substantial proportion of PV patients in our cohort, underscoring that tolerability remains a key challenge of interferon-based therapy. The relatively high rate of treatment discontinuation due to intolerance or toxicity observed in patients with PV may reflect disease-specific treatment characteristics rather than an intrinsically unfavorable safety profile of PEG-IFN-α. In routine clinical practice, patients with PV often receive PEG-IFN-α over prolonged periods, frequently extending beyond the durations reported in prospective clinical trials. Consequently, longer follow-up and cumulative drug exposure may increase the likelihood of delayed or cumulative interferon-related adverse effects, ultimately leading to higher discontinuation rates. These real-world findings underscore the importance of long-term tolerability monitoring when PEG-IFN-α is used to control chronic disease in PV.

In CML, PEG-IFN-α was administered as an adjunct to tyrosine kinase inhibitors in selected patients with suboptimal responses, resulting in hematologic and molecular responses without treatment-limiting toxicity. These observations align with previous studies suggesting that interferon-based strategies may enhance molecular responses in chronic-phase CML when combined with TKI therapy, although their role has diminished in the era of highly effective TKIs [1,19].

Our findings also align with prior reports demonstrating heterogeneous responses to PEG-IFN-α across Philadelphia chromosome–negative myeloproliferative neoplasms. In a phase II study by Jabbour et al., response rates were highest in ET, while outcomes in PV and MF were more variable, and treatment discontinuation due to toxicity was common [20]. These results are consistent with our real-world experience and emphasize the importance of disease subtype and patient selection.

Data regarding PEG-IFN-α in rare hematologic and histiocytic disorders remain limited. In myelofibrosis, responses are generally less frequent and heterogeneous; our single MF patient achieved a partial response without toxicity, consistent with larger cohorts showing modest hematologic responses but potential clinical benefit in selected patients [21,22].

In hypereosinophilic syndrome, available evidence suggests favorable efficacy and tolerability, and our patient achieved sustained remission with PEG-IFN-α following corticosteroid failure, supporting its role as a steroid-sparing option in selected cases [23,24]. Consistent with the available literature, our single HES patient—initially responsive to corticosteroids but experiencing loss of response—achieved sustained remission after 15 months of PEG-IFN-α therapy without treatment-related complications, supporting its role as an effective steroid-sparing option in selected HES cases.

In systemic mastocytosis, interferon-based therapy is no longer routine first-line treatment but may remain useful for symptom control or cytoreduction in selected patients. All three SM patients in our cohort experienced rapid clinical improvement without treatment-limiting toxicity, supporting a potential role for PEG-IFN-α in carefully selected cases [25,26]. In this context, all three SM patients in our cohort received PEG-IFN-α as first-line therapy and experienced rapid clinical improvement, particularly in pruritus and cutaneous symptoms, with a median time to response of two months and no treatment-limiting toxicity.

Evidence for interferon-based therapy in lymphomatoid granulomatosis is mainly derived from studies using conventional IFN-α. In a pivotal phase II National Cancer Institute study, IFN-α-2b achieved high and durable response rates in low-grade disease, supporting its EBV-driven immunologic nature. Experience with PEG-IFN-α is limited to case reports; however, sustained responses with improved tolerability have been described [27]. Consistent with these reports, our single LYG patient achieved a partial response without significant toxicity, suggesting PEG-IFN-α as a feasible option in selected low-grade cases.

Interferon-based therapy has historically been the main systemic treatment for Erdheim–Chester disease, with evidence supporting improved survival and organ responses. In our cohort, PEG-IFN-α was used as initial therapy in all ECD patients, resulting in disease stabilization or partial response in most cases and no treatment-related toxicity. These findings are consistent with existing literature supporting PEG-IFN-α as an effective conventional option in selected ECD patients, while highlighting the need for alternative or targeted therapies in cases of progression [5,28].

This retrospective, single-center study is limited by the relatively small and heterogeneous sample size. Longer and more standardized follow-up and the inclusion of a comparator group could further strengthen

the assessment of treatment outcomes. In addition, molecular response and patient-reported outcomes were not systematically collected.

Lastly, but perhaps most importantly, our findings also illustrate that proven clinical effectiveness and acceptable toxicity profiles alone may not be sufficient to ensure sustained use of a therapeutic agent in contemporary clinical practice. PEG-IFN-α is not available in Türkiye today. Beyond clinical efficacy and safety, our findings also highlight a broader regulatory and access-related paradox surrounding interferon-based therapies in hematologic neoplasms. Despite decades of evidence demonstrating the disease-controlling and, in some settings, disease-modifying potential of IFN-α—particularly in classical myeloproliferative neoplasms —PEG-IFN-α remains largely unlicensed for most hematologic indications and has become increasingly difficult to access in many countries following the decline in its use for viral hepatitis. In contrast, structurally and biologically related formulations, such as ropeginterferon alfa-2b, have obtained regulatory approval for selected indications (e.g., PV) based on contemporary phase III trial programs. While differences in pegylation and pharmacokinetics exist, these developments reflect regulatory and developmental trajectories rather than a fundamental shift in interferon biology. Consequently, clinically similar interferon-based strategies may now be available under different formulations with substantially different cost and accessibility profiles. Real-world data, such as those presented in this study, remain important to contextualize the clinical value of PEG-IFN-α beyond formal licensing frameworks, particularly in rare diseases and in patient populations for whom alternative therapies are limited, contraindicated, or associated with long-term safety concerns.

In this single-center, real-world cohort, PEG-IFN-α demonstrated measurable clinical activity across a heterogeneous group of hematologic neoplasms, including both classical myeloproliferative neoplasms and selected rare entities. Response patterns varied by disease type, and treatment-related toxicity remained a relevant clinical issue, leading to discontinuation in a substantial proportion of patients. Despite its prolonged half-life and simplified dosing schedule, the overall tolerability profile of PEG-IFN-α was comparable to that reported for conventional interferon-α in previous studies. These findings suggest that PEG-IFN-α represents a feasible therapeutic option in selected patients, particularly in settings where alternative treatments are limited or contraindicated.

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Author contributions

Conception: A.B.B., Y.B.; Design: Y.B.; Data acquisition: A.B.B., Y.B.; Data analysis: A.B.B.; Data interpretation: A.B.B.; Drafting of the manuscript: A.B.B.; Critical revision of the manuscript: Y.B.. All authors reviewed the results, approved the final version of the manuscript, and agreed to be accountable for all aspects of this study.

Ethical approval

This study was approved by the Hacettepe University Health Sciences Research Ethics Committee (Date: February 28, 2017, Decision/Protocol No: GO 17/206-03). Informed consent was obtained from all participants involved in this study.

Data availability statement

The data that support the findings of this study are available from the corresponding author upon reasonable request.

Conflict of interest

The authors declare that this study was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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Generative AI statement

The authors declare that no generative AI or AI-assisted technologies were used in the writing or preparation of this study.

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