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(CASE REPORT)

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# Intra-abdominal extramedullary plasmacytoma: A case report and literature review

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## Abstract

The incidence of extramedullary plasmacytoma (EMP) accounts for 3% to 4% of all cases of generalized plasmacytoma. Extramedullary plasmacytomas are predominantly located in the upper respiratory tract and nasopharynx, with only around 4% originating primarily in the gastrointestinal tract. The median age at diagnosis is approximately 55 years about 10 years earlier than in multiple myeloma and there is a higher incidence among males. These tumors exhibit a diverse phenotype that can result in varying degrees of mass effect and organ dysfunction based on their anatomical site. Although they display immunophenotypic characteristics similar to multiple myeloma, obtaining an in vivo tissue diagnosis can be particularly challenging for lesions situated within the abdominal cavity. Consequently, accurate diagnosis and effective treatment planning require a collaborative, multidisciplinary approach involving surgeons, hematologists, oncologists, radiologists, and pathologists.

We report the case of a 50 year old woman who presented an abdominal distension without any associated signs. An abdominal CT scan was performed who objectified a mass in the left hypochondrium and flank of 12 centimeters followed by a biopsy and a full blood test, which confirmed the diagnosis of extramedullary plasmocytoma kappa monotype. Surgery was not feasible, radiotherapy was thus carried out. After a follow up of 18 months, a TEP scan was performed showing the absence of any lesion suspected of progressive disease or residual metabolically active disease.

**Keywords:** Extramedullary Plasmocytoma; Intra-abdominal Plasmocytoma; Radiotherapy of Plasmocytoma; Plasmocytoma Kappa Monotype

## 1. Introduction

Extramedullary plasmacytoma (EMP) is a rare plasma cell neoplasm that most commonly affects the head and neck region, with gastrointestinal involvement occurring in less than 5% of cases. Typically diagnosed at around 55 years of age with a male predominance, EMP often presents with nonspecific symptoms related to local mass effects. Diagnosis relies on advanced imaging and histopathological confirmation of clonal plasma cells. Management usually requires a multidisciplinary approach, combining surgery, radiotherapy, and systemic chemotherapy. This case report, accompanied by a review of the literature, highlights current diagnostic and therapeutic challenges in the management of EMP.

## 2. Case Presentation

A 50-year-old female patient (7G-7P) with a pathological history of treated hepatitis B and arterial hypertension on medication presented with an abdominal distension without any associated signs. The patient had no history of

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malignancy or family history of haematological cancer. Physical examination found an abdominal mass in the flank and left hypochondrium. No inguinal nodes were palpable.

An abdomino-pelvic scan was performed showing a mass located in the left hypochondrium and flank, well limited, oval in shape, well circumscribed, hypodense, containing a few parietal calcifications, measuring 125 x 85 x 84 mm. Topographically: this mass is in intimate contact with the wall of the greater gastric curve and the tail of the pancreas with no separating fatty border. Externally, it comes into close contact in places with the spleen. Posteriorly, it displaces the left kidney, with loss of the separating border in places. Internally, it comes into close contact with the left renal vein, which remains permeable.

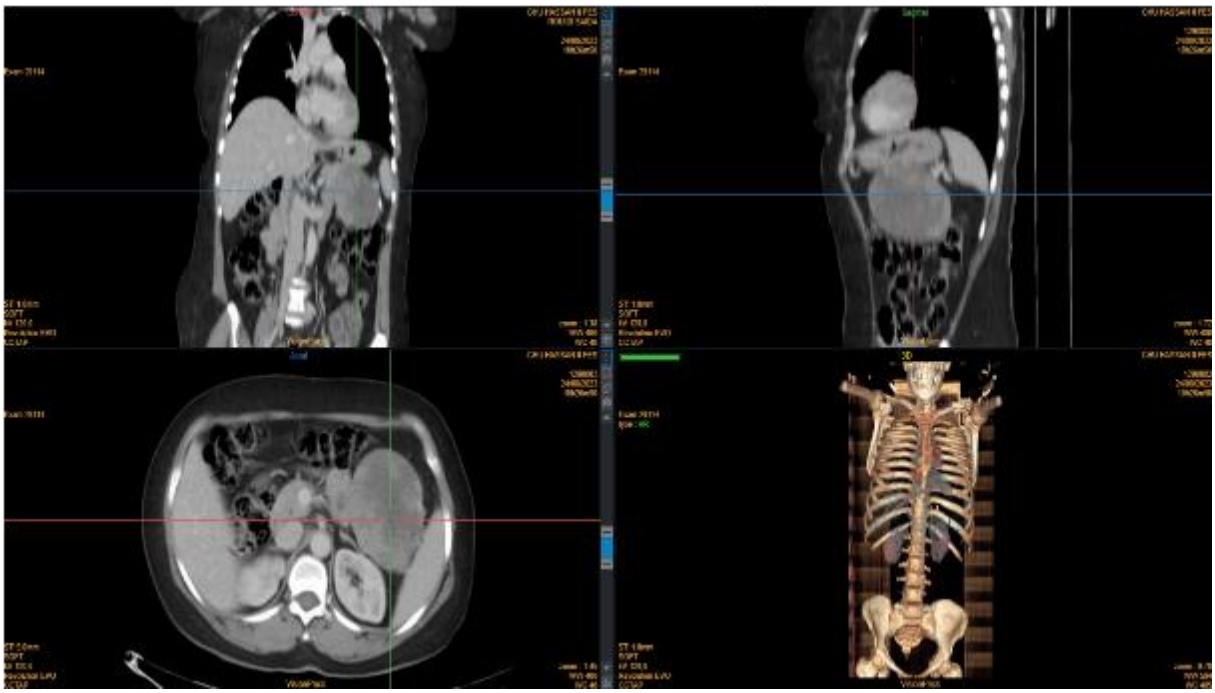

**Figure 1** CT scan: Axial, coronal and sagittal views with 3d reconstruction

Full blood test was carried out showing an increase in alpha and beta1 globulins and a polyclonal increase in gamma globulins with an EPP profile suggestive of inflammatory syndrome : Albumin: 40g/L, Protide: 78g/L, Alpha1: 3.9 g/L, Alpha 2: 9.3g/L, Beta 2: 5.5g/L, Beta2: 4.5g/L - Plasma protein electrophoresis shows no detectable monoclonal abnormality. - Bences jones proteinuria was negative - Light chain assay : Lambda: 2.2 g/L, Kappa: 4.5 g/L. - blood count, renal and liver function tests were normal.

A biopsy was carried out with an anatomopathological result and an immunohistochemical study which concluded in a plasmacytoma.

The patient's case was reviewed at a multidisciplinary consultation meeting: surgery was not feasible and the decision was made to carry out radiotherapy.

Our patient underwent radiotherapy at a total dose of 45gy in 25 fractions of 1.8gy per fraction for 5 weeks. During the radiotherapy, the patient's condition and appetite were acceptable with with grade I gastrointestinal toxicity. No abdominal pain, diarrhea, fever, or other symptoms were present. No adverse reactions were detected.

Regarding the follow-up of the patient, a history and physical examination was planned every 3 months for 2 years, and after that 2 times per year for 3 years and then, every 12 months. An abdomino-pelvic scan was done after 6 months of the end of radiotherapy who shows a regression in size of the intraperitoneal tumour mass.

After a follow up of 18 months, on clinical examination there is no palpation of the abdominal mass and a TEP scan was performed showing the absence of any lesion suspected of progressive disease or residual metabolically active disease.

## 3. Discussion

### 3.1. Epidemiology

Primary plasmacytomas are divided into solitary plasmacytomas of bone and solitary extramedullary plasmacytomas (EMP). EMP is a rare disease and is histopathologically characterized by infiltrates of plasma cells of diverse maturity and by their monoclonal immunoglobulin products (1). The disease occurs almost exclusively in the head, neck, and upper respiratory tract. EMPs in the gastrointestinal organs are uncommon (2). The next most frequent site of lesion occurrence is the stomach; however, this is also extremely rare, accounting for less than 5% of all EMPs (3). The median age at diagnosis is approximately 55 years about 10 years earlier than in multiple myeloma and there is a higher incidence among males. (4)

### 3.2. Clinical presentation

The main clinical symptoms of EMP are pain and those are caused by the compression exerted by local masses on the affected organs, for gastro-intestinal EMP change of bowel habit, and bowel obstruction can be can be suggestive symptoms. In addition, gastrointestinal bleeding has also been reported. (5,6) Nonetheless, there is no clinical specificity and systemic symptoms, such as those caused by multiple myeloma, including anemia, high serum calcium level, renal function damage, and bone destruction.

### 3.3. Imaging

Extramedullary plasmacytoma lesions are typically identified using axial imaging modalities or through direct visualization techniques such as laparoscopy or endoscopy. On non-contrast computed tomography (CT), these lesions usually appear as solitary, well-circumscribed masses often accompanied by areas of necrosis and cystic degeneration (7). Following contrast administration, CT scans demonstrate enhancement along the cystic wall as well as within any internal septations and solid components.

In a study by Ryu and Cohen-Hallaleh (8) involving 21 patients with extramedullary plasmacytoma (EMP), MRI findings demonstrated that these lesions are typically iso- to hypointense relative to muscle on T1-weighted images and range from iso- to hyperintense on T2-weighted images, with variable enhancement following contrast administration. Diffusion-weighted imaging consistently showed restricted diffusion within the tumor. However, for gastrointestinal tumors, CT and endoscopy are generally preferred over MRI due to the interference of bowel peristalsis artifacts.

Additionally, 18F-fluorodeoxyglucose positron emission tomography (FDG-PET) is useful for detecting areas of active disease, distinguishing extramedullary plasmacytoma from multiple myeloma, and monitoring treatment response (9).

### 3.4. Diagnosis:

The method for obtaining a definitive tissue diagnosis varies with the lesion's anatomical location. For example, endoscopic ultrasound-guided fine needle aspiration has been successfully performed for lesions in the pancreas (10), liver (11), and gallbladder (12), while gastroscopic biopsy is employed for gastric lesions (13). Given the risk of visceral injury associated with minimally invasive biopsy techniques in the abdomen, surgical resection or open/laparoscopic biopsy is often necessary to secure a formal diagnosis. Bone marrow and pathological biopsies are the indispensable examinations for confirmation of EMP. Bone marrow biopsy has no evidence of peripheral plasmacytosis or clonal marrow. Microscopic findings of pathological biopsies show the dense of neoplastic plasma cells and intranuclear immunoglobulin inclusion. (14). Emerging diagnostic approaches, such as assessing circulating plasma cell load and narrow (15) band imaging endoscopy (16), are also being explored.

Differential diagnoses for gastrointestinal extramedullary plasmacytoma include adenocarcinoma, gastrointestinal stromal tumors, multiple myeloma and lymphoplasmacytic lymphoma and immunoblastic lymphoma. Many cases of gastrointestinal plasmacytoma were misdiagnosed as low grade B-cell lymphoma with plasma cell differentiation (17). Histopathological confirmation requires the presence of clonal plasma cells, typically identified using immunohistochemical markers such as CD38, CD138, and CD79a, along with evidence of monoclonal kappa or lambda light chain expression. Although the immunophenotype closely mirrors that of multiple myeloma, the extramedullary distribution aids in differentiating the two conditions.

### 3.5. Management

Currently, there are no universally accepted guidelines for the management of intra-abdominal extramedullary plasmacytoma. Treatment planning is generally based on institutional experience and is informed by data from limited

case series. Therapeutic options include surgical resection, radiotherapy, systemic chemotherapy, or a combination of these modalities. Optimal patient outcomes rely on multidisciplinary discussions involving surgeons, hematologists, oncologists, radiologists, and pathologists. Systemic chemotherapy regimens are often analogous to those used in multiple myeloma, incorporating agents such as thalidomide, bortezomib, and high-dose corticosteroids. Various radiotherapy protocols have been described, including targeted approaches that deliver 40–50 Gy over a four-week period (18).

A larger case series by Li et al. (n = 38) suggested that radiotherapy alone might offer improved five-year local progression-free survival compared to a combined modality of surgery and radiotherapy. However, this series encompassed extramedullary plasmacytomas from a variety of anatomical sites—including head and neck, pulmonary, and abdominal primaries—without accounting for potential differences in tumor biology across tissues (19).

### 3.6. Prognosis

According to population-based registries (20), over 70% of extramedullary plasmacytomas eventually progress to multiple myeloma, with a median progression time of approximately 19 months, although reported intervals range from 7 to 293 months. This progression appears to be influenced by the specific expression profiles of chemokine receptors on malignant plasma cells, as well as by abnormalities in cellular adhesion molecules (21). Key predictors include Bartl's histological grade, tumor size, and an MIB1 proliferation index greater than 10% (22,23). Consequently, extramedullary plasmacytoma may initially present with clinical features suggestive of evolving multiple myeloma or disseminated disease, such as weight loss or cachexia, bone pain from intramedullary involvement, renal failure, anemia, or neurological symptoms. Five-year local overall survival rates are generally favorable—ranging between 78.4% and 87.4%—although significantly lower outcomes have been reported in patients with intra-abdominal disease. Furthermore, local recurrence, relapse, or progression to plasma cell myeloma may occur as late as 5 to 10 years after resection, underscoring the need for prolonged patient surveillance (13).

## 4. Conclusion

Our case report highlights a rare presentation of intra-oabdominal extramedullary plasmacytoma, emphasizing the diagnostic challenges and the critical role of a multidisciplinary approach. Advanced imaging modalities and targeted biopsy techniques facilitated a definitive diagnosis, underscoring the importance of considering EMP in the differential diagnosis of intra-abdominal mass lesions. Tailored treatment strategies that integrate surgical, radiotherapeutic, and chemotherapeutic modalities are essential for optimal patient outcomes. Given the risk of late local recurrence and progression to multiple myeloma, long-term surveillance remains crucial. Further studies with larger cohorts are needed to refine management guidelines and improve prognostic evaluation for this uncommon entity.

## Compliance with ethical standards

### *Disclosure of conflict of interest*

All authors have no conflict of interest to declare.

### *Statement of informed consent*

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

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