

Orbital Plasmacytoma in an Elderly Patient With Multiple Myeloma

Salma Aloui¹, Pir Abdul Ahad Aziz Qureshi², Vikram Rao Bollineni¹

Review began 01/24/2024

Review ended 02/05/2024

Published 02/08/2024

© Copyright 2024

Aloui et al. This is an open access article distributed under the terms of the Creative Commons Attribution License CC-BY 4.0., which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

1. Department of Radiology, Universitair Ziekenhuis Brussel, Brussels, BEL 2. Department of Radiology, Landspítali - The National University Hospital of Iceland, Reykjavík, ISL

Corresponding author: Pir Abdul Ahad Aziz Qureshi, abdulahad.q@gmail.com

Abstract

Orbital plasmacytomas are exceedingly rare neoplasms characterized by the proliferation of monoclonal plasma cells in the orbital soft tissues. Their presentation and clinical course can be diverse, making early diagnosis and management challenging. This case report sheds light on one such instance, emphasizing the diagnostic and therapeutic aspects of this uncommon condition. We present an 81-year-old patient with a prior diagnosis of multiple myeloma and bilateral orbital plasmacytomas, highlighting the importance of imaging in diagnosis and management.

Categories: Ophthalmology, Radiology, Oncology

Keywords: secondary malignancies, mri, ct, radiological features, multiple myeloma, orbital plasmacytoma

Introduction

Plasmacytoma, a pathological proliferation of plasma cells, represents a subtype of plasma cell dyscrasia, which can present itself as multiple myeloma, primary amyloidosis, or monoclonal gammopathy of undetermined significance (MGUS). Plasmacytoma can be primary or secondary to disseminated multiple myeloma, categorizing it into two distinct groups, i.e., medullary (about 2-5% of all plasma cell neoplasms) and extramedullary (about 3% of all plasma cell neoplasms). Extramedullary plasmacytoma usually manifests in the upper respiratory, head, and neck regions [1].

Case Presentation

An 81-year-old female patient who was diagnosed with multiple myeloma in 2019 presented with a three-month history of double vision, bulging of the right eye globe, restricted eye movement, and headache. On general physical examination, she had a high blood pressure of 160/95 mm Hg, and her pulse rate was 80 b/min. An ophthalmological examination revealed bilateral diplopia in all quadrants except inferiorly, mild proptosis of the right eye, and pupils that were normal bilaterally. Subsequently, a CT scan of the brain was advised to evaluate the patient's symptoms, which revealed well-defined bilateral intra-orbital tumors with homogenous contrast enhancement. No overlying bone erosion or destruction was identified. The largest mass was in the right retrobulbar region, measuring 26 mm (Figure 1).

How to cite this article

Aloui S, Qureshi P, Bollineni V (February 08, 2024) Orbital Plasmacytoma in an Elderly Patient With Multiple Myeloma. Cureus 16(2): e53823. DOI 10.7759/cureus.53823

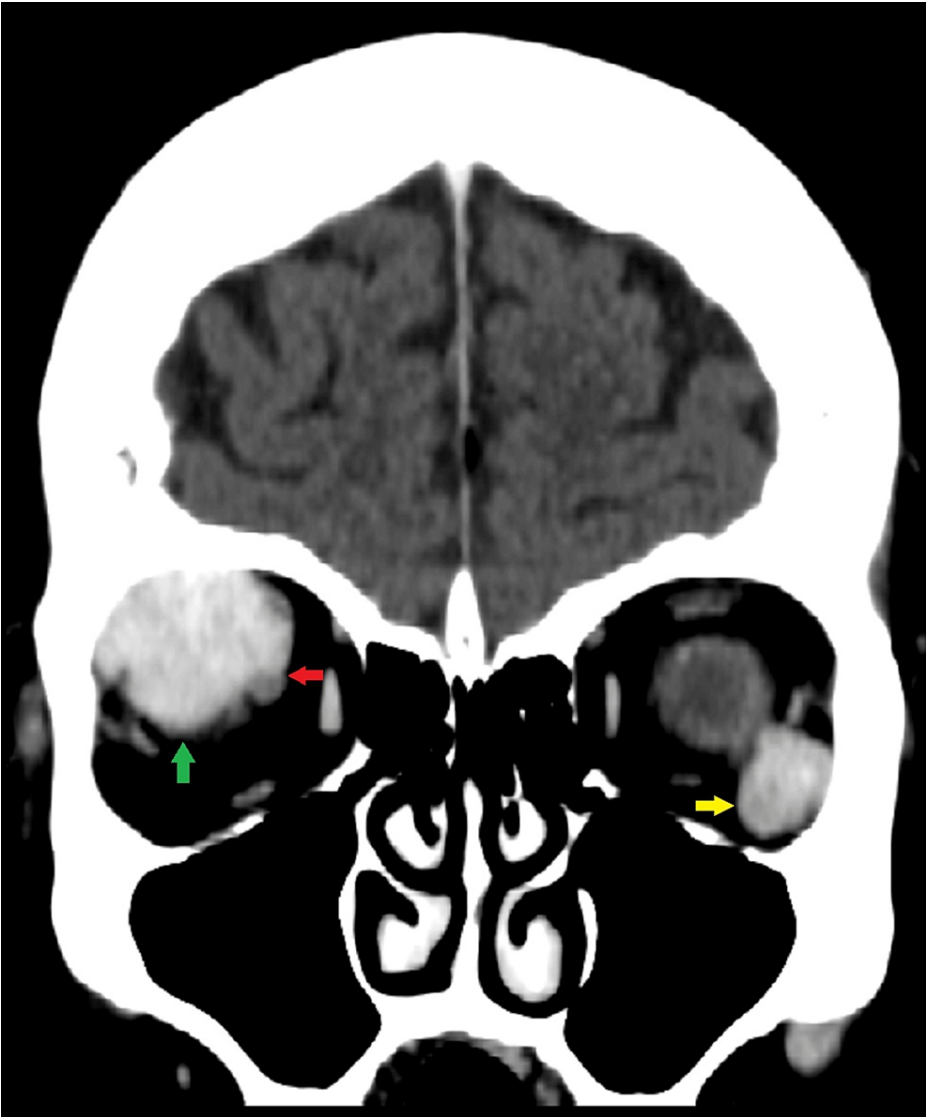


FIGURE 1: CT brain scan with contrast (coronal view)

Well-defined, bilateral intra-orbital tumors with homogenous contrast enhancement are seen (green and yellow arrows). The lesion in the right orbit is encasing and compressing the right optic nerve (red arrow).

Additionally, there were multiple scattered osteolytic lesions in the skull, spine, sternum, and probably in the ribs, pelvis, and scapula on the CT scan. An MRI was performed to better evaluate the lesions, their extent, and their relationship with surrounding structures. It identified three orbital well-circumscribed, homogeneously enhancing, diffusion-restrictive soft tissue lesions, with the largest lesion in the right eye measuring 4 cm, encasing the optic nerve with focal loss of intervening fat planes and exerting a mass effect on the conus (Figure 2).



FIGURE 2: T1 post-contrast brain MRI image (coronal view)

Uniformly enhancing, bilateral intra-orbital lesions are seen (red and yellow arrows). The lesions are closely abutting the adjacent extraocular muscles. Note the larger lesion in the right orbit encasing the optic nerve and compressing and displacing it inferiorly.

Based on the clinical history and imaging findings, the differentials of orbital plasmacytoma, lymphoma, and metastases were advised. Subsequently, the patient underwent excision of the lesions, and histopathology of the specimen revealed plasma cell infiltration in soft tissue where the cells are partly atypical and many binucleated plasma cells are seen. Appearance was compatible with plasmacytoma.

The patient was then further treated with chemotherapy and radiotherapy. Currently, the patient is being treated with chemotherapy and receiving bortezomib once a month and lenalidomide 10 mg for seven days every other week, which has kept the disease in remission clinically. No further imaging was requested by the treating physician at this point, and the patient is advised for monthly follow-up.

Discussion

Radiological assessment of orbital plasmacytomas often involves CT and MRI scans. Orbital plasmacytomas are typically localized within the orbital soft tissues, and imaging can visualize the tumor's location within the orbit. On CT and MRI scans, orbital plasmacytomas often appear as well-defined, enhancing soft tissue masses within the orbital space. These masses may be homogeneous in appearance.

When administering contrast, orbital plasmacytomas typically exhibit enhancement, signifying increased vascularity within the lesion. One distinctive characteristic of orbital plasmacytomas is their capacity to

induce bone destruction. Imaging reveals this as erosive changes in the adjacent orbital bones, representing an essential distinguishing factor from other orbital tumors [2-4].

In some cases, these tumors may infiltrate the surrounding orbital soft tissues, leading to displacement or compression of the adjacent structures, which is evident in imaging. Imaging plays a crucial role in monitoring the response to treatment. Following treatment (e.g., surgery and/or radiotherapy), serial imaging may show a reduction in the lesion size and a decrease in enhancement, indicating a positive response to the treatment [5].

In clinical practice, radiological assessment of orbital plasmacytomas plays a valuable role in initial evaluation but has inherent limitations. These limitations include the non-definitive nature of imaging findings, their potential lack of specificity that can lead to misdiagnosis, and the challenge of distinguishing between tumor recurrence and post-treatment effects over time. Furthermore, it is crucial to consider a range of differential diagnoses, including metastasis, vascular malformations, lymphoma, inflammatory conditions, and benign lesions. These conditions may share radiological characteristics with plasmacytomas, emphasizing the need for a multidisciplinary approach and histopathological confirmation to ensure precise diagnosis and appropriate management.

Conclusions

The coexistence of orbital plasmacytomas with multiple myeloma poses a diagnostic challenge and demands a multidisciplinary approach. Characterizing the extent of the orbital lesion and guiding treatment strategies through radiological assessment becomes crucial. In this case, focal radiation therapy effectively treated the patient, resulting in complete remission. The radiological characteristics of orbital plasmacytomas can vary from case to case. However, the presence of a well-defined, enhancing mass with bone destruction and soft tissue infiltration often serves as an indicative feature of this rare orbital neoplasm. Radiological evaluation is critical for initial diagnosis, surgical planning, and post-treatment monitoring of orbital plasmacytomas.

Additional Information

Author Contributions

All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work.

Concept and design: Pir Abdul Ahad Aziz Qureshi, Vikram Rao Bollineni, Salma Aloui

Critical review of the manuscript for important intellectual content: Pir Abdul Ahad Aziz Qureshi, Vikram Rao Bollineni

Supervision: Vikram Rao Bollineni

Acquisition, analysis, or interpretation of data: Salma Aloui

Drafting of the manuscript: Salma Aloui

Disclosures

Human subjects: Consent was obtained or waived by all participants in this study. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

References

1. Kim YJ, Moon SW, Park IK, Shin JH: Extradural plasmacytoma of the orbit with intracranial invasion: a case report. *Medicine (Baltimore)*. 2023, 102: [10.1097/MD.00000000000033920](https://doi.org/10.1097/MD.00000000000033920)
2. Ooi GC, Chim JC, Au WY, Khong PL: Radiologic manifestations of primary solitary extradural and multiple solitary plasmacytomas. *AJR Am J Roentgenol*. 2006, 186:821-7. [10.2214/AJR.04.1787](https://doi.org/10.2214/AJR.04.1787)
3. Ahn S, Cha SH, Cho WH: Spheno-orbital plasmacytoma as an initial presentation of multiple myeloma. *Brain Tumor Res Treat*. 2022, 10:270-4. [10.14791/btrt.2022.0026](https://doi.org/10.14791/btrt.2022.0026)
4. Permaisuari N, Anggraini N, Mahyuddin M, Kodrat E, Rajabto W: A case series of aggressive orbital plasmacytomas. *Ocul Oncol Pathol*. 2023, 8:197-202. [10.1159/000527273](https://doi.org/10.1159/000527273)
5. Soutar R, Lucraft H, Jackson G, Reece A, Bird J, Low E, Samson D: Guidelines on the diagnosis and management of solitary plasmacytoma of bone and solitary extradural plasmacytoma. *Clin Oncol (R Coll Radiol)*. 2004, 16:405-13. [10.1016/j.clon.2004.02.007](https://doi.org/10.1016/j.clon.2004.02.007)