Clin. Lab. 2026;72:XXX-XXX

CASE REPORT

A Case of Primary IgG-κ with κ Free Light Chain Plasma Cell Leukemia with Literature Review

Qingfeng Ma, Sai Zhang, Wei Qin

Department of Clinical Laboratory, Feicheng People's Hospital, Feicheng, China

SUMMARY

Background: **The aim of this study was to investigate the clinical and laboratory features of primary IgG-κ with κ free light chain plasma cell leukemia.**
Method: **We retrospectively analyzed the clinical and laboratory features of a case of primary plasma cell leukemia of the IgG-κ with κ free light chain type and reviewed the literature on patients with primary plasma cell leukemia.**
Result: **The patient's white blood cell count was 36.95 x 10^9/L, hemoglobin was 43 g/L, platelet count was 64 x 10^9/L. Push film review: the number of white blood cells was significantly increased, and a type of cell was seen, with medium cytosol, polarized nucleus, abundant cytoplasm, stained areas, and rounded inclusions, which accounted for 90% of the total number of white blood cells. IgG 89.8 g/L, IgA < 0.26 g/L, IgM < 0.26 g/L, complement C3 0.33 g/L, complement C4 0.09 g/L; blood β2 microglobulin > 24.4 mg/L, ferritin 429.72 ng/mL. Serum protein electrophoresis: M protein bands were found, and the M protein content was 71.84 g/L. Serum immunofixation electrophoresis: precipitating bands were found in the IgG lane, two precipitating bands were found in the κ lane, and the monoclonal immunoglobulin type was IgG-κ with κ free light chain type. Flow cytometry: plasma cells accounted for 69.61% of the total, and their immunophenotypes were CD28+, CD38+, CD138+, CD27+ partially, CD269+ in small amount, CD19-, CD20-, and intracellular immunoglobulin Kappa light chain restriction expression, suggesting primary plasma cell leukemia.**
Conclusions: **For primary plasma cell leukemia, we should pay attention to the changes in the abnormal morphology and number of plasma cells. With the help of bone marrow smear, flow cytometry and other tests, we can make a clear diagnosis as early as possible and actively carry out treatment at an early stage.**
(Clin. Lab. 2026;72:xx-xx. DOI: 10.7754/Clin.Lab.2025.250508)

Correspondence:
Wei Qin
Feicheng People's Hospital
Feicheng, Shandong Province 271600
China
Email: 13562880289@163.com

Case Report accepted May 18, 2025

KEYWORDS

primary plasma cell leukemia, diagnostic trait, prognosis

INTRODUCTION

Plasma cell leukemia (PCL), an extremely rare leukemia, is extremely aggressive and has a short patient survival. It accounts for only 2 - 4% of abnormal plasma cell diseases [1]. The distinctive feature of PCL is the emergence of large numbers of abnormal plasma cells in the peripheral blood and bone marrow, and the extensive infiltration of these cells into various organs and

tissues. According to the etiology, PCL can be classified into primary plasma cell leukemia (PPCL) and secondary plasma cell leukemia (SPCL). In this case, the PPCL patient did not have a clear history of multiple myeloma (MM), but rather had typical features of acute leukemia. SPCL mostly comes from the progression of diseases such as MM to end stage [2]. PPCL is more aggressive, less prognostic and difficult to treat compared to SPCL and MM. This article reports an exceptional case of primary plasma cell leukemia. In this case, the percentage of peripheral blood plasma cells was as high as 90% and the percentage of bone marrow plasma cells was as high as 95%, and its type was IgG-κ with κ free light chain type.

CASE REPORT

The patient is a 72-year-old woman who began to experience epigastric discomfort with loss of appetite without any obvious trigger 2 months ago, and then nausea and vomiting, peripheral fatigue, rib pain in the chest, and panicked and suffocating breathlessness on activity more than 10 days ago. Temperature, pulse, respiration, and blood pressure were approximately normal, clear, general spirit, anemic appearance, thick respiratory sounds in both lungs, no obvious dry or wet gong sounds, the heart rate was unison, no obvious murmurs in each valve auscultation area, sternal pressure pain, flat and soft abdomen, light pressure pain in the epigastrium, no rebound pain, liver and spleen were not palpated subcostally, no obvious oedema of the two lower limbs, and no obvious hemorrhage spots on the skin. Temperature, pulse, respiration, blood pressure were approximately normal, clear, general spirit, anemic appearance, thick respiratory sounds in both lungs, no obvious dry and wet gong sounds, heart rate was neat, no obvious murmurs in each valve auscultation area, sternal pressure pain, flat and soft abdomen, light pressure pain in the upper abdomen, no rebound pain, liver and spleen were not palpated subcostally, no obvious edema of the two lower limbs, and no obvious hemorrhage spots on the skin.

Routine blood tests: white blood cell count 36.95 x 10^9/L, hemoglobin 43 g/L, platelet count 64 x 10^9/L, monocyte percentage 77.1%. Upon re-examination of the slides, the red blood cells were seen to be arranged in a cord, the number of white blood cells was significantly increased, and one type of cell was seen, with a medium cytosol, an off-set nucleus, abundant cytoplasm, and visible areas of light staining, and rounded inclusion bodies, which accounted for 90% of the cells (Figure 1). Sedimentation is 25 mm/hour. Urine protein +. Preliminary diagnosis: cause of anemia to be investigated. Liver function tests: total protein 138.9 g/L, albumin 32.1 g/L, globulin 106.8 g/L; aminotransferases, cardiac kinases normal; fibrinogen 1.61 g/L; calcium 2.08 mmol/L, phosphorus 2.08 mmol/L, carbon dioxide 6 mmol/L, potassium, sodium, chloride ions normal; blood glucose normal, urea nitrogen 16.12 mmol/L. creatinine 492 μmol/L. Preliminary diagnosis: renal failure, metabolic acidosis, electrolyte metabolism disorder. It is recommended to improve autoantibody, immunofixation electrophoresis, serum light chain examination. IgG 89.8 g/L, IgA < 0.26 g/L, IgM < 0.26 g/L, complement C3 0.33 g/L, complement C4 0.09 g/L; blood β2 microglobulin > 24.4 mg/L, ferritin 429.72 ng/mL. Serum protein electrophoresis: M protein bands were found, and the M protein content was 71.84 g/L. Serum immunofixation electrophoresis was examined: precipitated bands were found in the IgG lane, two precipitated bands were found in the κ lane, and the type of monoclonal immunoglobulin was the IgG-κ with κ free light-chain type (Figure 2). Serum light chain quantification: Kappa light chain 32.70 g/L, Lamda light chain < 0.50 g/L, Kappa/Lamda ratio significantly elevated, suggesting restricted expression of Kappa light chain. Urine protein electrophoresis: M protein bands were found, and the urinary M protein content was 7,909.22 mg/24 hours. Urine Benjamin Chouin positive, type κ free light chain type. Consider plasma cell disease and perform a bone marrow aspiration and biopsy to assist in the diagnosis. Bone marrow smear morphology: proliferation is obviously active, plasma cells are obviously increased, accounting for 95%, and binucleated and multinucleated plasma cells can be seen. Granulocytes and erythrocytes were suppressed, and mature erythrocytes were arranged in coils. Peripheral blood cell morphology: leukocytes increased in number, plasma cells increased significantly, accounting for 90%, the proportion of granulocytes is reduced, mature erythrocytes arranged in coils. Diagnostic opinion: plasma cell leukemia is not excluded, combine with flow cytology.

Flow cytometry: 69.61% of plasma cells were seen, with an immunophenotype of CD28+, CD38+, CD138+, CD27+ partially, CD269+ in small amounts, CD19-, CD20-, and restrictive expression of the intracellular immunoglobulin Kappa light chain, suggesting monoclonal plasma cells (Figure 3). Bone marrow biopsy: nucleated cells with extremely active proliferation, hematopoietic volume of about 90%, plasma cells were diffusely distributed, the cell cytosol was oversized, the nucleus was rounded or irregular, often parabasic, and the cytoplasm was abundant. Immunohistochemistry: CD38 diffuse +, CD138 diffuse +, κ diffuse +, κ occasional +, CD56 occasional +, CD20 scattered +, CD19 scattered less +, Cyclin-D1 scattered less +. Combined with histomorphology and immunohistochemistry, a plasma cell tumor was considered, with approximately 95% tumor cells. Karyotype analysis: after inoculation and culture, the cells grew poorly and had no analyzable chromosome division phase. Combining the above findings, the diagnosis of plasma cell leukemia was made.

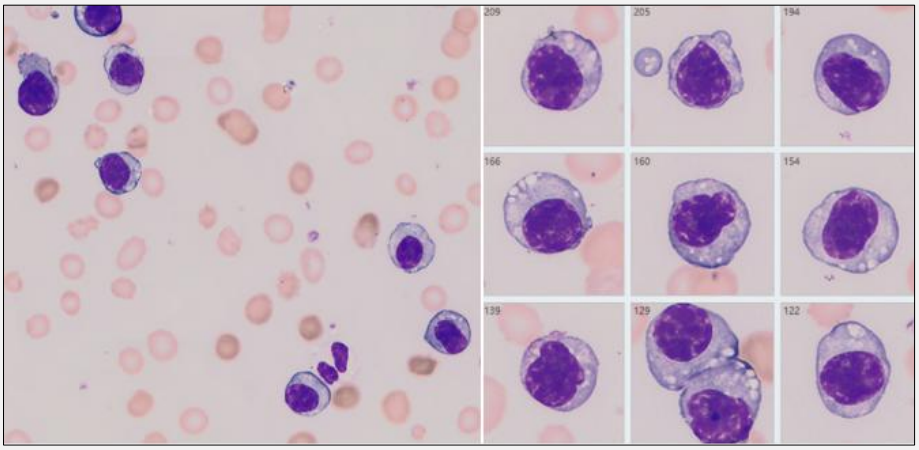

Figure 1. Blood smear showing a large number of cells with medium cytosol, deviated nuclei, abundant cytoplasm with areas of light staining and rounded inclusion bodies.

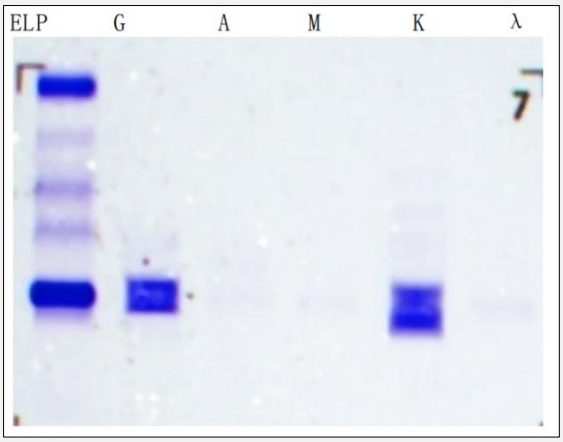

Figure 2. Serum immunofixation electrophoresis: precipitating bands were found in the IgG lane and two precipitating bands were found in the κ lane, and the monoclonal immunoglobulin type was IgG-κ with κ free light chain type.

DISCUSSION

Primary plasma cell leukemia is a rare and aggressive malignancy. The annual worldwide incidence is estimated to be 0.4 to $1.2/10^6$, and few studies have been reported [3]. Definitive diagnosis of plasma cell leukemia is considered to be a percentage of circulating plasma cells in the peripheral blood ≥ 20% and/or an absolute count of circulating plasma cells ≥ 2 x 10^9/L with abnormal morphological changes [4]. Usually, PPCL has an acute onset, with manifestations similar to acute leukemia, such as high fever, anemia, infection, enlargement of liver, spleen and lymph nodes, and sternal pressure pain. In this case, the patient had a peripheral blood plasma cell percentage of up to 90% and a bone marrow plasma cell percentage of up to 95%, with a diffuse distribution of plasma cells, with enlarged cytosol, rounded or irregularly shaped nuclei, deviated, and

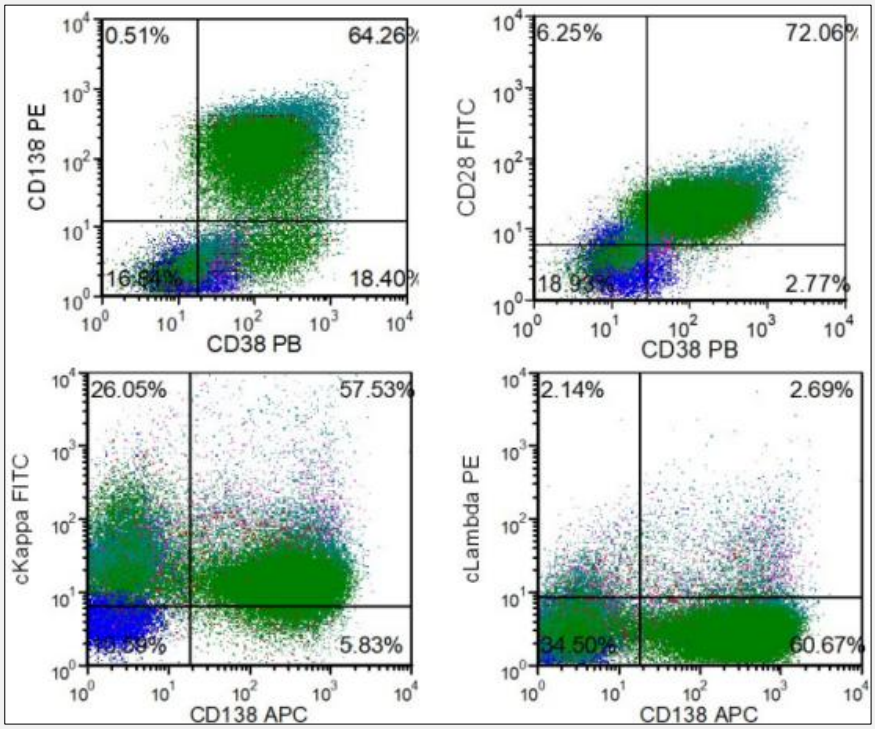

Figure 3. Flow cytometry showing plasma cells expressing high levels of CD138, CD38, and CD28 with restricted expression of intracellular immunoglobulin kappa light chain.

abundant cytoplasmic mass. It was also accompanied by clinical features similar to acute leukemia, which provided clues for the diagnosis of PPCL.

Studies have claimed that patients with PPCL have elevated leukocyte counts, blood calcium, β2-microglobulin, and lactate dehydrogenase with renal failure, and that these markers indicate the presence of a high tumor load in the patient and correlate with a high proliferation of tumors [5]. Usually, hypercalcemia is often associated with osteolytic lesions and renal insufficiency in patients, and osteolytic lesions are less common than in MM and renal insufficiency is more frequent in patients with PPCL [6]. The symptoms of this patient were consistent with the study report. The difference was that this patient had normal blood calcium despite bone pain and renal failure. Several studies have claimed that hypercalcemia is an independent risk factor for median overall survival in patients with PPCL [7]. This also predicts that the pathophysiologic processes in this patient have not yet significantly affected calcium ion homeostasis. However, when age ≥ 60 years, platelet count ≤ 100 x 10^9/L, and peripheral blood plasma cell count ≥ 20 x 10^9/L are found then patient survival is considered as poor [8].

In primary plasma cell leukemia (PPCL), immunoglobulin types of IgG and IgA are more common. In the case of the present study, it was detected by immunofixation electrophoresis as IgG-κ with κ free light chain type. CD38 and CD138 are important to recognize as typical markers of plasma cells. Compared with multiple myeloma (MM), PPCL patients tend to show higher levels of CD20, CD27, CD28, and CD45 expression on the cell surface. In contrast, CD9, CD56, CD117, and HLA-DR are usually in a state of non-expression in PPCL [9]. In this case analysis, the abnormal plasma cells showed expression of CD38, CD138, and CD28, providing a basis for the diagnosis of PPCL. Studies have shown that CD28 promotes plasma cell proliferation, so patients with high CD28 expression tend to have a poorer prognosis [10].

As yet, there are no standardized treatment protocols for PPCL due to its rapid progression and poor prognosis. Traditionally, chemotherapy is the mainstay, but nowadays, immunomodulators, proteasome inhibitors, targeted drugs such as CD38 monoclonal antibody, chimeric antigen receptor T cells and other new drugs, as well as hematopoietic stem cell transplantation, are mostly used to treat PPCL [11]. Although the survival of the patient was prolonged, his overall survival was still poor. This patient was not followed up and treated at our hospital

after diagnosis. Therefore, when we make a diagnosis of PPCL, we should pay attention to the changes in the abnormal morphology and number of plasma cells, and with the help of bone marrow smear, flow cytometry, and other tests, we should make a clear diagnosis as early as possible, and actively carry out treatment at an early stage.

Declaration of Interest:
None.

References:

1. Gong Z, Khosla M, Vasudevan S, et al. Current Status on Management of Primary Plasma Cell Leukemia. Curr Oncol Rep 2024;26(9):1104-12. (PMID: 38954316)

2. Hayman SR, Fonseca R. Plasma cell leukemia. Curr Treat Options Oncol 2001;2(3):205-16. (PMID: 12057120)

3. Chaulagain CP, Diacovo MJ, Van A, et al. Management of Primary Plasma Cell Leukemia Remains Challenging Even in the Era of Novel Agents. Clin Med Insights Blood Disord 2021;14: 2634853521999389. (PMID: 33716516)

4. Fernandez de Larrea C, Kyle RA, Durie BG, et al. Plasma cell leukemia: consensus statement on diagnostic requirements, response criteria and treatment recommendations by the International Myeloma Working Group. Leukemia 2013;27(4):780-91. (PMID: 23288300)

5. Gonsalves WI, Rajkumar SV, Dispenzieri A, et al. Quantification of circulating clonal plasma cells via multiparametric flow cytometry identifies patients with smoldering multiple myeloma at high risk of progression. Leukemia 2017;31(1):130-5. (PMID: 27457702)

6. Jurczyszyn A, Radocha J, Davila J, et al. Prognostic indicators in primary plasma cell leukaemia: a multicentre retrospective study of 117 patients. Br J Haematol 2018;180(6):831-9. (PMID: 29315478)

7. Nakaya A, Yagi H, Kaneko H, et al. Retrospective analysis of primary plasma cell leukemia in Kansai Myeloma Forum registry. Leuk Res Rep 2018;10:7-10. (PMID: 30013912)

8. Ravi P, Kumar SK, Roeker L, et al. Revised diagnostic criteria for plasma cell leukemia: results of a Mayo Clinic study with comparison of outcomes to multiple myeloma. Blood Cancer J 2018;8(12):116. (PMID: 30442928)

9. Guo R, Hu F, Weng Q, et al. Guiding T lymphopoiesis from pluripotent stem cells by defined transcription factors. Cell Res 2020;30(1):21-33. (PMID: 31729468)

10. Pellat-Deceunynck C, Bataille R, Robillard N, et al. Expression of CD28 and CD40 in human myeloma cells: a comparative study with normal plasma cells. Blood 1994;84(8):2597-603. (PMID: 7522634)

11. Tuazon SA, Holmberg LA, Nadeem O, Richardson PG. A clinical perspective on plasma cell leukemia; current status and future directions. Blood Cancer J 2021;11(2):23. (PMID: 33563906)