



Case Report *Neuroradiology/Head and Neck Imaging*

# Solitary extramedullary plasmacytoma of the nasopharynx: A case report

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## ABSTRACT

Solitary extramedullary plasmacytoma (SEP) is a rare form of plasma cell neoplasm that occurs in the soft tissue without systemic involvement. We report the case of an 81-year-old female patient who presented with intermittent dysphagia and was found to have a right nasopharyngeal mass diagnosed as a plasmacytoma. Diagnostic workup included endoscopy, biopsy, imaging, and laboratory analysis, which ruled out multiple myeloma. The patient underwent definitive radiation therapy, and at evaluation 4 months later, there was no metabolic or endoscopic evidence of any disease, except for minor toxicity limited to nocturnal xerostomia. This case underscores the importance of timely diagnosis and the role of radiotherapy in treating SEP, particularly in uncommon anatomical locations.

**Keywords:** Case report, Extramedullary plasmacytoma, Nasopharynx, Radiation therapy

## INTRODUCTION

Clonal plasma cell proliferative disorders represent a cytogenetically heterogeneous spectrum of diseases. According to the International Myeloma Working Group, a solitary plasmacytoma (SP) is a biopsy-proven solitary lesion of bone or soft tissue with evidence of clonal plasma cells, but without evidence of bone marrow involvement or end-organ damage that can be attributed to a lymphoplasmacytic proliferative disorder.<sup>[1]</sup> Typical examples of end-organ damage are calcium elevation, renal insufficiency, anemia, and bone lesions (criteria). On skeletal survey and computed tomography (CT)/magnetic resonance imaging (MRI) of the spine and pelvis, there should be no evidence of disease except for the primary solitary lesion.

SP is a rare plasma cell dyscrasia that can occur locally in the bone, known as solitary bone plasmacytoma (SBP) or in soft tissue, known as solitary extramedullary plasmacytoma (SEP). The clinical presentation of SEP can be diverse, often manifesting as localized masses in soft tissue that may be mistaken for other malignancies or benign conditions. The rarity of SEP and its often-nonspecific symptoms can delay diagnosis, making it important for clinicians to consider SEP in differential diagnoses when encountering soft-tissue tumors. In addition, SEPs' potential for progression to multiple myeloma (MM) necessitates careful monitoring and sometimes aggressive management.

This case report highlights the diagnosis and management of a patient with a rare presentation of SEP in the nasopharyngeal region, emphasizing the importance of timely intervention with appropriate imaging and comprehensive workup to exclude MM, and the role of radiation therapy

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(RT) in achieving optimal outcomes. The unique aspects of this case, including initial incidental finding and biopsy on esophagoscopy of a right nasopharyngeal mass, underscore the need for further investigation into the clinical behavior of SEP and early management. Written informed consent was obtained from the patient for publication of this case and accompanying images.

## CASE REPORT

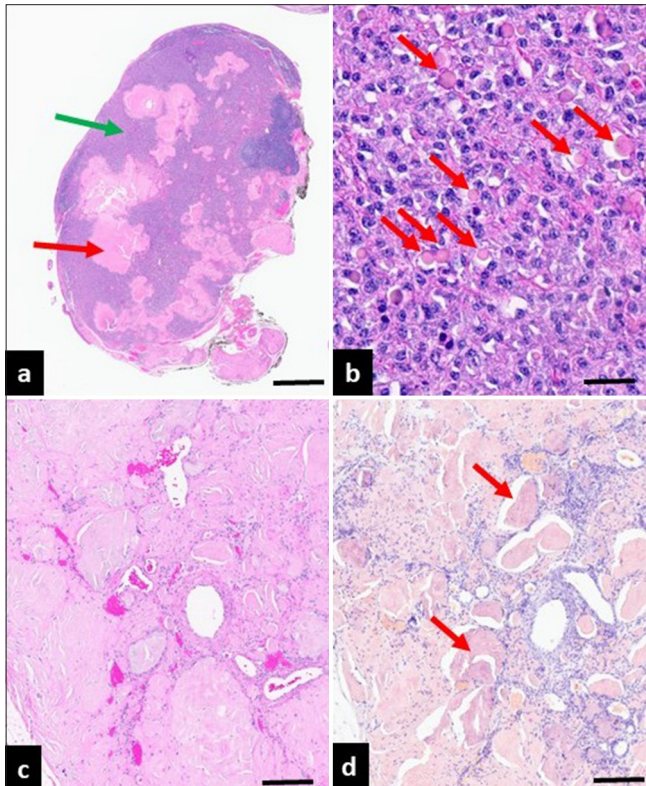
An 81-year-old female patient was diagnosed with SEP after presenting with intermittent dysphagia for 1 year. Initially, gastroenterological evaluation, including manometry and barium swallow, showed no structural abnormalities but identified a prominent cricopharyngeal bar. She was referred to otolaryngology for further evaluation and potential Botox.

Further investigation with esophagoscopy revealed a nasopharyngeal mass located on the right side, prompting

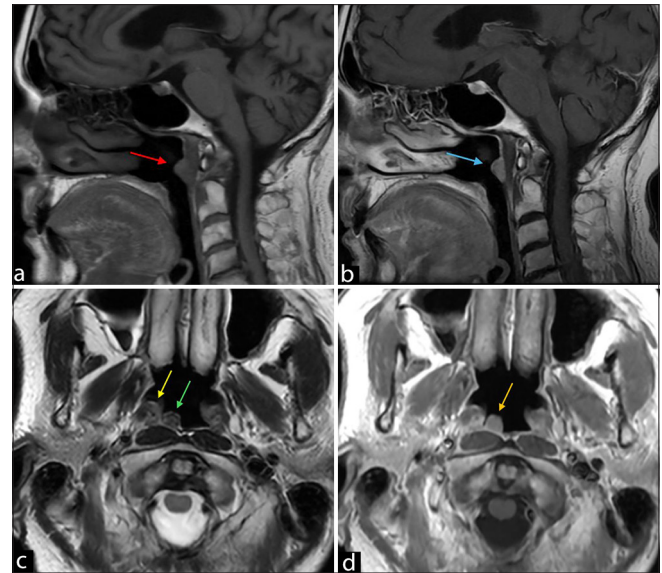
an excisional biopsy. Pathology confirmed a kappa-restricted plasma cell neoplasm, with Congo red staining positive for amyloid [Figure 1]. Laboratory tests, including complete blood count and comprehensive metabolic panel, were within normal limits. Serum electrophoresis revealed an M spike with paraprotein of 0.013 g/dL. Immunofixation showed IgG  $\lambda$  (possible biologic heterogeneity), while free light chain and immunoglobulin levels were within normal limits. N-terminal pro-B-type natriuretic peptide in the blood was elevated at 392 pg/mL, but urine studies were negative for proteinuria.

Immunohistochemistry demonstrated kappa light-chain restriction within the lesion, confirming a monoclonal plasma cell neoplasm. Serum immunofixation revealed a low-level IgG lambda paraprotein. Given the absence of systemic findings on bone marrow biopsy and PET/CT, this was interpreted as a localized clonal process rather than overt multiple myeloma.

The patient received further evaluation with an MRI with sagittal T1-weighted images of the brain and pharynx [Figure 2] without [Figure 2a] and with contrast [Figure 2b], which revealed a polypoid lesion in the nasopharynx that demonstrated intermediate signal intensity on non-contrast T1 (red arrow) [Figure 2a] and relatively homogeneous



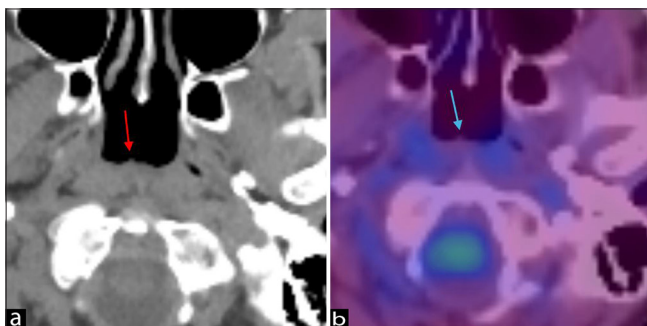
**Figure 1:** (a) Low-power hematoxylin and eosin (H&E) section of the mucosal tumor demonstrating a mass-like accumulation of plasma cells (Green arrow) with associated amyloid deposition (red arrow) (12.5 $\times$  magnification; scale bar = 100  $\mu$ m). (b) High-power H&E section showing abundant intracytoplasmic immunoglobulin inclusions (Russell bodies) within plasma cells (red arrows) (200 $\times$  magnification; scale bar = 20  $\mu$ m). (c) Medium-power H&E section demonstrating a separate amyloidoma composed of amorphous eosinophilic extracellular material (40 $\times$  magnification; scale bar = 100  $\mu$ m). (d) Congo red stain highlighting amyloid deposition (red arrow) (40 $\times$  magnification; scale bar = 100  $\mu$ m).



**Figure 2:** Sagittal T1-weighted MR images of the brain and nasopharynx in an 81-year-old female obtained (a) without and (b) with intravenous contrast demonstrate a polypoid mass within the nasopharynx. The lesion exhibits intermediate signal intensity on non-contrast T1-weighted imaging (red arrow) and relatively homogeneous enhancement on post-contrast T1-weighted imaging (blue arrow). (c) Axial T2-weighted and (d) axial post-contrast T1-weighted images reveal a protuberant soft tissue mass in the right nasopharynx, located medial to the fossa of Rosenmüller and torus tubarius (yellow arrow). The lesion demonstrates heterogeneous T2 signal intensity (green arrow) with relatively homogeneous post-contrast enhancement (orange arrow).

enhancement on post-contrast T1 (blue arrow) [Figure 2b]. On axial T2-weighted [Figure 2c] and axial post-contrast T1-weighted [Figure 2d] imaging, there is a protuberant lesion in the right nasopharynx located medially to the Fossa of Rosenmuller, and torus tubarius (yellow arrow), demonstrating heterogeneous T2 signal intensity (green arrow) [Figure 2c] and relatively homogeneous enhancement (orange arrow) [Figure 2d]. Positron emission tomography (PET)/CT demonstrated an irregular soft-tissue density in the right lateral nasopharyngeal wall and right maxillary sinus, with a small focus of mildly increased metabolic activity (maximum standardized uptake [SUVmax] 4.0). Although this small focus may have represented postsurgical changes, the high possibility of residual active disease could not be excluded. No evidence of other local or distant metastatic disease was observed [Figure 3]. Axial CT [Figure 3a] and PET [Figure 3b] images through the level of the pharynx after excision demonstrated minimal soft-tissue thickening on CT (red arrow) [Figure 3a] and barely discernible low-grade fluorodeoxyglucose avidity (blue arrow) [Figure 3b], representing post-treatment changes without residual or recurrent local disease. Bone marrow biopsy ruled out MM.

The patient was subsequently treated with volumetric-modulated arc therapy with treatment volumes including gross tumor volumes (GTV) initially as seen on imaging, plus a clinical target volume (CTV) generously covering the mucosa of the nasopharynx 0.5-3.0 cm around gross disease and a planning target volume (PTV) of 0.5 cm, using a daily cone-beam CT (CBCT) for image guidance. A dose of 50 Gy at 2 Gy per fraction in 25 fractions was prescribed to the PTV because the initial procedure was non-oncologic in the form of a biopsy. Treatment was well-tolerated with only mild oral soreness. At her 4-month follow-up visit, the patient reported nocturnal xerostomia, which may be attributable to irradiation of the minor ectopic salivary glands. PET/CT performed before this visit showed minimal soft-tissue



**Figure 3:** (a) Axial contrast-enhanced CT and (b) axial PET images obtained at the level of the nasopharynx following surgical excision demonstrate minimal residual soft tissue thickening on CT (red arrow) and faint, low-grade FDG uptake on PET (blue arrow). These findings are consistent with post-treatment changes without imaging evidence of residual or recurrent disease.

asymmetry in the right nasopharynx at the site of the excised tumor, but no significant hypermetabolic activity, suggesting post-surgical and radiation changes rather than recurrence. There were no hypermetabolic lymph nodes or distant lesions that would suggest nodal or metastatic recurrence. In addition, mucosal thickening was noted, more pronounced on the right side, along with periapical lucencies in the right upper molars extending into the right maxillary sinus, showing hypermetabolic activity (SUVmax 3.8). This was more consistent with odontogenic sinusitis than with active malignancy. A flexible fiberoptic direct nasal pharyngolaryngoscopy was also performed, which revealed no masses or lesions in the nasopharynx.

## DISCUSSION

SPs represent <5% of all plasma cell dyscrasias. They are categorized based on their locations into two types: SBP, which can progress to MM with rates as high as 60% within 3 years, and SEPs, which occur in soft tissue and have a lower progression rate of approximately 20% to MM within 3 years. SEPs are rarer than SBPs, making up about one-third of all SPs.<sup>[2]</sup> Unlike MM, plasmacytomas are considered “curable” and are of particular interest to radiation oncologists because the standard treatment with curative intent is definitive RT [Table 1]. These tumors are highly radioresponsive and radiocurable, with RT alone achieving excellent long-term local control rates of 79-91%. Conventional radiography has many limitations, including low sensitivity, so cross-sectional imaging with CT/MRI should be used for establishing a diagnosis, staging, and RT planning. Regarding MRI, SEP lesions are typically hypointense on T1-weighted images and enhancing with contrast, whereas they are hyperintense on T2-weighted and short tau inversion recovery (STIR) sequences, as seen particularly in our case. PET scans are very useful for local disease delineation, staging, and follow-up.

While the optimal RT dose is debated, durable responses are more commonly seen with doses >40 Gy.<sup>[3]</sup> SEPs are very radiosensitive, and excellent local controls can be achieved, as is also recommended by the National Comprehensive Cancer Network guidelines. The current guidelines recommend a dose range of 40-50 Gy. The Mayo experience reported no local failures when doses exceeded 45 Gy for SBP.<sup>[4]</sup> Notably, 23% of local failures came after 5 years, despite a median time to failure of 18 months. A French study found similar outcomes for SEPs.<sup>[5]</sup> In addition, the 2018 International Lymphoma Radiation Oncology Group (ILROG) guidelines recommend a total dose of 40-50 Gy for SEPs. In cases of small, well-defined, or post-excision with positive margins, 40 Gy is acceptable.

When nodal tissue is involved, definitive RT coverage is necessary, and elective coverage of adjacent nodes at risk should be considered. However, the benefit of prophylactic nodal irradiation is controversial, particularly when

**Table 1:** Unusual solitary extramedullary plasmacytoma (SEP) sites.

Site	Presentation	Imaging	Radiation therapy considerations	Dose range (Gy)
Orbit	Proptosis, diplopia, visual disturbances, periorbital swelling	CT: Homogeneous enhancement; MRI: Hypointense T1, hyperintense T2/STIR, enhances with contrast	Requires precise targeting to spare optic structures; IMRT/VMAT preferred	40-50
CNS (Intracranial/Dura/Skull Base)	Headache, neurological deficits, seizures, focal symptoms	MRI: Essential for extent; T1 hypointense, T2 hyperintense, enhances with gadolinium	Critical normal tissue constraints for brain, brainstem, optic pathways	40-50
Thyroid	Neck mass, dysphagia, voice changes, compressive symptoms	CT: Homogeneous enhancement; Ultrasound: Solid mass with increased vascularity	Consider proximity to recurrent laryngeal nerves, trachea, esophagus	40-50
Larynx/Throat	Hoarseness, dysphagia, stridor, voice quality changes	CT: Soft tissue mass; MRI: Better soft tissue delineation; Endoscopy: Direct visualization	Voice preservation critical; may require IMRT for organ sparing	40-50
Trachea/Large Airway	Dyspnea, stridor, cough, airway obstruction	CT: Intraluminal or extraluminal mass; Bronchoscopy: Direct assessment of airway involvement	Airway management crucial; may need bronchoscopic debulking first	40-50
Lung (endobronchial/pulmonary)	Cough, dyspnea, chest pain, hemoptysis	CT: Homogeneous soft tissue mass; PET/CT: FDG-avid lesion	Lung dose constraints important; consider respiratory motion	40-50
Mesentery/Abdominal Wall	Abdominal mass, organ compression, back pain	CT: Large heterogenous mass with enhancement; MRI: Better soft tissue contrast	Large treatment volumes; kidney/bowel dose constraints	40-50
Primary Cutaneous/Mucosal	Nodular lesions, ulceration, local mass effect	Clinical exam: Primary assessment; Ultrasound: Depth assessment for treatment planning	Superficial techniques; electron beam or low-energy photons	40-50
Breast (unilateral/bilateral)	Palpable mass, skin changes, nipple discharge	Mammography: Dense mass; MRI: Enhancing lesion; Ultrasound: Solid hypoechoic mass	Consider whole breast vs. partial breast approaches	40-50
Mediastinum/Heart/Pericardium	Chest pain, dyspnea, cardiac symptoms, superior vena cava syndrome	CT: Mediastinal mass; MRI: Cardiac/pericardial involvement; Echo: Functional assessment	Critical cardiac dose constraints; respiratory motion considerations	40-50
Spleen	Abdominal pain, splenomegaly, early satiety, cytopenias	CT: Splenic mass with enhancement; PET/CT: FDG-avid lesion	Splenic dose considerations; may require splenectomy	40-50
Adrenal Glands	Abdominal pain, mass effect, hormonal dysfunction	CT: Enhancing adrenal mass; MRI: Variable signal characteristics	Kidney dose constraints; consider bilateral involvement	40-50
Genitourinary Tract	Hematuria, urinary obstruction, pelvic pain, mass effect	CT/MRI: Enhancing soft tissue mass; Cystoscopy/Ureteroscopy: Direct visualization if applicable	Bladder/bowel sparing; fertility considerations in young patients	40-50
Testis/Scrotum	Scrotal mass, testicular enlargement, pain	Ultrasound: Solid testicular mass; CT/MRI: soft tissue enhancement	Fertility considerations; contralateral testis sparing	40-50

Contd...

**Table 1:** Unusual solitary extramedullary plasmacytoma (SEP) sites (*Continued*).

Site	Presentation	Imaging	Radiation therapy considerations	Dose range (Gy)
Retroperitoneum	Abdominal mass, organ compression, back pain	CT: Large heterogeneous mass with enhancement; MRI: Better soft tissue contrast; PET/CT: Metabolic activity	Large treatment volumes; kidney/bowel dose constraints	39.6-50
Salivary Gland	Painless mass, xerostomia if bilateral involvement	CT/MRI: Well-defined enhancing mass; Sialography: Ductal displacement	Contralateral gland sparing when possible; IMRT beneficial	40-50
Nasopharynx	Nasal obstruction, epistaxis, hearing loss, diplopia	MRI: Gold standard - T1 intermediate, T2 high signal, intense enhancement; CT: Homogeneous enhancement; Endoscopy: Direct visualization	Parotid sparing with IMRT/VMAT; excellent radiosensitivity	42-50 (good outcomes reported with higher doses of up to 67.5)
Nasal Cavity/Sinuses	Epistaxis, nasal obstruction, rhinorrhea, facial pain	CT: Soft tissue mass with possible bone erosion; MRI: Superior soft tissue detail; Endoscopy: Mucosal assessment	IMRT preferred for complex anatomy; optic pathway sparing	>45 preferred
Soft Palate/Oral Cavity	Dysphagia, speech changes, nasal obstruction	CT: Contrast-enhancing mass; MRI: Intense enhancement with gadolinium; Clinical exam: Oropharyngeal obstruction	Swallowing function preservation; consider chemotherapy alternative	Minimum 39.6 Gy

Size-Based Dosing International Lymphoma Radiation Oncology Group (ILROG Guidelines):

- Small lesions (<5 cm): 35-40 Gy acceptable for SBP; 40 Gy minimum for SEP
- Large lesions (≥5 cm): 40-50 Gy recommended
- Post-surgical positive margins: 40-50 Gy

modern imaging shows no evidence of nodal involvement, and especially when the SEP involves Waldeyer's ring structures such as the nasopharynx. Historically, before the advent of conformal RT and advanced imaging techniques, prophylactic cervical lymph node coverage was routinely practiced, with regional nodal failures being rare. Retrospective studies of patients who did not receive elective nodal radiation suggest that nodal recurrence rates remain low (~5%). With advancements in imaging, including MRI and PET/CT, the ILROG panel now recommends against elective lymph node coverage for SEPs unless there is strong clinical evidence of high-risk for nodal involvement, such as very bulky primary disease or proximity to the primary lesion, provided that adding nodal coverage would not significantly increase treatment toxicity.<sup>[3]</sup> Regarding typical radiation portals for localized disease, the target would cover GTV, plus CTV for subclinical microscopic disease around gross tumor of 0.5 cm-1.0cm-3.0 cm, depending on organ involved, and then finally adding a PTV of 5 mm around CTV, using a head and neck (H&N) mask for immobilization and daily reproducibility aided by a CBCT for image guided radiation therapy (IGRT).

The role of chemotherapy for SPs is controversial. It may be considered for SBP, given its high risk of progression to

MM, but there is no established role for chemotherapy in SEPs. A historical prospective study of 53 patients showed improved disease-free survival and overall survival when adding combined melphalan and prednisone to RT for SBP. However, more recent retrospective studies using newer agents have yielded mixed results.

On imaging, nasopharyngeal plasmacytoma can mimic more common entities such as nasopharyngeal carcinoma, lymphoma, or minor salivary gland tumors. MRI typically demonstrates a homogeneously enhancing soft tissue mass with intermediate T1 and T2 signal characteristics. Unlike nasopharyngeal carcinoma, plasmacytoma may show well-defined margins without extensive skull base invasion. PET/CT plays a critical role in excluding systemic disease and assessing for additional hypermetabolic lesions suggestive of multiple myeloma.

Overall, SEPs are highly curable, with progression-free survival rates ranging from 70% to 87% at 10-14 years after treatment with RT (with or without previous resection).<sup>[6-8]</sup> For patients whose SEP shrinks but does not resolve, and where the M protein persists after definitive RT, close follow-up is recommended. Surgery may be considered for SEPs located in accessible sites. Patients with SEP should be followed closely with periodic clinical examinations along with relevant lab work, including Serum Protein

Electrophoresis and Immunofixation Electrophoresis (SPEP/IFE)  $\pm$  free light chains, every 3-4 months for the first 2 years and then every 6-12 months with imaging as indicated, particularly if an M-protein persists.

## CONCLUSION

This case underscores the importance of advanced imaging in the diagnosis, staging, and surveillance of solitary extramedullary plasmacytoma. MRI and PET/CT are critical for local characterization and exclusion of systemic involvement. Early recognition and appropriate imaging-guided treatment can result in excellent local control and favorable outcomes.

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**Declaration of patient consent:** The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given consent for their images and other clinical information to be reported in the journal. The patient understand that the patient's names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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