GLOBAL JOURNAL
OF MEDICAL RESEARCH - K: INTERDISCIPLINARY
Volume 26 | 2026 | Article ID: GJMR-254727 | Language: English

Solitary Plasmacytoma of the Torus Tubarius: A Rare Case Report

Online First

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RECEIVED	ACCEPTED	ONLINE PUBLISHED	PEER REVIEW
2026-03-14	2026-08-03	2026-08-14	Double Blind

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Solitary plasmacytoma is a rare neoplasm characterized by localized proliferation of monoclonal plasma cells and excessive production of the paraprotein M immunoglobulin; it's generally asymptomatic. When plasmacytomas present as isolated lesions, they are referred to as solitary plasmacytomas (SP). Solitary plasmacytoma is distinguished from multiple myeloma by the absence of the International Myeloma Working Group (IMWG)–defined criteria (criteria as > 10% of clonal plasma cells within bone marrow or biopsy-proven bony or EMP, and at least 1 myeloma-defining event). We report an extremely rare case of a 59-year-old woman who presented to the emergency department of our hospital in Chivasso, in the province of Turin, for unrelated reasons. During flexible fiberoptic endoscopic evaluation, an incidental pedunculated mass arising from the inferior portion of the right torus tubarius was identified.

Solitary Plasmacytoma | Extramedullary Plasmacytoma | Torus Tubarius | Nasopharyngeal Neoplasm | Plasma Cell Disorder | Immunohistochemistry | Radiotherapy

AI USE STATEMENT
No generative AI was used for analysis or results.

FUNDING
No external funding was declared for this work.

CONFLICT OF INTEREST
The authors declare no conflict of interest.

DATA AVAILABILITY
Not applicable for this article.

ETHICS
No ethics committee approval was required for this article type.

CONSENT
Not applicable for this article.

TRIAL REG.
Not applicable.

Crossref DOI: 10.34257/GJMRK254727

How to Cite: Briguglio (2026). Solitary Plasmacytoma of the Torus Tubarius: A Rare Case Report. Global Journal of Medical Research, Ahead of Print, 1-15. DOI: 10.34257/GJMRK254727

AR Experience Web Page DOI

Print ISSN 0975-5888

Online ISSN 2249-4618

Under the strict compliance and defined process of

METADATA CONTINUATION

AUTHOR CONTACT QR LEDGER

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ARCHIVAL RECORD

GJMR · Vol 26 · 2026

Article ID GJMR-254727 · DOI 10.34257/GJMRK254727

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Global Journal of Medical Research - Section K
GJMR

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Keywords: *Solitary Plasmacytoma, Extramedullary Plasmacytoma, Torus Tubarius, Nasopharyngeal Neoplasm, Plasma Cell Disorder, Immunohistochemistry, Radiotherapy*

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1. Introduction

Plasmacytomas are rare tumors characterized by the monoclonal proliferation of plasma cells, which may localize either in bone or in soft tissues, generally without causing systemic manifestations, and they account for approximately 6% of all plasma cell disorders.

Plasmacytomas are classified into two categories—multiple or solitary—according to the number of detectable lesions, with a higher tendency toward progression to multiple myeloma in multiple forms. Solitary plasmacytomas are further subdivided based on their osseous or extramedullary localization.

The incidence of multiple myeloma has increased in recent years, with a concomitant rise in the occurrence of extramedullary plasmacytoma and solitary plasmacytoma, particularly among the elderly population. Although the pathogenesis of extramedullary plasmacytoma remains incompletely understood, several factors—including genetic predisposition, viral infections, and exposure to ionizing radiation—have been proposed as potential contributors.

Approximately 80% of extramedullary plasmacytomas arise in the head and neck region, with the nasal cavity and oropharynx among the most commonly involved sites. The patients can present symptoms such as headache, dysphagia, sore throat, epistaxis, or nasal obstruction. Extramedullary plasmacytomas involving the larynx, thyroid gland, parotid gland, or middle ear are considered particularly rare. The involvement of these uncommon regions like the larynx can cause dysphonia, dysphagia, wheezing, and airway obstruction.

1.1. Case Presentation

We report the case of a 59-year-old woman who presented to the emergency department of our hospital in Chivasso, in the province of Turin, complaining of a foreign body sensation in the pharyngeal region. The patient did not report any additional symptoms, including pharyngeal pain or odynophagia. She reported good general health, was not receiving any chronic medical therapy, and had no known allergies. Her only previous surgical procedure was uterine curettage following a spontaneous abortion approximately 20 years earlier.

On otorhinolaryngological examination, oropharyngoscopy was unremarkable; however, flexible fiberoptic endoscopic evaluation revealed a pedunculated mass in the nasopharynx, specifically arising from the distal portion of the right torus tubarius. A contrast-enhanced computed tomography (CT) scan of the maxillofacial region and neck was therefore requested and performed five days after the initial clinical evaluation.

CT imaging demonstrated asymmetry of the oropharyngeal air spaces related to the presence of an ovoid lesion located slightly inferior to the right torus tubarius, measuring approximately 10 × 11×7 mm. The lesion showed intense contrast enhancement, a broad base of attachment to the right oropharyngeal mucosa measuring approximately 1 cm, and a plunging appearance into the airway spaces, resulting in partial obliteration on the right side. No clear infiltrative features were observed, particularly with respect to the deep fat planes and the retropharyngeal space.

Following otorhinolaryngological reassessment and review of the CT findings, the patient was placed on the waiting list for surgical excision of the lesion. Preoperative laboratory tests were within normal limits, as was chest radiography. Approximately one month after the last ENT evaluation, the patient underwent surgery under general anesthesia. After placement of a self-retaining mouth gag, transoral excision of the lesion along with its pedicle was performed under endoscopic guidance using a 70° rigid endoscope, and the specimen was submitted for histopathological examination.

Histological analysis revealed a thin squamous epithelium covering a densely cellular lesion, vaguely subdivided into nests by thin fibrous septa and traversed by a rich vascular network with variably sized and shaped lumina and thin walls. The cellular component consisted predominantly of mature plasma cells with abundant cytoplasm and round eccentric nuclei, along with a smaller

proportion of more immature cells with small nucleoli. Immunohistochemical staining demonstrated diffuse expression of vimentin and CD31, extensive but not complete positivity for CD138, CD79α, and kappa light chain, and negativity for CD34, CD45, pan-cytokeratin (AE1/AE3), CD99, cyclin D1, smooth muscle actin, desmin, calponin, S100, synaptophysin, CD20, PAX5, and lambda light chain. Rare scattered CD45-positive lymphocytes were observed. The proliferative index assessed by MIB-1 was approximately 2%.

Based on these findings, the histopathological features were considered most consistent with a diagnosis of solitary plasmacytoma.

The patient was subsequently referred to the hematology team. A bone marrow biopsy was performed and yielded negative results, and a PET scan was carried out, which showed disease with high metabolic activity in the right nasopharyngeal region; therefore, she underwent radiotherapy to this area.

2. Discussion

Solitary plasmacytoma is a rare neoplasm characterized by the proliferation of plasma cells within a single anatomical site, such as bone or soft tissues, without evidence of bone invasion or systemic symptoms.

Soft tissue plasmacytomas are less prevalent than osseous forms, accounting for approximately 30% of all cases reported in the literature. Extramedullary plasmacytomas predominantly occur in adulthood, with a higher incidence between 50 and 65 years of age.

Although they may affect younger individuals, they are considered rare before the age of 40–50 years. They occur more frequently in men than in women, with an estimated male-to-female ratio of approximately 3:1.

The diagnosis of extramedullary disease in multiple myeloma can be very difficult, and there may be delays due to varying locations or subtle presentations.

Extramedullary plasmacytomas may arise in a wide range of soft tissues, often producing nonspecific symptoms that can delay diagnosis and, consequently, postpone the initiation of appropriate treatment.

The symptoms that arise may be attributed to other conditions, which can increase the risk that patients delay seeking evaluation by a hematologist.

The head and neck region represents the most common site of involvement for these tumors (about 80% of all cases).

The sinonasal cavity is the most common site of EMP in the upper aerodigestive tract (38% occur within this subsite); when occurring at this site, they enter the differential diagnosis with lymphomas, melanomas, inverted papillomas, nasal polyps, or fungal rhinosinusitis.

They typically present with symptoms such as nasal obstruction or epistaxis.

At the nasopharyngeal level, they usually appear as slowly growing masses and are generally associated with a favorable prognosis.

Plasmacytoma shares cellular type and morphological features with multiple myeloma; however, it does not exhibit the systemic manifestations typical of advanced disease.

A high histological grade and the presence of angiogenesis in plasmacytomas are associated with an increased risk of progression to multiple myeloma (estimated to range between 10% and 30%).

Extramedullary plasmacytoma is occasionally regarded as an intermediate stage between monoclonal gammopathy of undetermined significance (MGUS) and overt multiple myeloma.

In order to diagnose solitary plasmacytoma, multiple myeloma must be excluded, as up to 60% of solitary plasmacytomas are treated with local therapy alone, whereas the 5-year survival rate for multiple myeloma is approximately 35%.

The diagnostic criteria for solitary extramedullary plasmacytoma include histologically confirmed plasma cell neoplasia, less than 5% plasma cells on bone marrow biopsy from a separate site and the absence of myeloma-related end-organ damage.

Biopsy is essential to assess plasma cell infiltration and characterize the cellular phenotype. Histopathological evaluation confirms the presence of plasma cells, while immunohistochemistry (IHC) offers crucial information regarding cellular phenotype and clonality. Key IHC markers, including CD138, CD38, and MUM1, are routinely employed to establish the plasma cell origin of the lesion.

Even a complete skeletal survey to exclude intramedullary involvement is fundamental: current recommendations include the use of whole-body imaging modalities, such as low-dose computed tomography (CT), magnetic resonance imaging (MRI), or ^{18}F-FDG PET/CT (18-fluorodeoxyglucose positron emission tomography–CT), in accordance with local guidelines and interpreted alongside clinical and radiological findings.

PET-CT scans are a highly effective tool for identifying the localization of EMD lesions and assessing metabolic response to therapy; MRI is a very important technique used for detecting lesions in soft tissue caused by extramedullary plasmacytoma. Instead, CT helps assess bone lesions and is frequently used to evaluate MM cases. A better option is whole-body low-dose CT (WBLD-CT), which allows for the evaluation of the risk of pathological fractures.

In the present case, computed tomography was useful in defining the lesion's characteristics—particularly its intense contrast enhancement—and in excluding infiltration of deeper tissues.

Treatment options for solitary plasmacytoma include surgical resection, definitive radiotherapy, or surgery followed by adjuvant radiotherapy.

Radiotherapy is a treatment of choice and is frequently used in extramedullary disease. It achieves local control rates of approximately 80% in both solitary bone plasmacytomas and extramedullary plasmacytomas due to the radiosensitive nature of these lesions.

Surgical excision, either partial or complete, may be considered in cases of extramedullary or solitary bone plasmacytomas. Surgery is frequently performed when a newly identified tumor mass requires histopathological evaluation.

Chemotherapy is reserved for high-grade plasmacytomas or for extensive disease, primarily to reduce the risk of recurrence.

3. Conclusions

Although extremely rare, nasopharyngeal plasmacytoma should be considered in the differential diagnosis of other neoplasms, such as squamous cell carcinomas and lymphomas. Careful histopathological analysis is crucial for accurate diagnosis and appropriate management. In this context, close collaboration with a hematologist remains essential for optimal patient care.

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