

Solitary Extramedullary Plasmacytoma of the Sinonasal Region: A Case Report

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Abstract

We report the case of a 74-year-old woman who presented with left-sided nasal swelling and epistaxis. Clinical examination revealed a mass in the left nasal cavity with absent airflow through the affected side. No cervical lymphadenopathy was detected. Cervicofacial CT revealed a lesion occupying the left nasal cavity and ethmoid air cells, with destruction of the surrounding osteocartilaginous structures. MRI revealed a large mass in the left nasal cavity, measuring 45 × 27 × 45 mm, with extensive involvement of adjacent structures. Histopathological examination of the biopsy specimen established the diagnosis of plasmacytoma of the nasal cavity. The systemic evaluation did not establish a diagnosis of multiple myeloma. Radiotherapy was recommended. This case highlights the clinical presentation, treatment, and disease course of solitary extramedullary plasmacytoma in an older adult.

Categories: Medical Education, Pathology, Anatomy

Keywords: case report, multiple myeloma, plasmacytoma, radiotherapy, surgery

Introduction

Solitary extramedullary plasmacytoma is a rare malignant plasma cell neoplasm, accounting for about 3% of plasma cell tumors, with an estimated incidence of 0.04 cases per 100,000 individuals [1]. Most solitary extramedullary plasmacytomas arise in the head and neck region and represent about 0.5% of malignant tumors affecting the upper respiratory tract [2]. The main differential diagnosis includes multiple myeloma, reactive plasmacytosis, lymphoma, nasopharyngeal carcinoma, and other sinonasal malignancies [2]. This tumor predominantly affects adults older than 60 years and is more common in men. Although radiotherapy remains the standard treatment, the optimal radiation dose and target volumes remain subjects of debate [3]. We present the case of a 74-year-old woman with a solitary extramedullary plasmacytoma of the nasal cavity and review the clinical presentation, treatment, and disease course of this rare tumor.

Case Presentation

A 74-year-old woman with a history of treated hypertension, diabetes mellitus, and ischemic heart disease presented with left-sided nasal swelling, epistaxis, and progressively declining visual acuity over 10 months. Nasal endoscopy revealed an exophytic mass in the left middle meatus that bled easily on contact, accompanied by fullness of the left nasolabial fold and absence of airflow through the left nasal cavity (Figure 1). No cervical lymphadenopathy was detected. Cervicofacial CT revealed an isodense lesion occupying the left nasal cavity, left ethmoid air cells, and sphenoid sinus, with mild enhancement following contrast administration. The lesion caused destruction of the surrounding osteocartilaginous structures. MRI revealed a large mass in the left nasal cavity, measuring 45 × 27 × 45 mm, with extension into the left maxillary sinus and orbit. The mass compressed and encased the prechiasmatic optic nerve and infiltrated the left orbital muscles. Additional findings included destruction of the skull base, extension into the left sphenoid sinus, and infiltration of the left cavernous sinus and adjacent meninges (Figure 2). Histopathological examination and immunohistochemical analysis supported the diagnosis of a plasma cell neoplasm of the nasal cavity (Figure 3). Protein immunoelectrophoresis revealed monoclonal gamma-kappa immunoglobulin. Bence-Jones proteinuria was negative. Renal function, complete blood count, serum calcium, and serum β2-microglobulin levels were within normal limits. Bone marrow aspiration, bone marrow biopsy, and additional systemic imaging revealed no abnormalities. The systemic evaluation did not establish a diagnosis of multiple myeloma. CD138, CD38, MUM1, kappa/lambda light-chain restriction, CD20, CD3, CK, S100, EBER, Ki-67 proliferation index, and clonal plasma cells were all negative, and the diagnosis was solitary extramedullary plasmacytoma of the sinonasal region. Given the patient's general condition and the locally advanced tumor, radiotherapy was administered at a total dose of 45 Gy in 25 fractions over five weeks (1.8 Gy per fraction). At the 11-month follow-up, the patient remained in complete clinical and radiological remission, with no detectable monoclonal protein.

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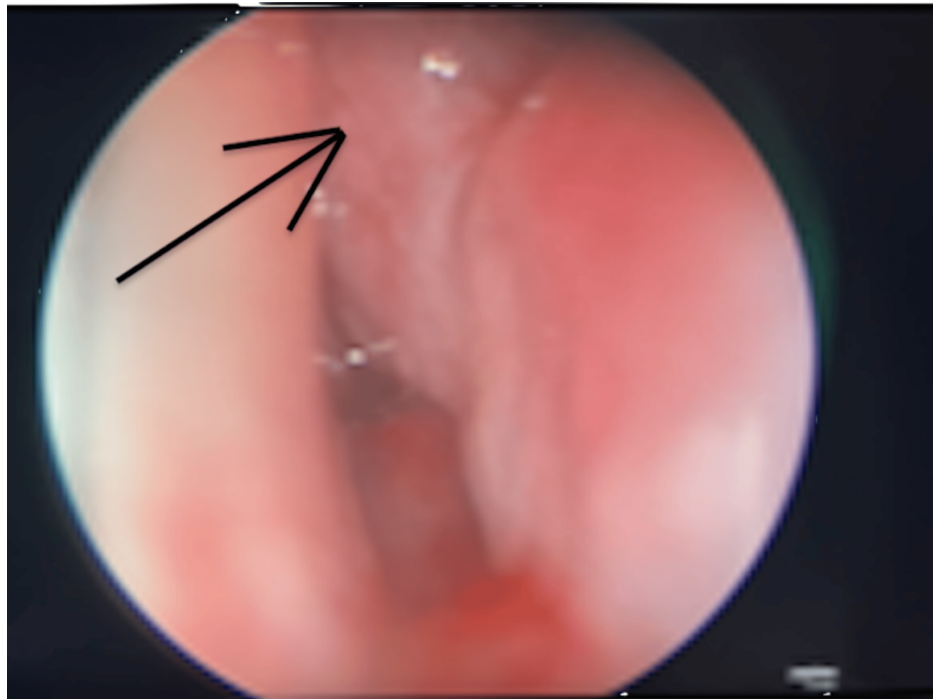


FIGURE 1: Nasal endoscopy showing an exophytic mass in the left middle meatus that bleeds easily on contact (black arrow)

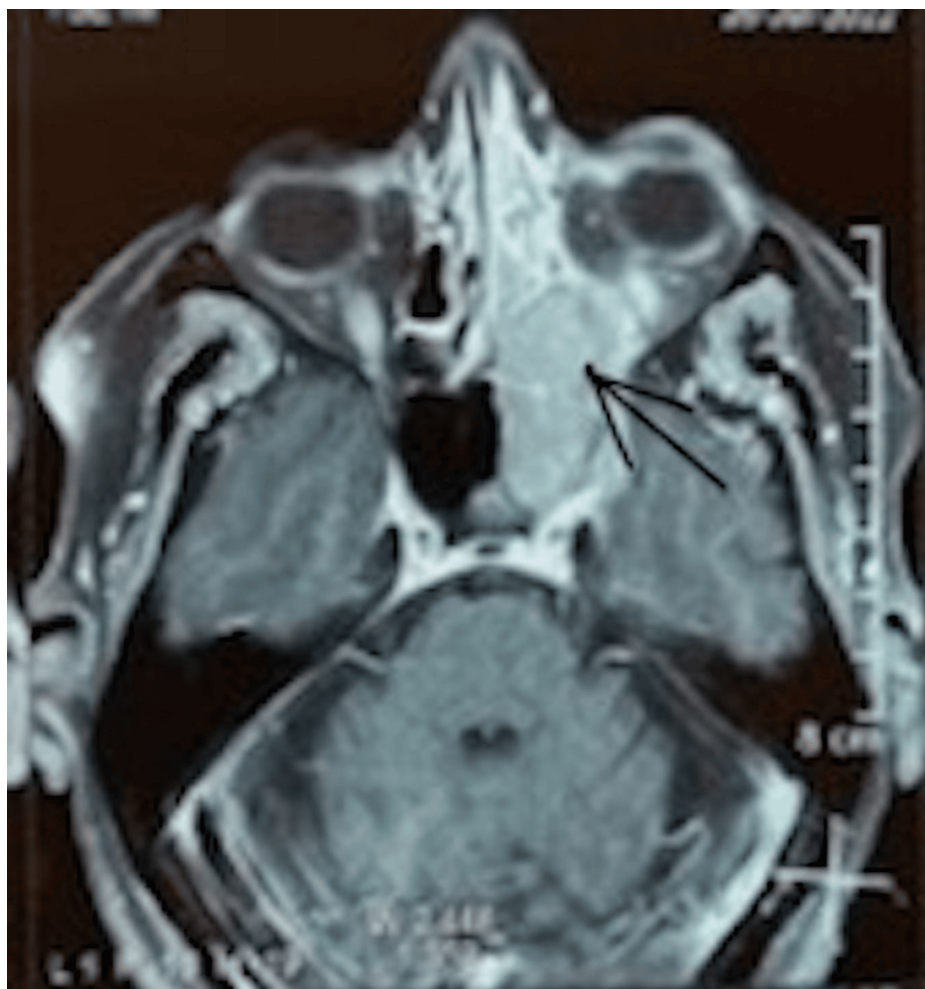


FIGURE 2: MRI showing a large mass in the left nasal cavity measuring $45 \times 27 \times 45$ mm, with extension into the left maxillary sinus, orbit, skull base, and cavernous sinus (black arrow)

MRI: magnetic resonance imaging

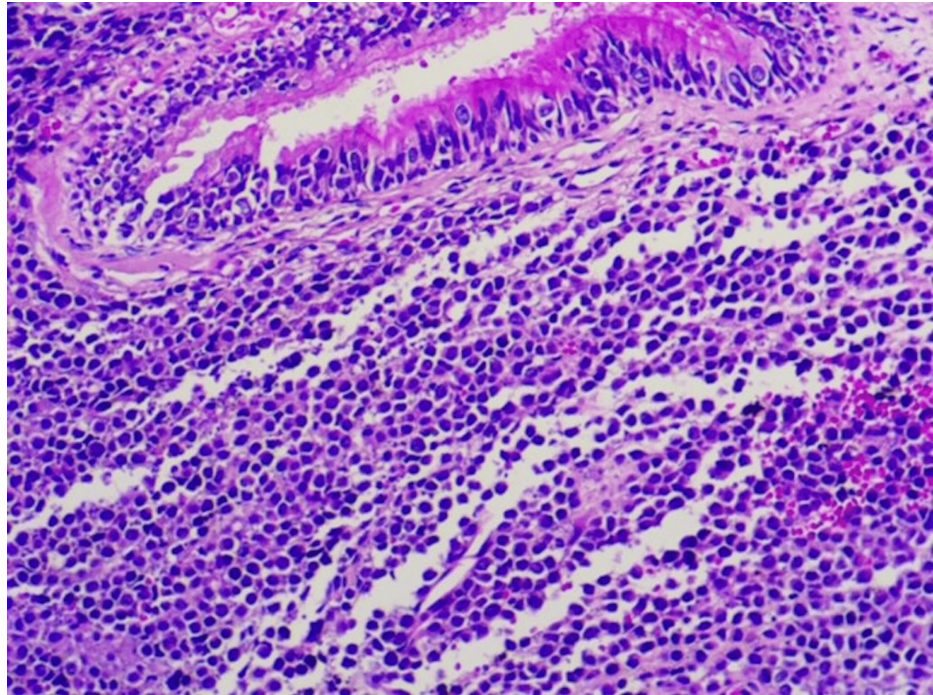


FIGURE 3: Photomicrograph showing infiltration of the lamina propria by mature and immature plasma cells, some exhibiting a high nuclear-to-cytoplasmic ratio (hematoxylin and eosin staining, ×200)

H&E: hematoxylin and eosin

Discussion

Solitary extramedullary plasmacytomas account for about 3% of solitary plasmacytomas and most commonly arise in the submucosal tissues of the upper aerodigestive tract, where they represent about 0.4% of malignant head and neck tumors. Histopathological examination of a biopsy or surgical specimen establishes the diagnosis of plasmacytoma. The diagnosis is made by exclusion, particularly after multiple myeloma has been ruled out [4].

Clinical manifestations may include headache and cranial nerve involvement, particularly of the oculomotor nerve [5]. However, plasmacytoma accounts for fewer than 4% of invasive cavernous sinus lesions [6]. In our patient, the disease involved both osseous and neurological structures, with extension into the cavernous sinus.

In otolaryngology practice, solitary extramedullary plasmacytoma may be diagnosed incidentally following biopsy [6]. In our patient, neither the clinical presentation nor the gross appearance of the nasal cavity lesion initially suggested plasmacytoma. The diagnosis was established through histopathological examination and systemic evaluation, including bone marrow biopsy and imaging to exclude multiple myeloma [1,7].

On CT imaging, the lesion usually appears as a mildly heterogeneous mass with moderate enhancement after contrast administration. Osteocartilaginous destruction may also be observed. However, these radiological findings are not specific and may resemble those seen in lymphomas or nasopharyngeal carcinomas. On MRI, the tumor is generally isointense on T1-weighted images, with marked enhancement after contrast and central heterogeneity [8].

Curative surgery is rarely indicated because of its potential morbidity and the marked radiosensitivity of these tumors [9]. Nevertheless, surgery may be considered a salvage option for patients who experience recurrence or disease progression following radiotherapy. Radiotherapy remains the standard treatment for solitary extramedullary plasmacytoma of the upper aerodigestive tract, either as the sole treatment or following surgery performed primarily for diagnostic purposes. Radiotherapy alone achieves local control in approximately 70%-100% of cases [9]. Local control appears to be associated with radiation dose, with doses of at least 45 Gy suggested in the cited literature. Prophylactic irradiation of regional lymph nodes has not been shown to provide a clear benefit in terms of locoregional control. Currently, there is insufficient evidence to support the routine use of adjuvant chemotherapy in the management of solitary

extramedullary plasmacytoma.

Long-term follow-up is necessary because of the risk of progression to multiple myeloma, which is a major prognostic factor alongside tumor size and lymph node involvement. Reported 10-year overall survival ranges from 50% to 80% [10].

Conclusions

Plasmacytoma should be considered in the differential diagnosis of invasive sinonasal lesions. Its clinical presentation is variable and may be nonspecific. Imaging findings can help suggest the diagnosis, but histopathological confirmation remains essential. Radiotherapy remains the standard treatment and may be administered alone or following surgery. The risk of progression to multiple myeloma is a major prognostic consideration. Therefore, regular clinical, laboratory, and imaging follow-up is essential.

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.

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