Open Access Original Research Article

MedPeer Publisher
Abbreviated Key Title: MedPeer
ISSN : 3066-2737
homepage: https://www.medpeerpublishers.com

Aggressive endonasal extramedullary plasmacytoma revealing multiple myeloma: a multidisciplinary challenge

DOI: 10.70780/medpeer.000QGTY

AUTHORS AND AFFILIATION

El Mehdi Harbili[1], Jalal Oubenjah[1], Mahdi Laarabi[1], Ahmed Rouihi[1], Mouad Moujoud[1], Mohamed Zalagh[1], Saloua Ouraini[1], Bouchaib Hemmaoui[1], Fouad Benariba[1], Noureddine Errami[1]

[1]Department of Otorhinolaryngology and Head and Neck Surgery,
Mohammed V Military Teaching Hospital, Rabat, Morocco.

Corresponding author : El Mehdi Harbili .

ABSTRACT

Extramedullary plasmacytoma is a rare plasma cell neoplasm, most frequently involving the upper aerodigestive tract. Endonasal localization is uncommon and may represent either a solitary lesion or the first manifestation of multiple myeloma. Clinical presentation is often nonspecific, which may lead to diagnostic delay. We report the case of an 83-year-old man presenting with chronic nasal obstruction and recurrent epistaxis. Endoscopic examination revealed a friable submucosal mass occupying both nasal cavities with extension to the hard palate. Imaging demonstrated an aggressive locally advanced endonasal lesion with skull base extension. Histopathological and immunohistochemical analysis confirmed a lambda light-chain plasmacytoma. Subsequent hematological investigations established the diagnosis of multiple myeloma. The patient received systemic therapy according to current hematological protocols. This case highlights the importance of considering plasmacytoma in the differential diagnosis of destructive endonasal masses and underscores the need for systematic hematological assessment.

KEYWORDS :

Plasmacytoma; Multiple Myeloma; Nose Neoplasms; Nasal Cavity; Skull Base; Palate, Hard; Immunoglobulin Lambda-Chains; Diagnosis, Differential.

MAIN ARTICLE

INTRODUCTION:

Plasma cell neoplasms encompass a spectrum of disorders ranging from monoclonal gammopathy of undetermined significance to multiple myeloma and extramedullary plasmacytoma. Extramedullary plasmacytoma represents less than 5% of plasma cell tumors and predominantly arises in the upper aerodigestive tract, particularly the nasal cavities, paranasal sinuses, and nasopharynx [1,2]. Endonasal involvement is rare but clinically significant because of its locally aggressive behavior and proximity to critical anatomical structures.

Extramedullary plasmacytoma may occur as a solitary lesion or as part of systemic multiple myeloma. Differentiating these entities is essential, as treatment strategies and prognosis differ considerably [3]. Several recent studies report progression to multiple myeloma in up to 30–40% of cases during follow-up [4]. We report a case of aggressive endonasal extramedullary plasmacytoma revealing multiple myeloma, emphasizing diagnostic challenges and therapeutic implications.

CASE PRESENTATION:

An 83-year-old man, retired from the Moroccan Armed Forces, was admitted for evaluation of chronic nasal obstruction associated with recurrent mild epistaxis evolving over six months. His medical history included hypertension, ischemic heart disease treated with beta-blockers, and hypothyroidism managed with levothyroxine. There was no history of occupational exposure to known carcinogens.

Symptoms progressively worsened, with bilateral nasal obstruction, anosmia, and extension to the oral cavity. There were no visual disturbances, neurological symptoms, or cervical pain. General condition was preserved at presentation.

Endoscopic nasal examination revealed inflammatory mucosa and a rounded, friable submucosal mass involving the anterior-inferior nasal septum and nasal floor, extending bilaterally to the lateral nasal walls and inferior meatuses. The lesion bled on contact and prevented visualization of the nasopharynx. Extension to the hard palate was noted. Cervical lymph node examination was unremarkable **(Figure 1).**

Contrast-enhanced computed tomography demonstrated a large heterogeneous tissue mass occupying the nasal fossae, extending posteriorly to the nasopharynx, with bone lysis of the medial maxillary sinus wall, ethmoidal invasion, destruction of the sphenoid sinus floor, and erosion of the hard palate **(Figure 2**). Magnetic resonance imaging confirmed a heterogeneous endonasal lesion with extension to the choanae, ethmoid cells, skull base (crista galli), and periorbital fat, without intracranial parenchymal invasion **(Figure 3).** Multiple endoscopic biopsies were performed. Histopathological examination with immunohistochemistry revealed a monoclonal plasma cell proliferation with lambda light-chain restriction, confirming the diagnosis of plasmacytoma. Subsequent hematological workup, including serum protein electrophoresis, immunofixation, bone marrow biopsy, and skeletal assessment, established the diagnosis of multiple myeloma.

Following multidisciplinary discussion, the patient was referred to the hematology department and initiated on systemic therapy according to current guidelines. Treatment consisted of a combination regimen including a proteasome inhibitor, an immunomodulatory agent, and corticosteroids, adapted to the patient's age and comorbidities. Local radiotherapy was not indicated given the systemic nature of the disease. The patient is currently under hematological follow-up.

DISCUSSION:

Extramedullary plasmacytoma of the head and neck region is rare but represents the most common site of extramedullary plasma cell tumors [1,2]. Endonasal localization poses a diagnostic challenge because of its nonspecific clinical presentation, often mimicking sinonasal carcinoma, lymphoma, melanoma, or inflammatory disease [5].

Radiologically, plasmacytomas typically present as aggressive soft-tissue masses with associated bone destruction. Computed tomography is useful for assessing osseous involvement, while magnetic resonance imaging provides superior evaluation of soft tissue extension, skull base invasion, and orbital or intracranial spread [6]. However, imaging features remain nonspecific, making histopathological confirmation mandatory.

Diagnosis relies on the demonstration of monoclonal plasma cell proliferation, confirmed by immunohistochemical evidence of light-chain restriction. Once plasmacytoma is diagnosed, systematic evaluation to exclude or confirm multiple myeloma is essential, including hematological, medullary, and skeletal assessment [3,7]. Multiple myeloma accounts for approximately 10% of hematological malignancies and predominantly affects elderly patients, with incidence increasing with age [3].

Progression from extramedullary plasmacytoma to multiple myeloma has been reported in up to 40% of cases, particularly in older patients and those with extensive local disease or bone involvement [4,8]. In the present case, advanced local extension and patient age were consistent with a high likelihood of systemic disease at diagnosis.

Therapeutic management depends on disease extent. Solitary extramedullary plasmacytoma is usually treated with radiotherapy, achieving excellent local control rates exceeding 90% [9]. In contrast, plasmacytoma associated with multiple myeloma requires systemic therapy. Recent advances in treatment, including proteasome inhibitors, immunomodulatory drugs, monoclonal antibodies, and corticosteroids, have significantly improved survival and quality of life, although the disease remains incurable [3,10]. Management should be individualized and multidisciplinary.

CONCLUSION:

Endonasal extramedullary plasmacytoma is a rare but aggressive entity that should be considered in the differential diagnosis of destructive nasal cavity masses. Histopathological confirmation and systematic hematological evaluation are mandatory to distinguish solitary disease from multiple myeloma. Early diagnosis and multidisciplinary management are essential to optimize patient outcomes in this challenging condition.

FIGURES :

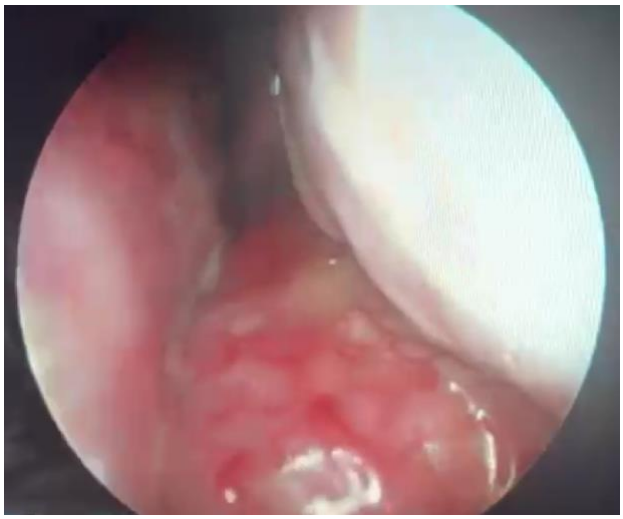

Figure 1: Nasal endoscopy revealed a friable submucosal mass involving both nasal fossae, taking the nasal septum and both inferior meatuses; the middle meatuses were free and the nasopharynx was not visualized due to the mass.

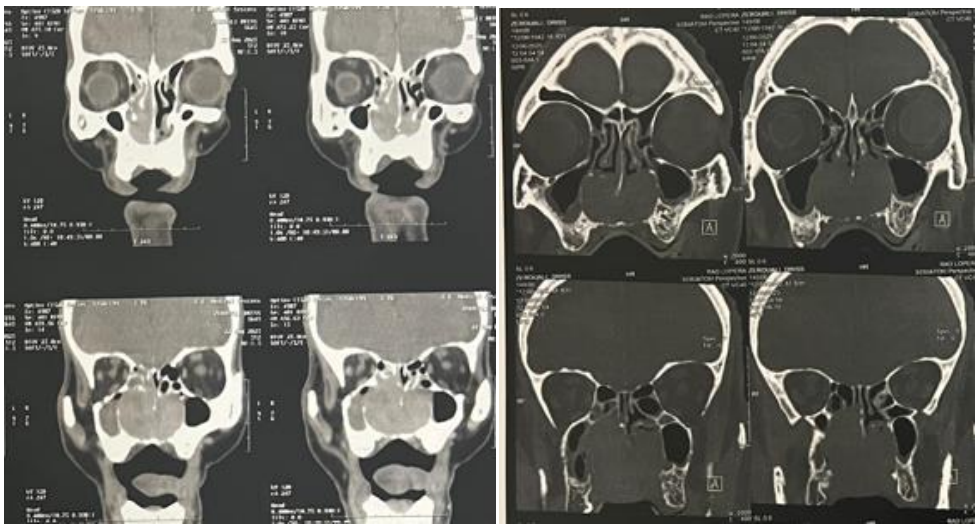

Figure 2: Contrast-enhanced CT showed a large heterogeneous sinonasal mass with nasopharyngeal extension, ethmoidal invasion, erosion of the maxillary sinus medial wall, sphenoid sinus floor, and focal hard palate destruction.

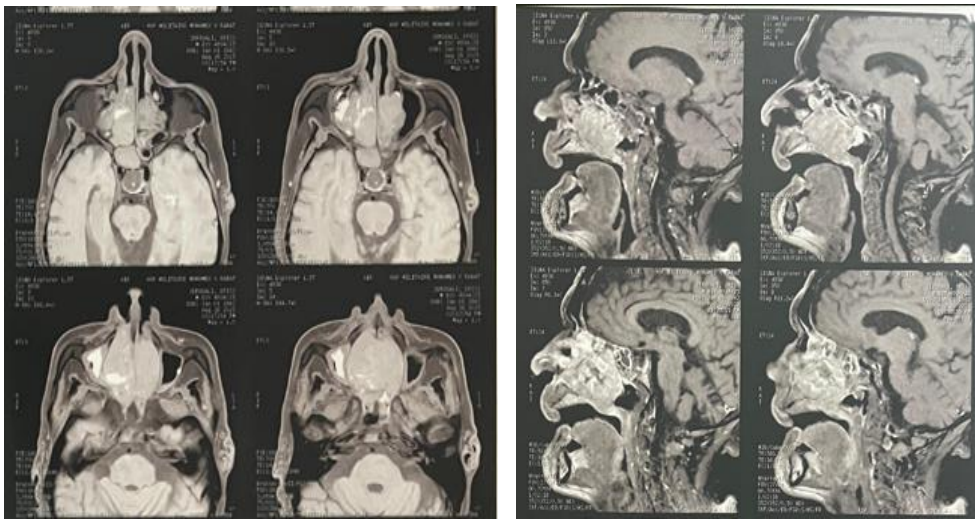

Figure 3: MRI (sagittal and coronal views) showing a heterogeneously enhancing sinonasal tumor with choanal, ethmoidal, skull base (crista galli), right orbital, and maxillary sinus medial wall extension, without intracerebral involvement.

ACKNOWLEDGEMENTS

Funding
No funding was received for this study.

Conflicts of Interest
The authors declare no conflicts of interest.

Patient Consent
Written informed consent was obtained from the patient for publication of this case and any accompanying images.

REFERENCES

1. Paiva B, et al. Extramedullary disease in multiple myeloma. Blood. 2021.

2. Varettoni M, et al. Clinical relevance of extramedullary disease in plasma cell neoplasms. Haematologica. 2022.

3. Dimopoulos MA, et al. Multiple myeloma: ESMO Clinical Practice Guidelines. Ann Oncol. 2021.

4. Rajkumar SV. Multiple myeloma: 2023 update on diagnosis and management. Am J Hematol. 2023.

5. Katodritou E, et al. Extramedullary plasmacytoma of the upper aerodigestive tract. Cancers (Basel). 2023.

6. Lee JY, et al. Imaging of head and neck plasmacytomas. Neuroradiology. 2021.

7. NCCN Clinical Practice Guidelines in Oncology. Multiple Myeloma. Version 2024.

8. Touzeau C, Moreau P. How I treat extramedullary myeloma. Blood. 2022.

9. Tsang RW, et al. Radiotherapy for solitary plasmacytoma. Int J Radiat Oncol Biol Phys. 2021.

10. Kumar SK, et al. Novel therapies in multiple myeloma. Lancet Oncol. 2022.