

Primary plasma cell leukaemia presenting as unilateral proptosis

Irene T. Schrijver¹ | Esta Kalikmanov-Mikhaylovskaya² | Yorick Sandberg¹ 

¹Department of Internal Medicine, Maastad Hospital, Rotterdam, The Netherlands

²Department of Ophthalmology, Maastad Hospital, Rotterdam, The Netherlands

Correspondence

Yorick Sandberg, Department of Internal Medicine, Maastad Hospital, Maastadweg 21, Rotterdam 3079 DZ, The Netherlands.

Email: SandbergY@maastadziekenhuis.nl

KEYWORDS

diplopia, haematology, primary plasma cell leukaemia, proptosis, radiology

1 | CASE

A 61-year-old woman was referred because of bone pain and unilateral proptosis of the left eye (Figure 1A) with diplopia but without vision loss. Elevation, adduction and abduction were impaired. The plica was thickened and a distention was palpable above the eye. Fundoscopy showed no papilledema or vascular abnormalities. Computed tomography imaging showed a large intraorbital mass (Figure 1B) and extensive lytic skeletal lesions. Peripheral blood (PB) examination

showed hypercalcaemia, anaemia, thrombocytopenia and leukocytosis ($28.8 \times 10^9/L$). A more thorough examination of PB smears illustrated aberrant plasma cells with nucleoli (May–Grünwald–Giemsa stain, original magnification $\times 1000$; Figure 1C). Flow cytometry detected a clonal plasma cell population of 32%, positive for CD138 and CD38 and negative for CD56 and CD19. The bone marrow aspirate smear identified nearly complete replacement of normal haematopoietic cells by clonal plasma cells. Immunohistochemistry revealed CD56 negativity and CyclinD1 positivity. The serum-free light chain kappa level

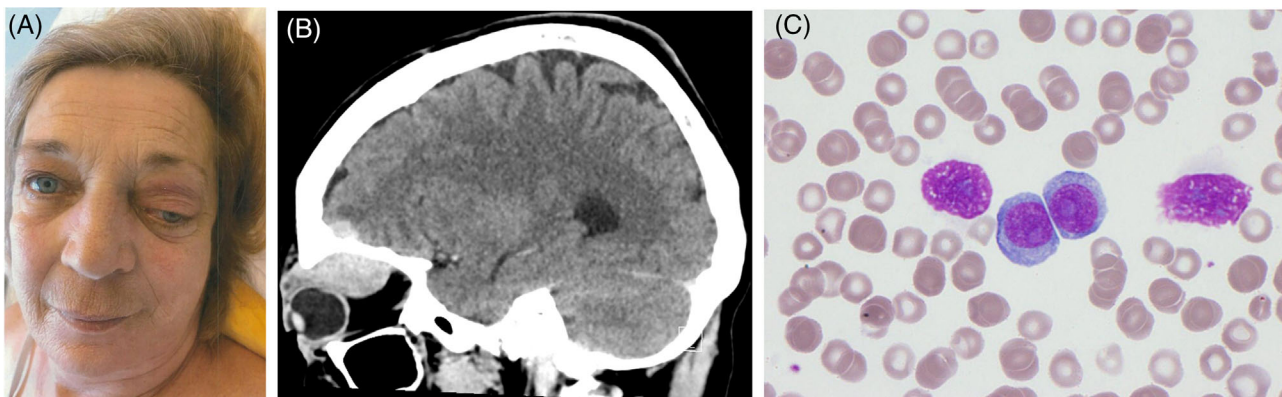


FIGURE 1 FIGURE 1 (A) Photograph showing proptosis of the left eye with inferior displacement. (B) Computed tomography (CT) imaging showing a large intraorbital mass extending intracranially with caudal and lateral displacement of the bulbus oculi with substantial proptosis. (C) Peripheral blood smear with May–Grünwald–Giemsa staining shows atypical plasma cells with a large nucleolus.

Abbreviation: pPCL, primary plasma cell leukaemia.

This is an open access article under the terms of the [Creative Commons Attribution-NonCommercial-NoDerivs](https://creativecommons.org/licenses/by-nc-nd/4.0/) License, which permits use and distribution in any medium, provided the original work is properly cited, the use is non-commercial and no modifications or adaptations are made.

© 2023 The Authors. *eJHaem* published by British Society for Haematology and John Wiley & Sons Ltd.

was 470 mg/L. Further cytogenetic exploration showed t(11;14) and deletion of 17p13. All findings were consistent with primary plasma cell leukaemia (pPCL). Induction therapy with daratumumab plus velcade (bortezomib)/lenalidomide/dexamethasone (D-VRD) resulted in complete remission, and full recovery of proptosis. However, there was early relapse after initial therapy, and the patient died because of severe sepsis.

Unilateral proptosis rarely manifests in haematologic malignancies and has not been reported as a presenting symptom in pPCL [1]. Increasing awareness of this phenomenon may aid clinicians in a fast diagnostic workup, including imaging and PB cytology and immunophenotyping.

Although the prognosis of patients with pPCL has significantly improved with daratumumab-based quadruplets, del17p(+) predicts poor outcome [2].

AUTHOR CONTRIBUTIONS

Irene T. Schrijver wrote the manuscript. All the authors revised the manuscript.

CONFLICT OF INTEREST STATEMENT

The authors declare they have no conflicts of interest.

ETHICS STATEMENT

The authors have confirmed ethical approval statement is not needed for this submission.

PATIENT CONSENT STATEMENT

Written informed consent was obtained from the patient.

CLINICAL TRIAL REGISTRATION

The authors have confirmed clinical trial registration is not needed for this submission.

ORCID

Yorick Sandberg  <https://orcid.org/0000-0002-4210-1331>

REFERENCES

1. Dsouza S, Kandula P, Kamath G, Kamath M. Clinical profile of unilateral proptosis in a tertiary care centre. *J Ophthalmol.* 2017;2017:8546458.
2. Katodritou E, Kastritis E, Dalampira D, Delimpasi S, Spanoudakis E, Labropoulou V, 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(5):730–38.

How to cite this article: Schrijver IT, Kalikmanov-Mikhaylovskaya E, Sandberg Y. Primary plasma cell leukaemia presenting as unilateral proptosis. *eJHaem.* 2023;4:1172–1173. <https://doi.org/10.1002/jha2.809>