Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Internal Medicine

Inaugural Ophthalmologic Involvement Revealing IgM Multiple Myeloma: A Case Report and Literature Review

Ikram Khoussar[1]*, Widad Rhandour[1], Rime Felk[1], Naoual Oubelkacem[1], Noufissa Alami[1], Zineb Khammar[1], Rizhlane Berrady[1]

[1]Department of Internal Medicine, Faculty of Medicine, Pharmacy and Dental Medicine of Fez, Université Sidi Mohamed Ben Abdellah, Fez, Morocco

DOI: https://doi.org/10.36347/sjmcr.2026.v14i02.033 | **Received:** 06.01.2026 | **Accepted:** 19.02.2026 | **Published:** 27.02.2026

***Corresponding author:** Ikram Khoussar
Assistant Professor, Department of Internal Medicine, Faculty of Medicine, Pharmacy and Dental Medicine of Fez, Université Sidi Mohamed Ben Abdellah, Fez, Morocco

Abstract | **Case Report**

***Introduction*:** IgM multiple myeloma is an extremely rare entity, accounting for less than 1% of all myelomas, and differs from Waldenström macroglobulinemia despite the shared secretion of monoclonal IgM. Its presentation with ophthalmologic involvement is exceptional and may threaten visual prognosis. ***Case Report*:** We report the case of a 38-year-old woman admitted for sudden bilateral visual acuity loss associated with headaches and constitutional symptoms. Ophthalmologic examination revealed bilateral retinal vein occlusion with papilledema and serous retinal detachment. Laboratory investigations showed profound aregenerative anemia, marked hyperproteinemia, hypercalcemia, and hyperviscosity syndrome. Serum protein electrophoresis demonstrated a massive monoclonal IgM spike. Bone marrow evaluation, including aspiration and biopsy, revealed 30% medullary plasmacytosis with CD138+ plasma cell infiltration, without diffuse lymphoplasmacytic proliferation. Testing for the MYD88 mutation, recommended to differentiate from Waldenström macroglobulinemia, could not be performed for financial reasons. Nevertheless, the overall findings supported a diagnosis of IgM multiple myeloma. The patient received chemotherapy according to the VCD protocol, with clinical, biological, and ophthalmologic improvement, followed by maintenance therapy with bortezomib and indication for autologous hematopoietic stem cell transplantation. ***Conclusion*:** Ophthalmologic involvement may reveal IgM multiple myeloma, highlighting the importance of systematic evaluation and prompt management to improve functional and overall prognosis.

Keywords: IgM Multiple Myeloma, Waldenström Macroglobulinemia, Ophthalmologic Involvement, Hyperviscosity.

INTRODUCTION

Multiple myeloma is a malignant hematologic disorder characterized by clonal proliferation of bone marrow plasma cells secreting a monoclonal immunoglobulin, most commonly IgG or IgA [1, 2]. IgM multiple myeloma is an exceptional entity, representing less than 1% of myelomas, and poses a major diagnostic challenge because of its clinical, biological, and immunological overlap with Waldenström macroglobulinemia [3, 4].

Ophthalmologic manifestations in monoclonal gammopathies are rare but potentially severe. They are most often secondary to hyperviscosity syndrome, leading to retinal microcirculatory abnormalities that may compromise visual prognosis [5, 6]. In this context, fundoscopic examination may provide a crucial indirect diagnostic clue. We report an unusual case of IgM multiple myeloma revealed by severe bilateral ophthalmologic involvement, illustrating the importance of early recognition of this complication.

CLINICAL OBSERVATION

A 38-year-old woman with no significant medical history was admitted for sudden bilateral visual acuity loss evolving over one week, associated with intense headaches. She also reported marked asthenia and exertional dyspnea for approximately three weeks.

Ophthalmologic examination revealed visual acuity limited to finger counting at 2 meters in both eyes. Fundoscopy showed dilated and tortuous retinal veins, multiple flame-shaped and blot intraretinal hemorrhages, bilateral stage III papilledema, and a poor macular reflex. Macular optical coherence tomography (OCT) demonstrated bilateral serous retinal detachment.

Laboratory investigations revealed profound macrocytic anemia (hemoglobin 4.9 g/dL), aregenerative, with rouleaux formation on peripheral smear. Total serum protein was elevated at 132 g/L, associated with corrected hypercalcemia of 120 mg/L. Serum protein electrophoresis showed a massive monoclonal spike in the gamma region. Quantitative immunoglobulin testing demonstrated markedly elevated IgM levels (128.48 g/L) with suppressed IgG and IgA, suggestive of hyperviscosity syndrome.

Brain imaging (CT and MRI) was unremarkable. Whole-body CT scan revealed bilateral osteolytic lesions of the iliac wings.

Bone marrow evaluation included aspiration and trephine biopsy. The myelogram showed 30% medullary plasmacytosis composed of morphologically mature plasma cells. The bone marrow biopsy revealed no diffuse lymphoplasmacytic proliferation. Immunohistochemical analysis demonstrated strong CD138 expression by plasma cells, without significant B-lymphoid marker expression.

Given the secretion of monoclonal IgM, testing for the MYD88 L265P mutation was recommended to exclude Waldenström macroglobulinemia; however, this analysis could not be performed due to financial constraints. Nevertheless, the overall clinical, biological, morphological, and radiological findings — including 30% medullary plasmacytosis, absence of lymphoplasmacytic infiltration, presence of lytic bone lesions, and plasma cell immunophenotype — supported the diagnosis of IgM multiple myeloma.

Urgent treatment was initiated with VCD chemotherapy (bortezomib, cyclophosphamide, dexamethasone). The patient showed rapid clinical improvement, progressive normalization of biological parameters, and ophthalmologic improvement with regression of papilledema and partial recovery of visual acuity. Maintenance therapy with bortezomib was started, with indication for autologous hematopoietic stem cell transplantation.

DISCUSSION

IgM multiple myeloma is an extremely rare entity, accounting for less than 1% of myelomas, and differs from Waldenström macroglobulinemia despite the shared secretion of monoclonal IgM [3, 4]. In our case, the patient presented with sudden bilateral visual loss due to severe ophthalmologic involvement secondary to hyperviscosity syndrome, an unusual but potentially serious mode of presentation [5-8]. This type of initial manifestation highlights the importance of systematic ophthalmologic evaluation in any rapid visual acuity decline associated with hyperproteinemia, as it may lead to early diagnosis of an underlying hematologic malignancy.

The differential diagnosis with Waldenström macroglobulinemia is particularly challenging, especially when the MYD88 L265P mutation is not investigated. In our case, this test could not be performed for financial reasons, which could theoretically complicate the distinction [3-8]. However, several morphological and biological arguments supported the diagnosis of IgM myeloma. The 30% medullary plasmacytosis observed on myelogram, absence of lymphoplasmacytic proliferation on bone marrow biopsy, and strong CD138 expression by plasma cells favor a pure plasma cell proliferation, in contrast to Waldenström disease, which typically shows mixed lymphoplasmacytic infiltration [2-7]. Furthermore, the presence of lytic bone lesions and hypercalcemia, characteristic of multiple myeloma, reinforced this diagnosis [1-4].

The ophthalmologic findings in our patient — retinal venous stasis, intraretinal hemorrhages, papilledema, and bilateral serous detachment — clearly illustrate hyperviscosity syndrome related to markedly elevated IgM levels [5,6,8]. Although rare, these severe abnormalities may constitute the initial presentation of IgM myeloma. Early recognition is crucial to prevent irreversible visual sequelae and to initiate appropriate hematologic treatment, potentially including plasmapheresis in severe cases [1-8].

Management with combination chemotherapy using the VCD protocol (bortezomib, cyclophosphamide, dexamethasone) led to rapid improvement in general condition, progressive normalization of laboratory parameters, and partial visual recovery. Maintenance therapy with bortezomib was initiated, with indication for autologous stem cell transplantation to consolidate response. This favorable evolution underscores the importance of a multidisciplinary approach and early recognition of ophthalmologic manifestations to improve both functional and overall prognosis [1-8].

In summary, this case illustrates the rarity of IgM multiple myeloma and the diagnostic difficulty in distinguishing it from Waldenström macroglobulinemia, particularly in the absence of MYD88 mutation testing. It also highlights the possibility of severe ophthalmologic involvement as the initial presentation, emphasizing the need for clinical vigilance and rapid diagnosis to initiate appropriate management and limit complications.

CONCLUSION

Ophthalmologic involvement may represent a rare but severe presenting feature of IgM multiple myeloma. In cases of bilateral retinal vein occlusion associated with profound anemia and hyperproteinemia, an underlying hematologic malignancy should be systematically considered.

REFERENCES

1. Rajkumar SV. Multiple myeloma: 2023 update on diagnosis, risk-stratification, and management. *Am J Hematol*. 2023;98:123–145.
2. Kyle RA, Rajkumar SV. Plasma cell disorders. *Hematology Am Soc Hematol Educ Program*. 2022;2022:1–11.
3. Kastritis E, et al. IgM multiple myeloma: biology, diagnosis and treatment. *Leukemia*. 2022;36:1–10.
4. Dimopoulos MA, et al. Waldenström's macroglobulinemia and IgM-related disorders. *N Engl J Med*. 2023;388:640–654.
5. Vignal-Clermont C, et al. Ocular manifestations of plasma cell dyscrasias. *Surv Ophthalmol*. 2021;66:234–245.
6. Stone MJ, et al. Hyperviscosity syndrome in plasma cell dyscrasias. *Br J Haematol*. 2021;195:344–356.
7. WHO Classification of Tumours Editorial Board. *Haematolymphoid tumours*. 5th ed. IARC; 2022.
8. Castillo JJ, et al. Diagnosis and management of Waldenström macroglobulinemia. *Blood*. 2022;139:285–296.