World Journal of Advanced Research and Reviews
eISSN: 2581-9615 CODEN (USA): WJARAI
Cross Ref DOI: 10.30574/wjarr
Journal homepage: https://wjarr.com/

(CASE REPORT)

ORBITAL PLASMACYTOMA REVEALING MULTIPLE MYELOMA: CASE REPORT

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World Journal of Advanced Research and Reviews, 2026, 31(03), 474–480

Publication history: Received on 31 July 2026; revised on 07 September 2026; accepted on 09 September 2026

Article DOI: https://doi.org/10.30574/wjarr.2026.31.3.2313

ABSTRACT

Multiple myeloma (MM) is a malignant hematologic disorder in which orbital localization is exceptional (< 1%). We report the case of an orbital mass revealing multiple myeloma with an atypical radiological semiology. Contrary to classic descriptions, computed tomography (CT) showed integrity of the bone walls without bone lysis, thus simulating a lymphoproliferative process. However, magnetic resonance imaging (MRI) demonstrated a soft tissue mass with frank hyperintensity on diffusion-weighted imaging (DWI), reflecting tumor hypercellularity. The diagnosis was confirmed by histopathological and immunohistochemical examination showing plasmacytic infiltration with kappa monotypia, consistent with systemic multiple myeloma after biological workup. This case emphasizes that the absence of bone lysis does not exclude a plasmacytoma and recalls the importance of systemic evaluation in the presence of any solid orbital mass.

Keywords: Multiple myeloma; Orbital plasmacytoma; Magnetic resonance imaging; Extramedullary disease

1 INTRODUCTION

Multiple myeloma is a malignant hematologic disorder characterized by clonal proliferation of plasma cells within the bone marrow. It accounts for approximately 1% of all malignant tumors and nearly 10% of hematologic malignancies. The disease classically manifests with diffuse bone involvement responsible for lytic lesions, found in more than 80% of patients at the time of diagnosis (1).

Extramedullary manifestations of multiple myeloma are relatively rare and result from the dissemination of tumoral plasma cells outside the bone marrow. They can affect various organs, notably soft tissues, lymph nodes, liver, skin, or the central nervous system. Among these localizations, orbital involvement remains exceptional, representing less than 1% of cases according to published series (2).

Clinically and radiologically, these orbital localizations can mimic a wide range of tumoral or inflammatory pathologies, such as orbital lymphomas, inflammatory pseudotumors, or certain metastases, which sometimes makes the diagnosis difficult. In this context, imaging plays a key role in diagnostic orientation, allowing evaluation of the local extension of the lesion and guiding the diagnostic and therapeutic strategy.

We report here a rare case of orbital localization of multiple myeloma with an atypical radiological presentation characterized by the absence of orbital bone lysis, which may simulate a lymphoproliferative process.

2 CASE REPORT

We report the case of a 58-year-old patient who consulted at the ophthalmic emergency department of Hassan II University Hospital in Fez for exophthalmos with decreased visual acuity in the right eye evolving for 15 days.

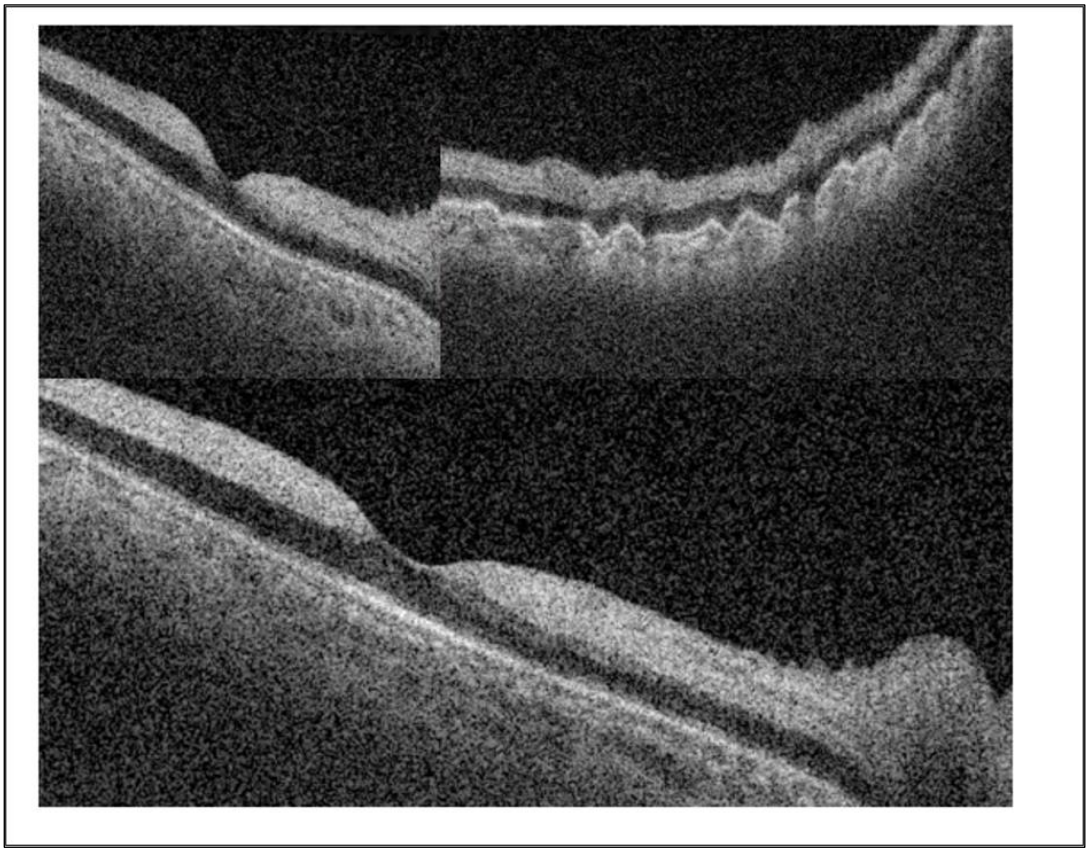

Figure 1: Hhyperreflectivity of the ganglion cell, plexiform, and inner nuclear layers, presence of chorioretinal folds and papillary edema.

Clinical examination of the right eye revealed positive light perception in the inferotemporal quadrant, non-axial irreducible non-pulsatile exophthalmos of 24 mm, limitation of ocular motility, significant inferior chemosis, and conjunctival hyperemia. Fundus examination showed papillary edema, tortuous vessels with ischemic retina, and chorioretinal folds limiting a nasal chorioretinal bulge from 1:30 to 4:00.

Systemic examination found multiple cutaneous and subcutaneous masses in the thorax and abdomen.

On optical coherence tomography (OCT): hyperreflectivity of the ganglion cell, plexiform, and inner nuclear layers, presence of chorioretinal folds and papillary edema (Figure 1).

Cranio-orbital CT showed an intraorbital and intraconal soft tissue process on the right, centered on the superior oblique muscle, oval-shaped, well-circumscribed, heterogeneously enhanced after contrast injection, measuring 31 × 22 × 25 mm in diameters (AP × T × H).

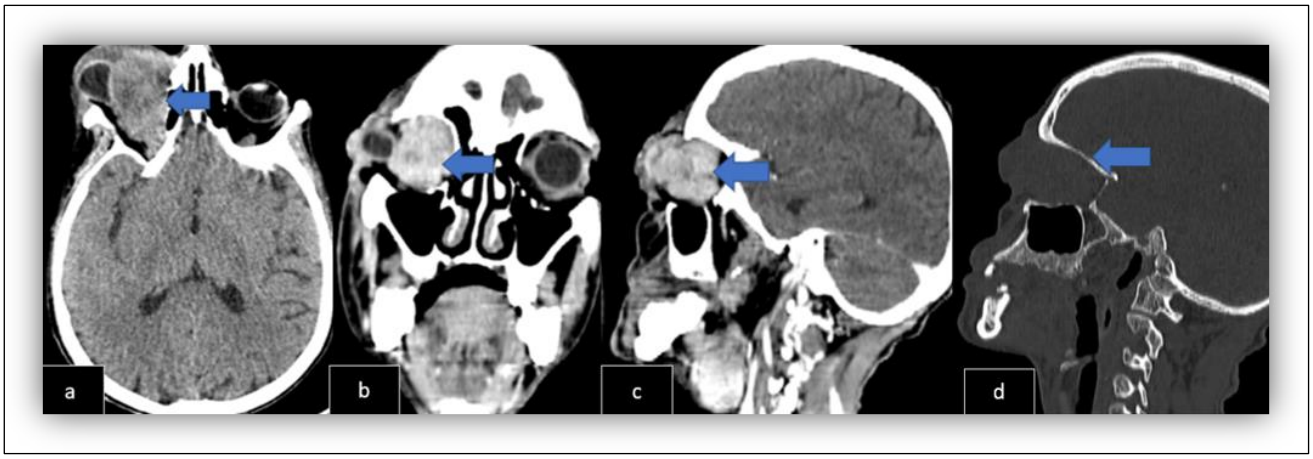

Figure 2: Cranio-orbital CT shows an intraorbital and intraconal soft tissue process on the right centered on the superior oblique muscle, oval-shaped, well-circumscribed, heterogeneously enhanced after contrast injection, measuring 31 × 22 × 25 mm in diameters (AP × T × H). This process is in contact with the orbital roof without signs of bone lysis in front of it (image d), remains distant from the orbital floor (images b and c), displaces and stretches the optic nerve, is in contact with the ipsilateral lamina papyracea without signs of bone lysis, and displaces the globe with loss of the separating fat plane in places, responsible for grade III exophthalmos (image a).

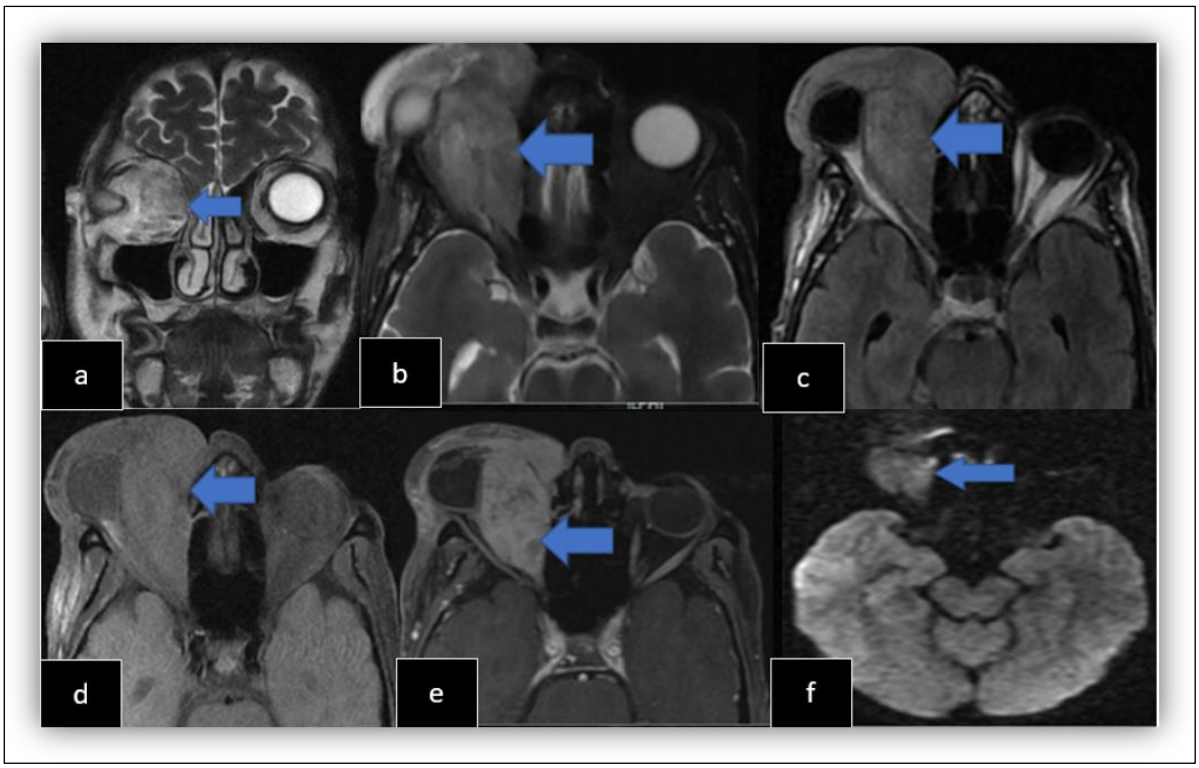

Figure 3: Cranio-orbital MRI shows an intraorbital and intraconal soft tissue process on the right centered on the superior oblique muscle, oval-shaped, well-circumscribed, described as heterogeneously hyperintense on T2 (arrows a, b and c), discretely heterogeneously hyperintense on T1 (arrow d), with restricted diffusion (arrow f), heterogeneously enhanced after contrast injection (arrow e), measuring 62 × 37 × 33 mm in diameters (AP × T × H).

Cranio-orbital MRI: intraorbital and intraconal soft tissue process on the right centered on the superior oblique muscle, oval-shaped, well-circumscribed, described as heterogeneously hyperintense on T2, discretely heterogeneously hyperintense on T1, hyperintense on DWI, heterogeneously enhanced after contrast injection, measuring 62 × 37 × 33 mm in diameters (AP × T × H).

Staging cervico-thoraco-abdomino-pelvic CT: peritoneal, thoracic and abdominal wall secondary localizations. Multiple axial bone lesions of the pelvic and scapular girdle consistent with multiple myeloma.

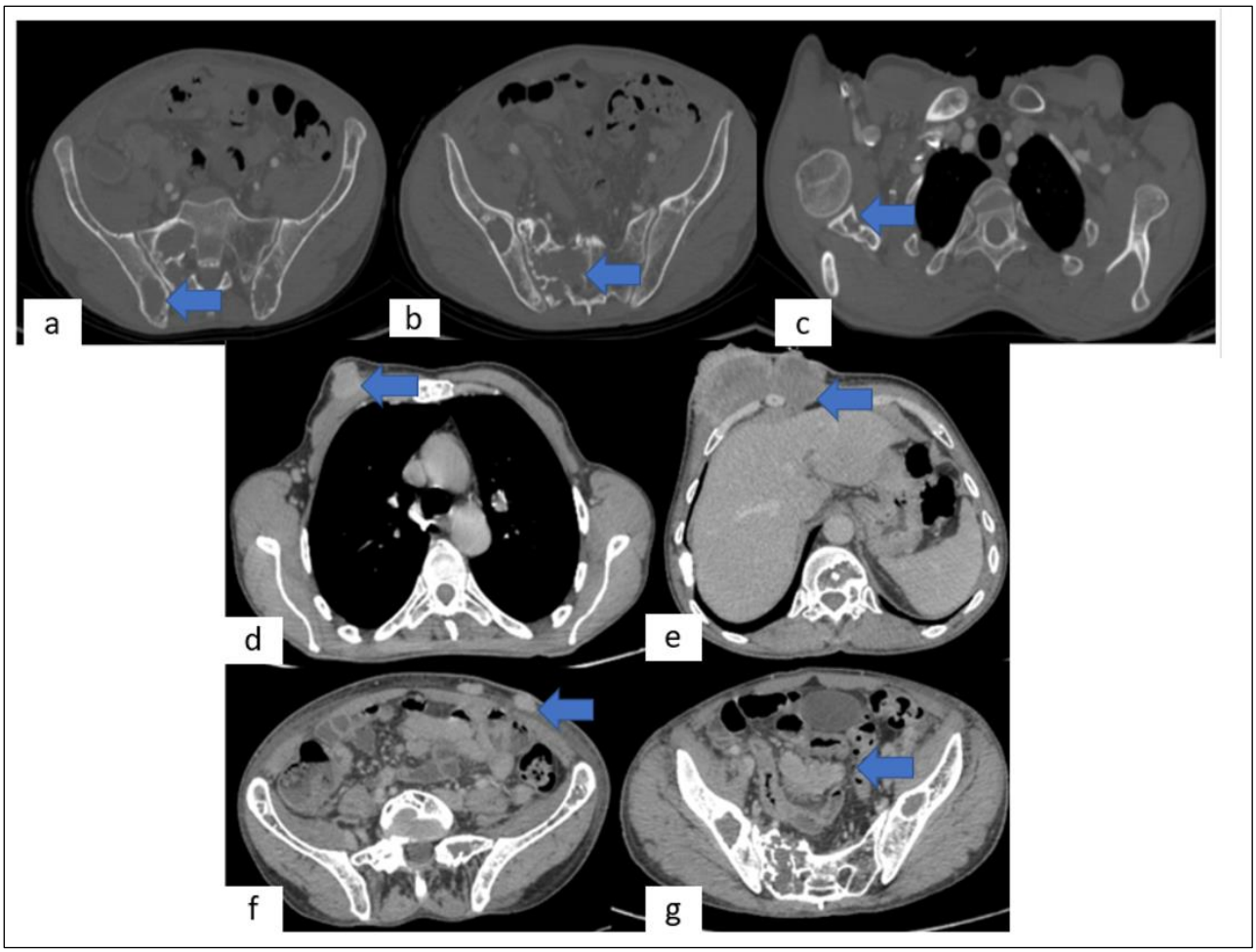

Figure 4: Thoraco-abdomino-pelvic CT shows the presence of masses and nodules in the parietal region in front of the right pectoral muscle and left axillary region, oval-shaped, well-circumscribed, heterogeneously enhanced after contrast injection (images d and e), multiple intra- and retroperitoneal soft tissue masses and nodules, oval-shaped, confluent in some cases, heterogeneously enhanced after contrast injection (image g), associated with multiple anterior abdominal wall soft tissue masses and nodules, oval-shaped, confluent in some cases, heterogeneously enhanced after contrast injection (images d and f). There are also multiple enhancing bone lesions involving the sternum, scapular girdle, ribs, and pelvic girdle, the largest of which involves the right sacral ala, invading the ipsilateral dorsal sacral foramina (images a, b and c), consistent with peritoneal, thoracic and abdominal wall secondary localizations and multiple axial bone lesions of the pelvic and scapular girdle in relation to multiple myeloma.

Histopathological study of the biopsy of an abdominal wall mass revealed histological and immunohistochemical features consistent with plasmacytoma with kappa monotypia.

The patient received 6 cycles of VDT+Z chemotherapy associated with palliative analgesic radiotherapy for intense bone pain.

Control cranial CT showed good clinical evolution with near-complete regression of the right intraorbital and intraconal soft tissue process centered on the superonasal quadrant, previously described on the old imaging, replaced by hypodense intraconal infiltration at this level.

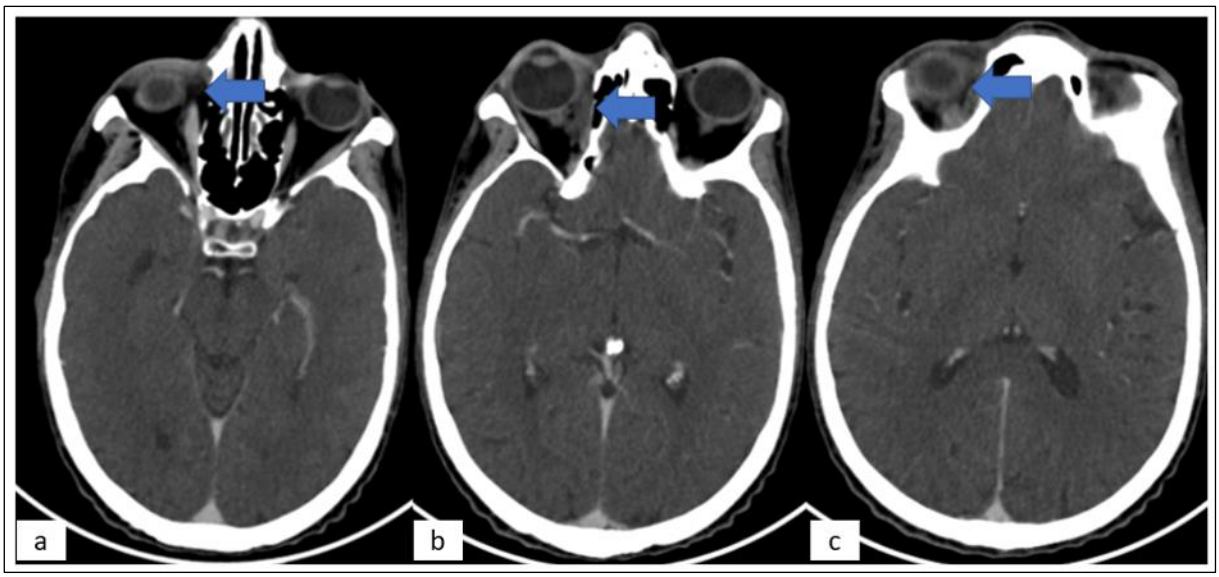

Figure 5: Control CT shows near-complete regression of the right intraorbital and intraconal soft tissue process centered on the superonasal quadrant, replaced by hypodense intraconal infiltration at this level (arrows a, b and c).

3 DISCUSSION

Multiple myeloma is a malignant hematologic disorder characterized by clonal proliferation of plasma cells within the bone marrow. Extramedullary localizations represent a relatively rare but well-described manifestation of the disease, observed in approximately 7 to 18% of patients during the course of the disease (3). Among these, orbital involvement remains exceptional, representing less than 1% of extramedullary localizations reported in the literature (2). It may manifest either as a solitary orbital plasmacytoma (4) or in the context of known or revealing systemic multiple myeloma (5).

Pathophysiologically, extramedullary involvement in multiple myeloma is linked to a phenomenon of clonal escape of malignant plasma cells (6). These cells lose the expression of certain adhesion molecules essential for retention in the bone marrow niche, notably CD56, VLA-4 and LFA-1, which favors their hematogenous dissemination to peripheral tissues (7). This dissemination is also facilitated by alterations in the bone marrow microenvironment and by overexpression of metalloproteinases favoring tissue invasion. In the orbit, propagation may occur by direct extension from adjacent bones, notably the diploë of the skull bones and the lateral orbital wall, or by hematogenous dissemination to orbital soft tissues (2).

Clinically, orbital involvement most often manifests as progressive exophthalmos, sometimes associated with diplopia, decreased visual acuity, orbital pain or palpebral edema (4). In certain cases, as in our patient, compression of intraorbital structures may lead to compressive optic neuropathy and fundus abnormalities such as papillary edema or chorioretinal folds (5).

Imaging plays a fundamental role in diagnostic orientation. Computed tomography (CT) classically reveals a soft tissue mass associated with typical lytic bone lesions of multiple myeloma, described as “punched-out” lacunae (2). However, our observation illustrates an atypical presentation characterized by the absence of orbital bone lysis, constituting a real diagnostic trap. This integrity of the orbital walls may initially orient toward other etiologies such as orbital lymphomas or inflammatory pathologies (8).

Magnetic resonance imaging (MRI) provides essential information in this context, notably for the analysis of soft tissues and intraorbital extension (2). Orbital plasmacytomas generally appear iso- or slightly hypointense on T1 and iso- to hyperintense on T2, with marked enhancement after gadolinium injection (9). The diffusion sequence constitutes a particularly useful tool (10). High tumor cellularity leads to diffusion restriction with a low ADC coefficient, reflecting the high cellular density and high nucleocytoplasmic ratio of tumoral plasma cells (10). This characteristic helps orient toward a malignant lesion and distinguish plasmacytomas from certain inflammatory or benign lesions (10).

The main differential diagnosis remains orbital lymphoma, which represents the most frequent primary malignant tumor of the orbit in adults. Unlike myeloma, lymphoma tends to mold the orbital structures without causing bone destruction and also presents diffusion restriction on MRI, which may complicate radiological distinction (8). Other

diagnoses must also be considered, notably orbital metastases from carcinomas (breast, lung, prostate), idiopathic inflammatory pseudotumors, or systemic inflammatory diseases such as IgG4-related disease (2).

Diagnostic confirmation relies imperatively on histopathological and immunohistochemical examination, demonstrating a monoclonal proliferation of plasma cells expressing plasmacytic markers such as CD138 and CD38, with kappa or lambda light-chain restriction (11). In our case, the presence of kappa monotypia associated with multiple localizations was compatible with extramedullary involvement of a known multiple myeloma.

From a prognostic point of view, extramedullary localizations of multiple myeloma are generally associated with more aggressive disease and a less favorable prognosis, often reflecting partial resistance to the bone marrow microenvironment and conventional treatments (6). Their presence is frequently correlated with high tumor burden and advanced stages of the disease according to the Durie-Salmon classification or the International Staging System (3).

Therapeutic management relies on a multidisciplinary approach, combining systemic treatment of multiple myeloma and local treatment of orbital lesions (12). Current protocols generally include combinations of chemotherapy containing proteasome inhibitors, such as bortezomib, often combined with immunomodulators, monoclonal antibodies, and corticosteroids (12). Orbital radiotherapy constitutes an effective treatment for local tumor control, notably in compressive or symptomatic forms, allowing rapid reduction of tumor mass and improvement of visual symptoms (13). In our observation, the near-complete regression of the orbital mass after chemotherapy illustrates the good sensitivity of plasmacytomas to systemic treatment.

Thus, this observation underlines the importance of including multiple myeloma in the differential diagnosis of any solid orbital mass, even in the absence of scanographic bone lysis. The association of clinical, radiological, and histological data remains indispensable for establishing a precise diagnosis and guiding therapeutic management.

4 CONCLUSION

Orbital localization of multiple myeloma can be misleading due to its scanographic bone integrity. Diffusion MRI is the key element to suspect malignancy. However, only histological proof with immunohistochemistry (kappa monotypia) allows confirmation of the plasmacytoma, then imposing a complete CRAB workup for therapeutic management.

STATEMENTS ON COMPLIANCE WITH ETHICAL STANDARDS

Acknowledgements

The authors would like to express their sincere gratitude to all those who contributed to this work. Their support and valuable advice were essential to the completion of this study.

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of ethical approval

This case report was conducted in accordance with institutional ethical standards.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

REFERENCES

[1] Kyle RA, Rajkumar SV. Multiple myeloma. Blood. 2008;111(6):2962-72. doi: 10.1182/blood-2007-10-078022. PMID: 18332230.

[2] Burkat CN, Van Buren JJ, Lucarelli MJ. Characteristics of orbital multiple myeloma: a case report and literature review. Surv Ophthalmol. 2009;54(6):697-704. doi: 10.1016/j.survophthal.2009.04.012. PMID: 19709708.

[3] Bladé J, Beksac M, Caers J, et al. Extramedullary disease in multiple myeloma: a systematic literature review. Blood Cancer J. 2022;12(3):45. doi: 10.1038/s41408-022-00643-3. PMID: 35314675.

[4] Rodman HI, Font RL. Orbital involvement in multiple myeloma. Review of the literature and report of three cases. Arch Ophthalmol. 1972;87(1):30-5. doi: 10.1001/archopht.1972.01000020032006. PMID: 4550269.

[5] Panda BB, Kar A, Moharana B, Mishra P. Orbital involvement in long-standing multiple myeloma. BMJ Case Rep. 2023;16(5):e255585. doi: 10.1136/bcr-2023-255585. PMID: 37160376; PMCID: PMC10174002.

[6] Usmani SZ, et al. Extramedullary disease portends poor prognosis in multiple myeloma and is over-represented in high-risk disease even in the era of novel agents. Haematologica. 2012;97(11):1761-7. PMID: 22665544.

[7] Gupta S, et al. Extramedullary Multiple Myeloma: A Patient-Focused Review. World J Oncol. 2022;13(5):258-266. doi: 10.14740/wjon1521. PMCID: PMC9635794.

[8] Haradome K, et al. Orbital Lymphoproliferative Disorders: Value of MR Imaging for Differentiating Lymphoma from Benign OPLDs. AJNR Am J Neuroradiol. 2014;35(10):1976-82.

[9] Aloui S, et al. Orbital Plasmacytoma in an Elderly Patient With Multiple Myeloma. Cureus. 2024;16(2):e538xx. PMCID: PMC10924626.

[10] Rodríguez-Laval V, et al. Imaging of Multiple Myeloma: Present and Future. J Clin Med. 2024;13(1):264. doi: 10.3390/jcm13010264.

[11] Slama N, et al. Extramedullary plasmacytoma of the orbit complicating the evolution of multiple myeloma in complete remission. Clin Case Rep. 2024. doi : 10.1002/ccr3

[12] Bansal R, et al. Extramedullary disease in multiple myeloma. Blood Cancer J. 2021;11(9):161. doi: 10.1038/s41408-021-00527-y. PMID: 34588423.

[13] Thoumazet F, et al. Orbital and muscle involvement in multiple myeloma. Can J Ophthalmol. 2006;41(6):733-6.