

Mediastinal Extramedullary Plasmacytoma: A Case Report and Literature Review

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DOI: <https://doi.org/10.36347/sjmcr.2026.v14i09.027>

| Received: 17.07.2026 | Accepted: 02.09.2026 | Published: 22.09.2026

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Abstract

Case Report

Introduction: Extramedullary plasmacytomas (EMP) correspond to plasma cell tumor localizations outside the bone marrow. They can present as a localized solitary form or as a manifestation of systemic multiple myeloma [1]. Their mediastinal location remains exceptional [2]. Diagnosis is often challenging, particularly when conventional biological markers of multiple myeloma are absent. For localized forms, radiotherapy constitutes the reference therapeutic option [3]. We present a case of EMP developing in the anterior mediastinum and propose, through a literature analysis, a reflection on management strategies in radiation oncology. **Case Report:** A 32-year-old female patient, with no notable medical history, consulted for progressive onset of dry cough and dyspnea. Chest imaging revealed a large anterior mediastinal mass measuring 12.4 cm with displacement of mediastinal structures. Histological examination of CT-guided biopsy revealed morphological and immunohistochemical features consistent with a plasma cell neoplasm. Laboratory tests showed 2% plasma cells on bone marrow aspirate. The diagnosis of solitary mediastinal extramedullary plasmacytoma was retained. The patient was referred to a medical oncology department for discussion of therapeutic options. **Conclusion:** Radiotherapy plays a predominant role in the treatment of solitary EMP, with excellent local control rates [4]. Nevertheless, the distinction between solitary EMP and EMP associated with systemic multiple myeloma is fundamental, as it guides management toward radically different approaches [5]. Serum free light chain assay is an essential diagnostic tool, particularly when serum protein electrophoresis shows no abnormality [6].

Keywords: Extramedullary plasmacytoma, mediastinum, radiotherapy, multiple myeloma, free light chains.

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INTRODUCTION

Plasma cells are terminally differentiated B-lineage cells responsible for immunoglobulin production. Their abnormal monoclonal proliferation gives rise to a spectrum of diseases grouped under the term monoclonal gammopathies, ranging from monoclonal gammopathy of undetermined significance (MGUS) to multiple myeloma, including solitary bone or extramedullary plasmacytomas [1].

Extramedullary plasmacytoma (EMP) corresponds to a monoclonal plasma cell tumor localization outside the bone marrow [1]. It can present in two forms:

- **Solitary form:** unique tumor without significant marrow involvement (< 10% clonal plasma cells) nor other signs of multiple myeloma [1];

- **A manifestation of multiple myeloma:** presence of extramedullary plasma cell lesions in a patient meeting diagnostic criteria for multiple myeloma (marrow plasmacytosis $\geq$ 10%, bone lesions, monoclonal peak, etc.) [1].

EMPs are rare tumors, representing only 3% of malignant plasma cell proliferations [2]. They predominantly affect males (sex ratio 3:1), with a median age of 55 years [2]. Although more frequent in men, EMPs can also occur in women, as in our observation.

Mediastinal topography is particularly rare. Indeed, more than 80% of EMPs develop in the upper aerodigestive tract, preferentially affecting the nasopharynx, paranasal sinuses, or nasal cavity [2]. Localizations in the lower respiratory tract and mediastinum are exceptional [3]. Less than 15% of EMPs occur outside the ENT sphere [2].

Citation: Ennadif, A., Mimouni, I., Ennafaa, S., Limouni, N., Touzri, S., Hamidi, F., Zaggha, N., Benlamlih, M., Hommadi, M., Marnouch, E. A., Maghous, A., Bazine, A., Lalya, I., Andaloussi, K., Haddadi, K., & El Marjany, M. (2026). Mediastinal Extramedullary Plasmacytoma: A Case Report and Literature Review. *Sch J Med Case Rep*, 14(9), 2071-2074. <https://doi.org/10.36347/sjmcr.2026.v14i09.027>

The reference treatment for localized EMP is radiotherapy, which achieves optimal local control with doses of 40 to 50 Gy [4]. Exclusive radiotherapy offers curative prospects in extramedullary forms, with excellent local control rates [4].

However, the distinction between solitary EMP and EMP associated with active multiple myeloma is of paramount importance because therapeutic approaches diverge fundamentally [3]. While solitary EMP requires curative local irradiation, multiple myeloma mandates systemic management with chemotherapy or targeted agents [5]. Recognition of EMP associated with active multiple myeloma is crucial to avoid missing systemic disease [3].

Serum free light chain (FLC) assay is a major diagnostic tool in this context, particularly when serum protein electrophoresis is normal [6]. The combination of this assay with electrophoresis and immunofixation allows optimal detection of monoclonal gammopathies [6].

We report the case of a 32-year-old female patient with a solitary anterior mediastinal EMP, and we discuss, through a literature review, the diagnostic and therapeutic challenges.

CASE REPORT

Patient and Context

A 32-year-old female patient, with no notable comorbidities, was seen in pulmonology consultation for evaluation of progressive exertional dyspnea and persistent dry cough for two months. She reported no fever, significant weight loss, or bone pain.

Clinical Examination Findings

Physical examination was unremarkable, apart from superficial tachypnea. There was no focal neurological deficit. Peripheral lymph node areas were free. There was no hemorrhagic syndrome or palpable hepatosplenomegaly.

Diagnostic Workup

Chest radiography showed mediastinal widening and a large pleural effusion. Chest CT scan revealed a large anterior mediastinal mass measuring 12.4 cm, displacing mediastinal structures to the right and causing collapse of the left lung and a large reactive pleural effusion.

A CT-guided percutaneous biopsy of the mediastinal lesion was performed. Histopathological examination revealed an atypical plasma cell proliferation, with CD138 and CD56 expression on immunohistochemistry, consistent with a plasmacytoma.

Laboratory tests showed:

- Normal serum protein electrophoresis, without monoclonal peak;
- Balanced bone marrow aspirate with 2% plasma cells (normal rate).

PET-CT showed FDG-avid uptake of the large mediastinal mass, with no other suspicious foci.

The diagnosis of solitary mediastinal extramedullary plasmacytoma was retained.

Therapeutic Management

Given the diagnosis of solitary EMP, the patient was transferred to a specialized onco-hematology center to discuss therapeutic options. Radiotherapy was proposed as the reference treatment but was not performed due to the large size of the lesion (12.4 cm), making it difficult to respect dose constraints to organs at risk (lung and heart). Systemic chemotherapy was initiated.

Patient Outcome

Following transfer to the specialized onco-hematology department, the patient initiated systemic chemotherapy. However, she did not return for follow-up after the first cycle and was subsequently lost to follow-up. No further clinical or radiological data are available.

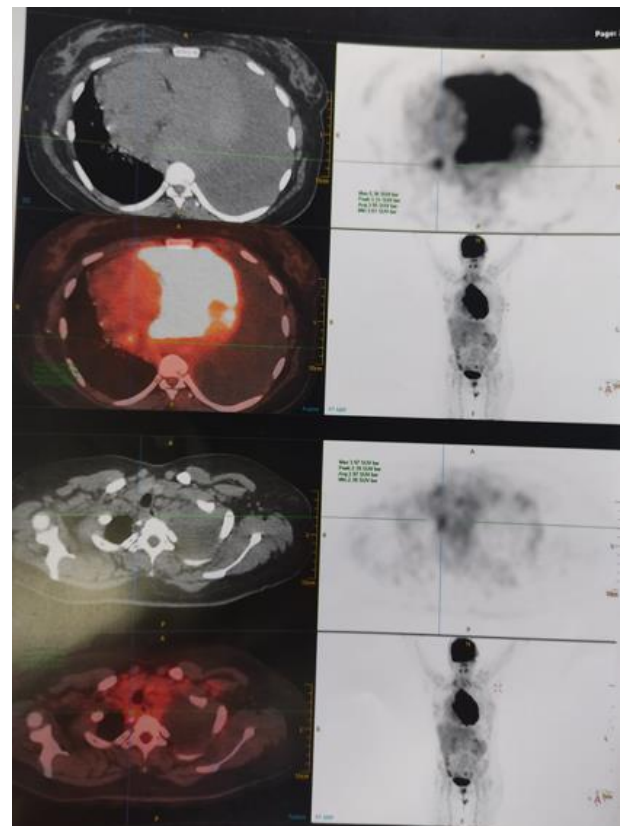


Figure 1: PET/CT imaging showing a large solitary anterior mediastinal mass with intense FDG uptake, without any other suspicious distant foci

DISCUSSION

Epidemiological Data

EMPs are rare tumors, representing only about 3% of plasma cell neoplasms [2]. They predominate in males with a sex ratio of 3:1, and most commonly occur between 50 and 60 years of age [7]. In our observation, the age of 32 years is younger than the median age reported in the literature (55 years), but remains within the range of described cases [7]. The female sex makes this case rarer, although EMPs can occur in both sexes.

Mediastinal localization is exceptional [3]. As highlighted by Alexiou *et al.*, more than 80% of EMPs develop in the upper aerodigestive tract [2]. Yamaguchi *et al.*, described a case of anterior mediastinal EMP in an 80-year-old man, successfully treated with irradiation [7]. Lee *et al.* reported a posterior mediastinal EMP in a 64-year-old man, initially mistaken for a neurogenic tumor, and treated with local radiotherapy [8]. Yilmaz *et al.* also published a comparable case in a 54-year-old female patient, confirming the rarity of this topography [9].

Diagnostic Challenges

EMP falls within the spectrum of monoclonal gammopathies. The diagnosis of solitary EMP rests on three essential elements: histological evidence of monoclonal plasma cell proliferation, absence of significant medullary plasmacytosis (less than 10%), and absence of multiple myeloma criteria (anemia, renal failure, lytic bone lesions, serum or urinary monoclonal peak) [1].

In our observation, all criteria for solitary EMP are met:

- **Histology:** plasma cell proliferation with CD138 and CD56 expression;
- **Bone marrow aspirate:** 2% plasma cells, well below the 10% threshold;
- **Absence of multiple myeloma criteria:** normal electrophoresis, no lytic bone lesions, no anemia or renal failure.

Serum free light chain assay, although useful in this context [6], was not performed in our patient, as all diagnostic criteria were already in favor of solitary EMP. Nevertheless, this assay could be discussed to refine the initial workup and formally exclude light chain multiple myeloma [6].

The distinction between solitary EMP and multiple myeloma with extramedullary localization is crucial because therapeutic strategies are radically opposed [3]. As noted by Proskuriakova *et al.*, the prognosis is significantly more favorable in the absence of associated multiple myeloma [10].

It is important to note that extramedullary plasmacytomas can also appear as disease progression in patients already treated for multiple myeloma,

corresponding to an aggressive form resistant to systemic treatments, often associated with a more reserved prognosis [11].

Role of Radiotherapy

Radiotherapy represents the reference treatment for solitary EMPs [4]. Due to their radiosensitivity, doses of 40 to 50 Gy achieve very satisfactory local control rates, with approximately 70% of patients in remission at 10 years [1].

In the literature, several observations confirm the efficacy of radiotherapy in mediastinal EMPs. Yamaguchi *et al.* reported significant tumor reduction and normalization of serum IgA after irradiation of an anterior mediastinal EMP in an 80-year-old patient [7]. Lee *et al.* described favorable outcomes after local radiotherapy for a posterior mediastinal EMP [8]. Yilmaz *et al.* planned radiotherapy after diagnostic confirmation in a 54-year-old patient [9]. Quilodrán *et al.* also reported a case of mediastinal EMP successfully treated with irradiation [12].

In our observation, radiotherapy was proposed as the reference treatment but could not be performed due to the large size of the tumor (12.4 cm), making it difficult to respect dose constraints to organs at risk (lung and heart). A therapeutic alternative with chemotherapy was considered.

Prognosis and Follow-up

The prognosis of solitary EMPs is generally good, with approximately 70% of patients in remission at 10 years [1]. However, transformation to multiple myeloma is observed in 10 to 30% of cases [1].

As highlighted in the 2025 review on solitary plasmacytomas, these rare tumors carry a risk of transformation to multiple myeloma, requiring optimal management and meticulous follow-up [13].

Prolonged surveillance is imperative, combining regular clinical examination, serum protein electrophoresis, free light chain assay, and imaging controls [6].

Limitations of the Case Report

Our observation has several limitations. It is a single case report, and the patient was lost to follow-up after initiating chemotherapy, preventing any assessment of treatment response, disease evolution, or long-term outcomes. Nevertheless, this case illustrates the rarity of mediastinal EMPs and the therapeutic challenges posed by bulky tumors, particularly in young patients where organ-at-risk sparing is critical.

CONCLUSION

EMP falls within the spectrum of monoclonal gammopathies, which can be a localized form or a

manifestation of systemic multiple myeloma. Mediastinal EMP is a rare tumor whose diagnosis requires a multidisciplinary approach combining imaging, pathology, and laboratory workup. Radiotherapy remains the reference treatment for solitary forms, offering excellent local control at doses of 40 to 50 Gy. The search for associated multiple myeloma is an indispensable prerequisite before initiating exclusive irradiation. Serum free light chain assay is an essential diagnostic tool, particularly when serum protein electrophoresis is normal. The prognosis is generally favorable for solitary forms, but prolonged clinical and biological follow-up is necessary to detect early any evolution toward systemic disease.

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