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# Unusual presentation of a soft palate mass: A rare case report of solitary extramedullary plasmacytoma

B. Merzouqi[a,*], S. Halily[a], Y. Oukessou[a], M. Regragui[b], R. Abada[a], M. Mahtar[a]

[a] Department of Oto-Rhino-Laryngology, Head and Neck Surgery, Casablanca University Hospital, Casablanca, Morocco
[b] Pathology Department, Casablanca University Hospital, Casablanca, Morocco

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ABSTRACT

*INTRODUCTION:* Plasmacytoma is a rare clonal neoplastic disorder of bone marrow that originates from plasma cells. It usually presents as a multiple myeloma (MM). Less than 5% of patients present with either a single bone lesion as a solitary bone plasmacytoma (SBP) or, even more rarely, as a soft tissue mass of monoclonal plasma cells representing a solitary extra medullary plasmacytoma (SEP).
*CASE PRESENTATION:* We report a case of a 59-year-old man presenting with a mass of the soft palate evolving for a year. Physical examination showed an extension to the nasal cavity. Biopsy with immunohistochemical study demonstrated sheets of mononucleated plasmacytoid cells diffusely expressing CD138. The plasma cells showed monoclonal light chain Kappa. Further investigations did not show any other locations including bone and bone marrow. Thus, diagnosis of solitary extramedullary plasmacytoma of the soft palate was established. The patient was treated with chemotherapy with total remission on his one year follow-up.
*DISCUSSION:* SEP may arise in any organ, either as a primary tumor or as part of a MM. Almost 90% of SEP arise in the head and neck, especially in the upper respiratory tract. Primary treatment for most patients is radiotherapy, but surgery may also be required, and multidisciplinary decision between surgeon, hematologist and radiotherapist is crucial for planning optimum care.
*CONCLUSION:* SEP is an extremely rare condition which requires diagnostic and therapeutic management in the same level of MM. Prognosis is better than the two other forms (MM and SBP).


## 1. Introduction

Plasmacytoma is a rare clonal neoplastic disorder of bone marrow that originates from plasma cells which is the last maturation stage of B lymphocytes [1]. It usually presents as a multiple myeloma (MM: systemic disease with multiple bone localizations). However, less than 5% of patients present with either a single bone lesion as a solitary bone plasmacytoma (SBP) or, even more rarely, as a soft tissue mass of monoclonal plasma cells representing a solitary extramedullary plasmacytoma (SEP). Clonal plasma cells involved in plasmacytoma frequently produce a monoclonal immunoglobulin as well as kappa or lambda free light chains [2,3]. We report a case of a 59-year-old man presenting with a solitary extramedullary plasmacytoma of the soft palate treated by chemotherapy with good outcomes. This case report has been reported in line with the SCARE Criteria [18].

## 2. Case presentation

A 59-year-old man with no medical history, consulted in the otolaryngology clinic for dysphagia to solids and breathing difficulty evolving for a year with no significant weight loss. The patient is a non-smoker and does not use alcohol or drugs.

Physical examination revealed a mass of the soft palate obstructing the oropharynx (Fig. 1). Rhinoscopy showed a cystic mass in both nasal cavities, obstructing both choanae, arriving to the middle turbinate (Fig. 2). He had no palpable cervical lymph nodes.

CT scan revealed a contrast-enhancing mass of the soft palate filling the nasopharynx lumen without invading its walls or the deep spaces of the face (Fig. 3).

MRI showed a 62 × 34 × 40 mm tumor of the nasopharynx, extending to the oropharynx which was partially obstructed. The mass did not invade the skull base. It was intensely enhanced after gadolinium injection (Fig. 4).

Pathology report of the biopsy with immunohistochemical study demonstrated sheets of mononucleated plasmacytoid cells with perivascular pink (Fig. 5) with amorphous deposits plasmacytoid cells diffusely expressing CD138 (clone MI15) (Figs. 5 and 6). The plasma cells showed monoclonal light chain Kappa (Polyclonal antibody) (Fig. 7).

* Corresponding author at: 6, Rue Lahcen Al Aarjoune, Casablanca, 20250, Morocco.
*E-mail address:* merzouqiboutaina@gmail.com (B. Merzouqi).

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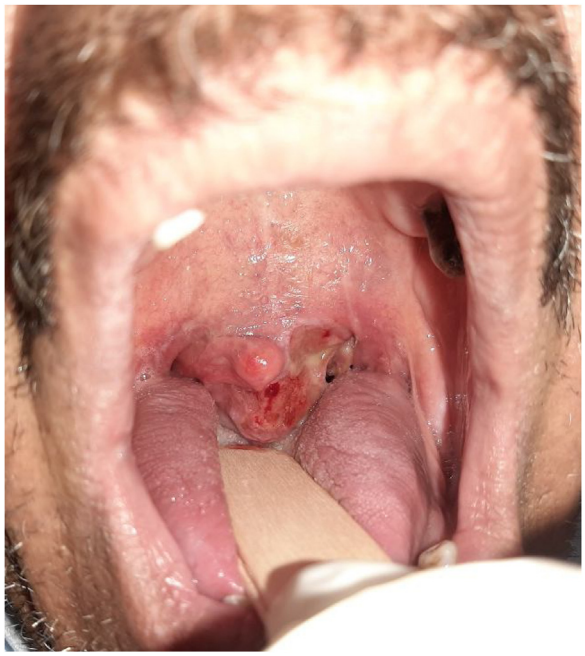

**Fig. 1.** Clinical aspect of the soft palate tumor arriving to the base of the tongue.

We performed other investigations which did not reveal any abnormalities. The patient had a normal examination of his bone marrow biopsy. Bone radiographies did not reveal any suspicious lesions. Urine test did not show Bence Jones protein. He also had a normal blood count, normal calcium level and renal function. No monoclonal immunoglobulins were detected in his blood. PET scan did not reveal any other localizations. Thus, the diagnosis of multiple myeloma is then ruled out and that of a solitary extramedullary plasmacytoma of the soft palate extending to the nasal cavity and the oropharynx was established.

The patient received chemotherapy as he refused the surgery as a mutilating procedure. He was disease free on his one-year follow-up with no significant complications or side effects.

## 3. Discussion

Solitary plasmacytoma (SP) is a solitary lesion without clinical, histological or radiological evidence of MM. It is subdivided into two entities (SBP and SEP) depending on whether the lesion originates from the bone or soft tissue [4]. It may be isolated or the first manifestation of a subsequent systemic disease (MM) [3,5]. The incidence of SBP is approximately 40% higher than SEP [6]. The isolated form (SBP or SEP) of plasmacytoma seems to have a better prognosis compared to the systemic one (MM) [3,5,7]. The median age at diagnosis of SP is 55–60 years, significantly lower than in MM patients, the male to female ratio varies from 1,2:1 to 2:1 [4,6,8].

SEP may arise in any organ, either as a primary tumor or as part of a MM. It is usually associated with a better prognosis and a higher cure rate compared to SBP. Almost 90% of SEP arise in the head and neck, especially in the upper respiratory tract (nasal cavity, sinuses, oropharynx, salivary glands and larynx) followed by the second most frequent site which is the gastrointestinal tract [9,10].

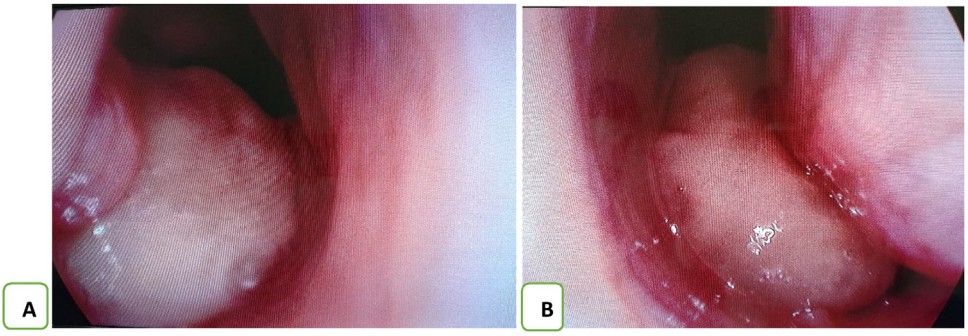


**Fig. 2. A)** Soft tissue mass invading the right nasal cavity. **B)** Soft tissue mass invading the left nasal cavity.

**Fig. 3.** Sagittal images of CT scan showing a mass of the soft palate filling the oropharynx and nasopharynx lumen without invading its walls.

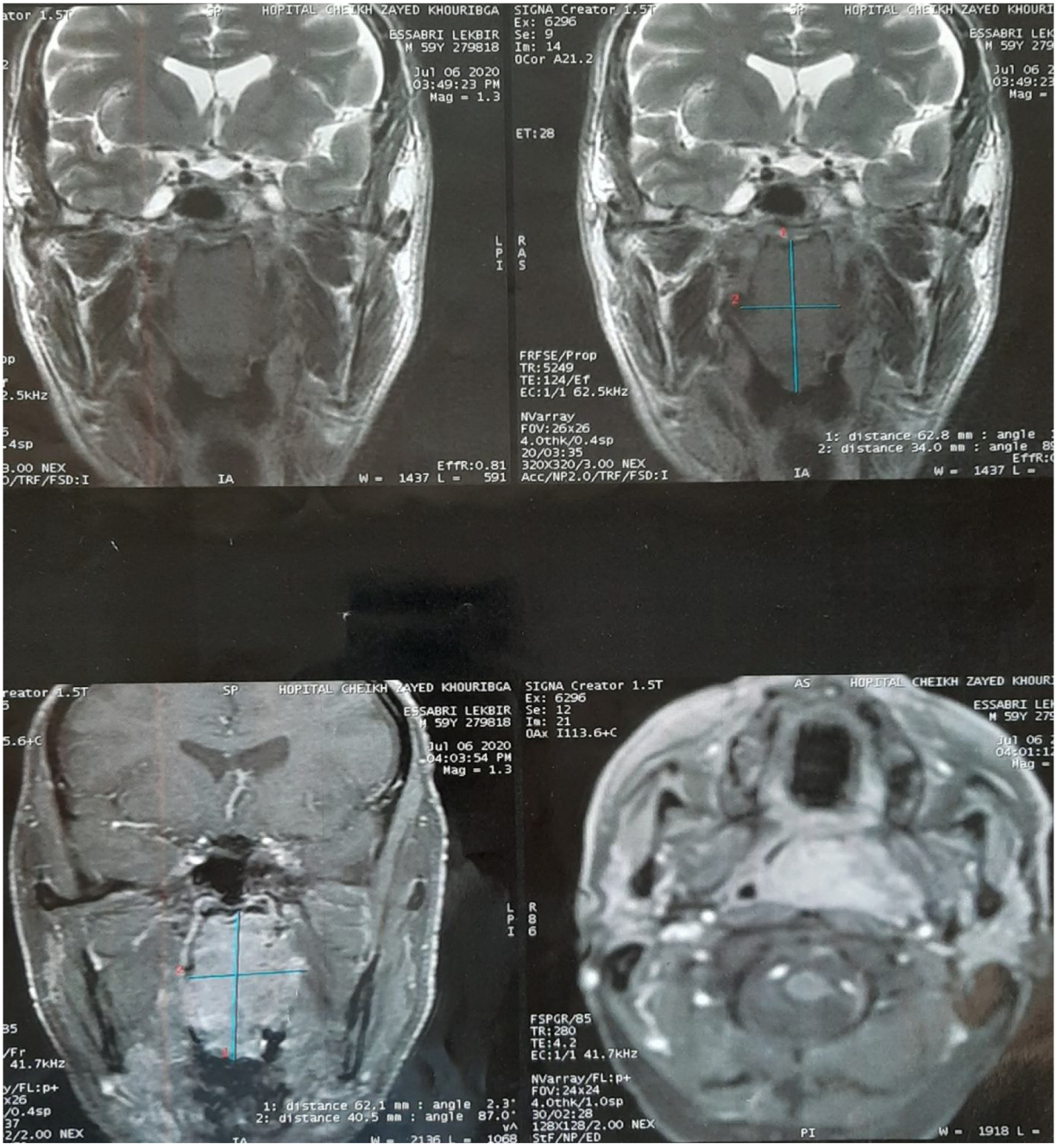


**Fig. 4.** MRI images showing an intensely enhanced tumor of the nasopharynx, extending to the oropharynx which was partially obstructed without the skull base invasion.

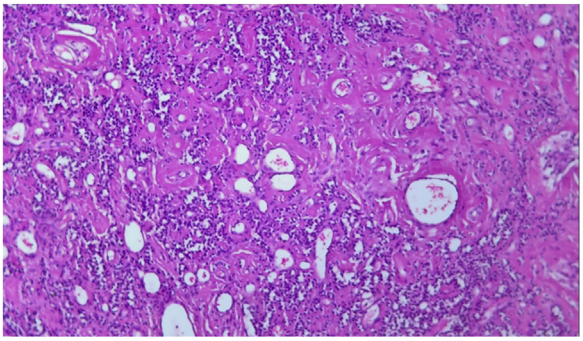

**Fig. 5.** HEx100: Sheets of mononucleated plasmacytoid cells with perivascular pink amorphous deposits.

Other sites may rarely be involved, such as bladder, urethra, breast, ovary, lung, pleura, thyroid, orbit, brain and skin [10–13].

Diagnostic criteria of a SEP include a solitary lesion, histopathological confirmation, negative bone marrow examination or clonal plasma cells infiltration less than 10% of all nucleated cells, normal results on skeletal survey, including radiology of long bones or negative PET CT, negative urine test for Bence Jones protein, absence of anemia, hypercalcemia or renal impairment due to plasma cell dyscrasia, absent or low serum or urinary level of monoclonal immunoglobulins [10,14,15]. Our case meets all these criteria for the diagnosis of SEP of soft palate.

The diagnosis and management of patients with solitary plasmacytoma requires the same range of clinical and laboratory expertise as for patients with multiple myeloma [16]. The primary treatment for most patients is radiotherapy, but surgery may

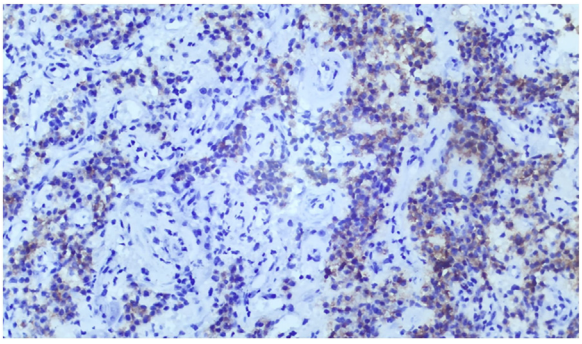

**Fig. 6.** Plasmacytoid cells diffusely expressing CD138 (clone MI15).

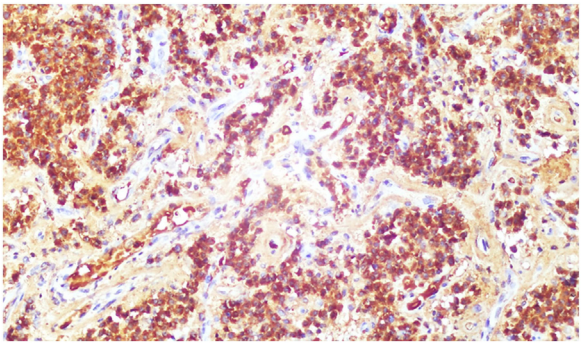

**Fig. 7.** The plasma cells show monoclonal light chain Kappa (Polyclonal antibody).

also be required, and multidisciplinary decision between surgeon, hematologist and radiotherapist is crucial for planning optimum care [10].

In a curative goal, radical surgery is a mutilating procedure which is not indicated as the tumors of head and neck are generally highly radiosensitive and most patients are cured with radiotherapy. Local control rates of 80–100% are consistently reported with moderate doses of radiotherapy [17]. Adjuvant chemotherapy is preferred in patients with lymphadenopathy or with radiation resistant tumors [10]. In our case, the patient chose to have chemotherapy instead of other treatment options discussed.

SBP has a high risk of progression to MM. Conversely, SEP is almost always localized and has a high cure rate with local treatment [10].

## 4. Conclusion

To our knowledge, SEP is an extremely rare condition which requires diagnostic and therapeutic management in the same level of MM. Prognosis is better than the two other forms (MM and SBP) and radiotherapy remains the preferred option according to the literature. Due to the rarity of this disease, efforts should be made to determine which treatment should be given to which patient.

## Declaration of Competing Interest

None.

## Funding

None.

## Ethical approval

The study is exempt from ethical approval in our institution as it is a “Case report” and not a research study.

## Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

## Registration of research studies

Not Applicable.

## Guarantor

B. Merzouqi.

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## CRediT authorship contribution statement

**B. Merzouqi:** Investigation, Resources, Writing - original draft, Writing - review & editing, Visualization. **S. Halily:** Investigation, Resources, Writing - review & editing. **Y. Oukessou:** Writing - review & editing. **M. Regragui:** Investigation, Resources. **R. Abada:** Validation, Supervision. **M. Mahtar:** Validation, Supervision.

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