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(CASE REPORT)

Solitary extramedullary plasmacytoma of the nasopharynx: A case report and literature review

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Abstract

Solitary plasmacytoma (SP) is a plasma cell disorder characterized by a localized accumulation of neoplastic monoclonal plasma cells in bone or soft tissues, without systemic involvement or end-organ damage. SPs are classified into two groups: solitary bone plasmacytoma (SBP) and solitary extramedullary plasmacytoma (SEP). SEP, a less common presentation (20% to 30% of cases), occurs predominantly in the head and neck region (such as the nasal cavity, paranasal sinuses, and nasopharynx) but can also arise, though rarely, in soft tissues, the gastrointestinal tract, skin, and lymph nodes. Cervical lymph node involvement is observed in 10-15% of head and neck SEP cases. Common symptoms include epistaxis, rhinorrhea, nasal obstruction, sore throat, dysphonia, and hemoptysis. Definitive radiation therapy is the standard treatment, offering long-term local control and potentially curative outcomes in extramedullary cases. This case report presents a 39-year-old female with persistent nasal obstruction, later diagnosed with nasopharyngeal extramedullary plasmacytoma. The patient underwent exclusive radiation therapy, resulting in a satisfactory outcome. Additionally, we review the current literature on the definition, diagnosis, management, follow-up procedures, and perspectives of solitary extramedullary plasmacytoma, with a focus on ILROG recommendations guiding best practices in the use of radiation therapy for SEP.

Keywords: Solitary Extramedullary Plasmacytoma; Radiation Therapy; Local Control; ILROG

1. Introduction

Plasma cell neoplasms are malignancies of mature B cells, comprised of clonal plasma cells that are terminally differentiated and characterized by immunoglobulin secretion. While the majority of plasma cell tumors are diagnosed as multiple myeloma (MM) and typically affect older adults, around 5% to 6% present as solitary plasmacytomas (SPs), located either in bone or extramedullary tissues (SEP). Unlike MM, which primarily requires systemic chemotherapy, solitary plasmacytomas are managed with definitive radiation therapy (RT), known for providing durable local control with curative intent. SPs are highly relevant to radiation oncology due to their radiosensitivity and the high efficacy of localized RT in treating these tumors. However, with recent advances in imaging and RT techniques, along with the development of new systemic therapies, there is a recognized need to update guidelines for the integration of RT in the management of SEPs. The International Lymphoma Radiation Oncology Group (ILROG) has formed an expert panel to review the literature and develop consensus guidelines for RT planning in these conditions [1]. This case report illustrates the effective use of exclusive RT in a 39-year-old female with a nasopharyngeal solitary extramedullary plasmacytoma, highlighting the importance of radiation therapy in the treatment of these rare tumors.

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2. Case report

A 39-year-old woman was referred to our Department of Radiation Therapy at HASSAN II University Hospital (Fez, Morocco) with a 24-month history of persistent nasal obstruction and intermittent epistaxis. Physical examination revealed a tumor on the right posterolateral wall of the nasopharynx, which was prone to bleeding upon contact. Computed tomography showed a mass measuring approximately 32 x 20 x 33 mm in the right posterolateral wall of the nasopharynx, extending to the oropharynx and parapharyngeal space without bony involvement or endocranial extension. Bilateral jugulocarotid lymphadenopathy was also detected, with the largest lymph node measuring 15 mm on the left (**Fig. 1**). Histopathological examination of the biopsy revealed a low-grade malignancy consistent with lymphoplasmacytic lymphoma, plasma cell type. Immunohistochemical staining confirmed the presence of plasma cell markers (Mum1 and CD138) and the absence of CD3, CD20, CD34, synaptophysin, and HMB45. Additionally, tumor cells expressed low levels of monoclonal immunoglobulin G with kappa light chains. The patient underwent a cerebro-thoraco-abdomino-pelvic CT scan, which showed no distant lesions. There were no signs of CRAB criteria: complete blood count, calcium, creatinine, and uric acid levels were within normal limits, and a whole-body bone survey showed no detectable osteolytic lesions. The patient received exclusive external radiation therapy using intensity-modulated radiation therapy (IMRT), targeting the primary tumor and lymph node regions with a total dose of 60 Gray, administered in 30 fractions of 2 Gray each, delivered once daily over five days per week (**Fig. 2**).

A follow-up CT scan performed three months after the end of radiation therapy showed partial tumor regression (20x15mm compared to 32x20x33mm initially), and another scan at six months showed further regression (16x14mm compared to 20x15mm). An 18FDG PET scan one year after radiation therapy indicated complete remission, with no abnormal fixation observed.

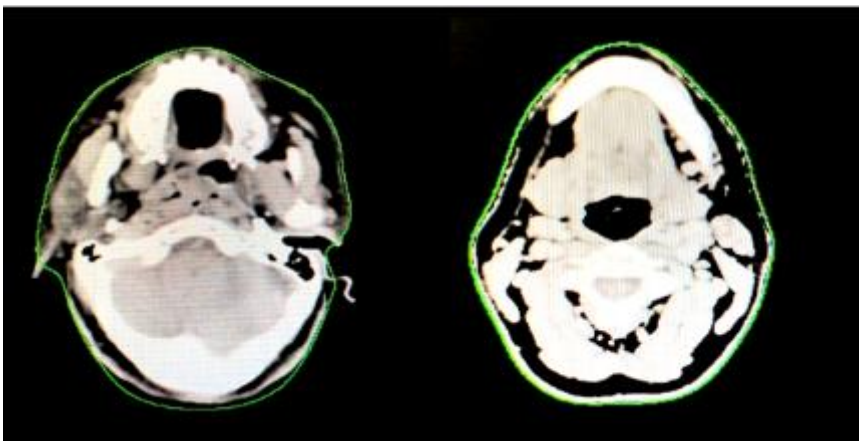

Figure 1 CT images of a solitary extramedullary plasmacytoma in the nasopharynx with nodal involvement

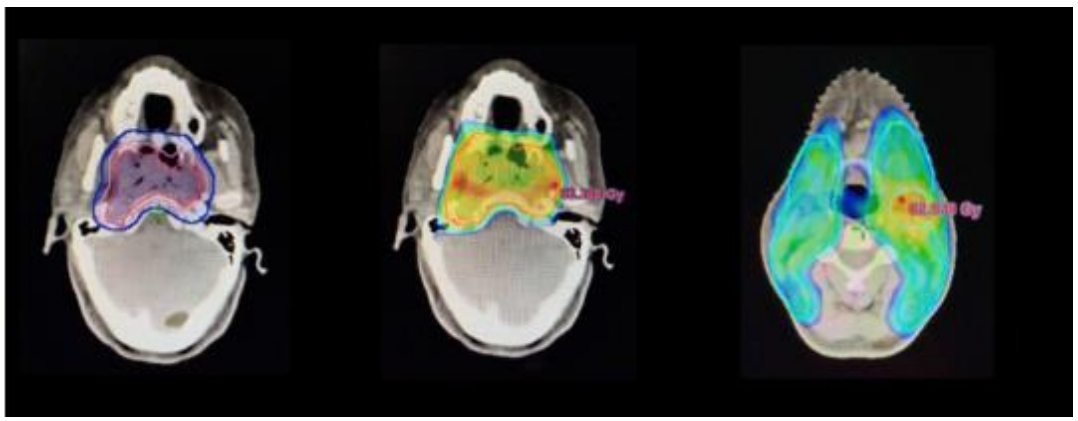

Figure 2 Radiation Dose Distribution to Target Volumes

3. Discussion

Solitary plasmacytoma (SP) is a plasma cell disorder marked by a localized accumulation of neoplastic monoclonal plasma cells, either in bone or in soft tissues without systemic involvement [2]. SPs are categorized based on location: solitary bone plasmacytoma (SBP) and solitary extramedullary plasmacytoma (SEP). SEP, a less common form (20-30% of cases)[3.4], primarily occurs in the head and neck region, such as the nasal cavity, paranasal sinuses, and nasopharynx, but can also rarely appear in soft tissues, the gastrointestinal tract, skin, and lymph nodes [3.4]. Our patient's case is particularly intriguing, as she presents with a rare SEP located in the nasopharynx, along with lymph node involvement—an occurrence that is even more uncommon and scarcely documented in the literature.

SEPs are often localized and show a favorable response to local therapies like radiation therapy (RT), resulting in long-term control with a higher cure rate compared to SBP [3.4]. In rare cases, SP can be associated with POEMS syndrome, a condition involving polyneuropathy, organomegaly, endocrinopathy, a monoclonal plasma cell disorder, and skin abnormalities [5]. In patients with POEMS syndrome, RT may improve or alleviate symptoms associated with the syndrome in about half of cases [6].

The International Myeloma Working Group has established diagnostic criteria for solitary plasmacytomas to distinguish them from systemic plasma cell disorders [2]. Diagnosis of solitary bone plasmacytoma or solitary extramedullary plasmacytoma requires [1.2]:Biopsy-proven solitary lesion, Bone marrow biopsy and aspiration should show an absence of clonal plasma cells. A normal skeletal survey and magnetic resonance imaging (MRI) (or computed tomography, CT) of the spine and pelvis, except for the identified solitary lesion. If available, a positron emission tomography/computed tomography (PET/CT) should confirm the presence of a solitary lesion, supporting the absence of additional disease. And, absence of CRAB criteria.

In cases with minimal bone marrow involvement (under 10% clonal plasma cells), these criteria apply, though even minor marrow involvement may increase the risk of progression to multiple myeloma [1.2].

Furthermore, Solitary bone plasmacytomas carry a high risk of progression to multiple myeloma (MM), with rates reaching 65-84% within 10 years and nearly 100% by 15 years [4, 7, 8]. In contrast, solitary extramedullary plasmacytomas have a lower progression risk of 10-30% over 10 years but a slightly higher local recurrence risk [4, 8]. Therefore, the optimal treatment for SEPs aims for durable local control and standard treatment is definitive local radiation therapy, which provides high local control rates of 85-90%, potentially leading to long-term remission and even cure [9]. SEPs commonly arise in the head and neck region, and about 25% of these cases may show regional nodal disease on imaging using MRI and PET/CT [7.10.11.12]. In these cases, definitive RT coverage of involved lymph nodes is required, and elective coverage of at-risk adjacent nodes should be considered. However, the role of prophylactic nodal irradiation is still debated, particularly when the SEP involves structures like Waldeyer's ring, as the benefit of prophylactic nodal irradiation is uncertain when no nodal involvement is seen on modern imaging [1]. Prior to the development of conformal RT and advanced imaging, prophylactic cervical lymph node irradiation was common, and regional nodal failures were rare [7.10.11].

In terms of radiation dose, The optimal radiation dose for solitary plasmacytoma (SP) is not well established, as most studies have weak dose-response data due to small patient numbers and a limited range of doses. A large multi-institutional study (n = 258) did not show a dose-response relationship beyond 30 to 35 Gy [4]. Despite this, it is common practice to use doses between 40 and 45 Gy [13]. Mendenhall et al. reported a local control rate of 94% with doses of 40 Gy or above, compared to 69% with doses lower than 40 Gy in a retrospective review of 81 patients [14]. Several studies using larger radiation doses (45–60 Gy) did not demonstrate any clear advantage, with sporadic local failures still occurring even at doses of 50 to 60 Gy [10.15.16]. The ILROG panel recommends, in the case of SEPs, a total dose of 40 to 50 Gy for optimal tumor control, based on evidence and expert consensus [1]. For small, well-defined tumors or cases post-excision with positive margins, 40 Gy may be sufficient. These guidelines are based on daily fractions of 1.8-2 Gy [12, 17].

The determination of target volumes is essential for precise radiation therapy delivery. The gross tumor volume (GTV) is outlined using primary imaging and should be fused with simulation studies (e.g., PET/CT or MRI) to accurately define it. The clinical target volume (CTV) includes the GTV and potential microscopic disease, with a margin of 0.5 to 3 cm in all directions for definitive radiation therapy. For soft tissue tumors, such as in the head and neck, a 0.5 to 1 cm expansion may be sufficient. Elective regional nodal irradiation for solitary extramedullary plasmacytoma (SEP) in the head and neck is debated. Prophylactic nodal irradiation is not recommended when imaging shows no involvement of cervical lymph nodes, except when there is known regional involvement. The planning target volume (PTV) includes the CTV or internal target volume (ITV), expanded to account for treatment uncertainties. The margin size depends on

factors like immobilization and patient compliance. In hematologic malignancy treatment, careful consideration of organs at risk (OARs) is crucial, with radiation doses adjusted to minimize damage. Advanced techniques like IMRT or VMAT may offer better sparing of critical structures, and image guidance during treatment is beneficial for tumors near vital organs. In head and neck cases, immobilization with a thermoplastic mask ensures precise planning and treatment [1].

The role of systemic therapy, except in cases of progression to multiple myeloma, remains uncertain. While Suh et al. observed a positive trend for improved MMFS (median metastasis-free survival), the absolute 5-year MMFS rate was worse by almost 50% in patients who did not receive chemotherapy after radiation therapy ($p = 0.08$)[18]. However, due to the small size of the group, this observation cannot be used as a basis for making changes to the treatment algorithm in any significant way. At most, it could be seen as an attempt to identify a subgroup at high risk of local treatment failure, such as those with bulky tumors or incomplete remission [19]. Considering the effectiveness of radiation therapy in controlling local disease, and the higher incidence of secondary hematological malignancies caused by melphalan-based chemotherapy, there is no strong evidence from large randomized multicenter studies to support the use of chemotherapy in clinical practice [20].

Follow-up procedures involve regular clinical assessments, as well as serum and urine tests to monitor for persistent M-protein, typically every six months. In patients with SP and detectable M-protein prior to definitive radiation therapy, successful treatment is marked by the disappearance of M-protein, though this may take several months post-RT. Persistent M-protein indicates a high risk of progression to multiple myeloma [10.21.22]. Routine monitoring includes complete blood counts, serum chemistry tests [such as calcium and creatinine], and occasional skeletal surveys to detect any progression to MM [1].

Reimaging is crucial for assessing the response to SP treatment, ideally performed 3 to 6 months after RT. MRI or PET/CT scans are preferred for evaluating soft tissue responses. Minimal abnormalities on CT, even on MRI, may persist for several months after definitive RT and should not be mistaken for active disease. It may take 6 to 8 months for SP to reach its maximum therapeutic response. PET/CT scanning, when available, can offer earlier metabolic response detection, even when routine imaging shows persistent abnormalities [23.24]. For our patient, both the first CT scan at 3 months and the follow-up CT at 6 months showed partial remission, but the PET-CT at 1 year revealed a complete remission, highlighting that such residual abnormalities can occur and should not be misinterpreted as treatment failure. Reimaging every 4 to 6 months may be considered for any residual tumor mass until a complete response is achieved, or until any remaining abnormalities stabilize. It is generally unnecessary to continue imaging minor stable residual abnormalities unless clinically indicated [1].

Combining novel agents with RT, especially for bulky plasmacytomas, has not been extensively studied. The addition of adjuvant therapies like proteasome inhibitors or immunomodulatory drugs [Lenalidomide] to RT could improve local control and potentially eliminate subclinical disease, thus preventing the development of systemic MM. Preliminary data indicate that such combinations are feasible and effective [25.26]. A phase 3 study in the UK is investigating the potential benefits of adding lenalidomide and dexamethasone to RT to enhance progression-free survival [27].

4. Conclusion

In conclusion, this case study highlights the effectiveness of exclusive radiation therapy in treating solitary extramedullary plasmacytoma in the nasopharyngeal region, a rare localization, offering durable local control and potential cure without the need for systemic treatments. It is also important to adopt a rigorous therapeutic approach based on advanced imaging techniques for accurate tumor volume delineation, minimizing safety margins, and sparing surrounding organs at risk. Our findings support the ILROG guidelines, emphasizing that although SEP carries a low risk of progression to multiple myeloma, continuous monitoring is crucial to detect any local recurrence or systemic progression.

Compliance with ethical standards

Disclosure of conflict of interest

All authors have no conflict of interest to declare.

Informed Consent

The patient provided informed consent for the publication of this case report.

Statement of ethical approval

This case report was conducted in accordance with ethical guidelines. Informed consent was obtained from the patient for publication.

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