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Case report

# Anal canal plasmacytoma—An uncommon presentation site

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ABSTRACT

*Background:* Extramedullary plasmacytomas (EMP) are rare plasma cell tumors that arise outside the bone marrow. They are most often located in the head and neck region, but may also occur in the other locations. The lower gastrointestinal EMP represents less than 5% of all cases, and location in the anal canal is exceedingly rare.
*Aim:* We present an exceedingly rare case of anal canal plasmacytoma, aiming to achieve a better understanding of this rare entity.
*Methods:* We report a case of a 61-year-old man with a bulky mass in the anal canal. The lesion measured about 6 cm and invaded in all layers of the anal canal wall. The biopsy was performed and revealed a round and plasmocitoid cell population with a solid growth pattern and necrosis. The tumoral cells have express CD79a and CD138 with lambda chains. There was no evidence of disease in other locations and these features were consistent with the diagnosis of an extra-osseous plasmacytoma. The patient was submitted to conformal radiotherapy 50.4 Gy total dose, 1.8 Gy per fraction. After 24 months, the patient is asymptomatic and the lesion has completely disappeared.
*Conclusions:* EMP accounts for approximately 3% of plasma cell malignancies. The median age is about 60 years, and the majority of patients are male. The treatment of choice for extramedullary plasmacytoma is radiation therapy in a dosage of about 50 Gy. Patients should be followed-up for life with repeated bone marrow aspiration and protein studies to detect the development of multiple myeloma.



## 1. Background

Solitary plasmacytomas (SP), a monoclonal proliferation of plasma cells without evidence of significant bone-marrow plasma-cell infiltration, is comprised of two main groups: solitary plasmacytoma of the bone (SPB) and extramedullary plasmocytoma (EMP). SP is a rare disease, and accounts for about 10% of plasma cells tumors.[1–3] The clinical presentation of the former generally includes bone involvement, with pain, neurological deficit and, sometimes, pathological fractures. The latter often presents as a mass which becomes symptomatic if compresses adjacent structures.

EMPs are most often located in the head and neck region, mainly in the upper aerodigestive tract, but may also occur in the gastrointestinal tract, urinary bladder, central nervous system, thyroid, breast, testes, parotid gland, lymph nodes,

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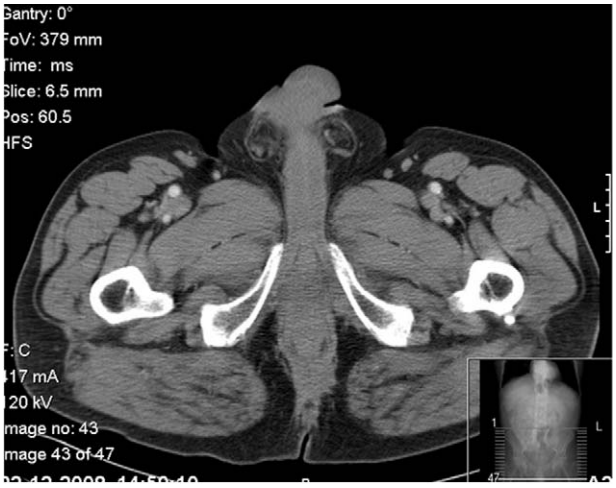


**Fig. 1 – Axial CT.**

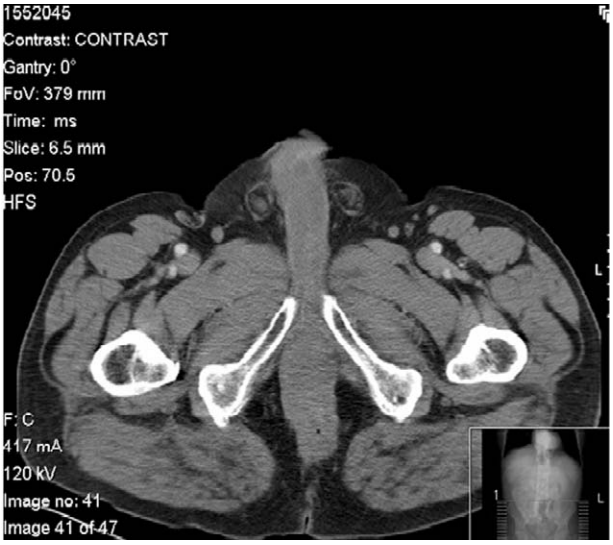


**Fig. 2 – Axial CT.**

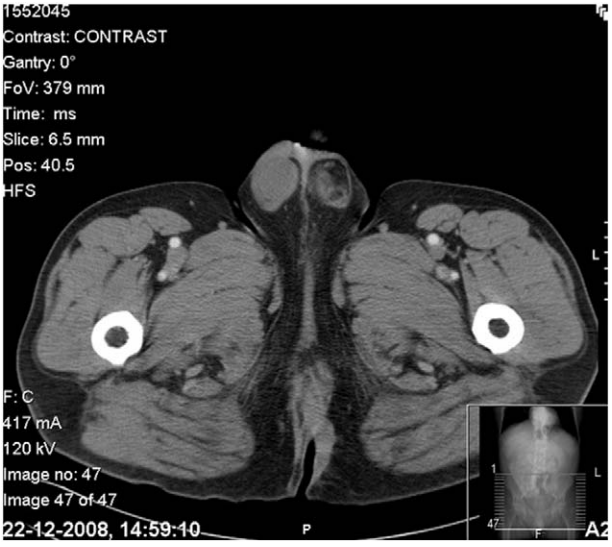


**Fig. 3 – Axial CT.**

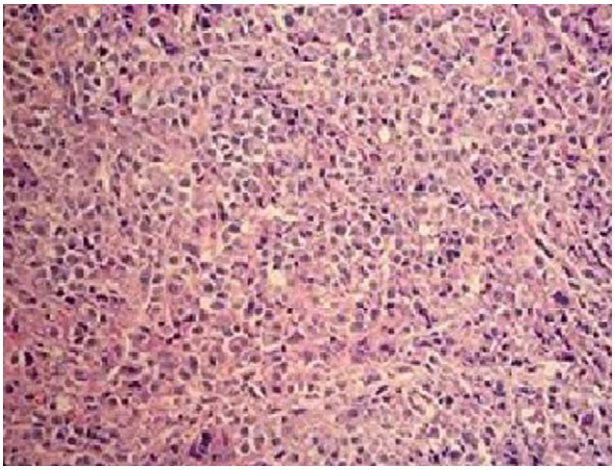

**Fig. 4 – Photomicrograph.**

and skin. The lower gastrointestinal EMP represents less than 5% of all cases.

We report an exceedingly rare case of anal canal plasmacytoma, aiming to achieve a better understanding of this rare entity.

## 2. Case report

A 61-year-old white man presented with a 4-month history of abdominal discomfort, tenesmus and perineal pain. On physical examination, he was in a good general status (IK 90%, PS 1). The digital rectal examination (DRE) revealed an impinging mass, of about 4 cm, hard consistency, at 2 cm of the anal margin, in the left posterior margin, without any palpable nodes. The blood testing showed haemoglobin 11.9 g/dL, white blood cells $5.73 \times 10^9$/L, β 2 microglobulin was slightly elevated (3.13 mg/L; Ref. 0.8–1.8 mg/L), and the protein electrophoresis revealed a slender elevation of the IgG. The pelvic CT revealed a bulky mass in the anal canal of about 6 cm that invaded all layers (Figs. 1–3). The biopsy was performed and histological examination identified a round and plasmocitoid cell population with solid growth pattern and necrosis. The tumoral cells expressed CD79a and CD138 with lambda light chains (Fig. 4).

There was no evidence of disease in other locations and these features were consistent with the diagnosis of an extramedullary plasmocyoma (EMP) according to Internacional Myeloma Working Group (IMWG).[4]

The patient was referred to our department and submitted to conformal 3D external radiotherapy (RT) directed to the lesion with a total dose of 50.4 Gy, 1.8 Gy/F, 5F/week. The irradiated PTV[1] included the tumor identified in the clinical exam and planning TC (GTV[2]) and a margin for subclinical disease (CTV[3]) and for variations and uncertainties (PTV). During the treatment, the patient complained about dysuria and occasional hematuria, which were treated with nonsteroidal

[1] PTV – planning target volume.
[2] GTV – gross tumor volume.
[3] CTV – clinical target volume.

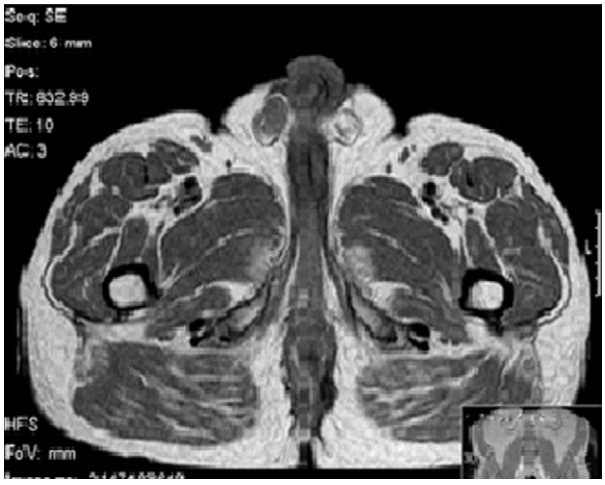


**Fig. 5 – Axial MRI.**

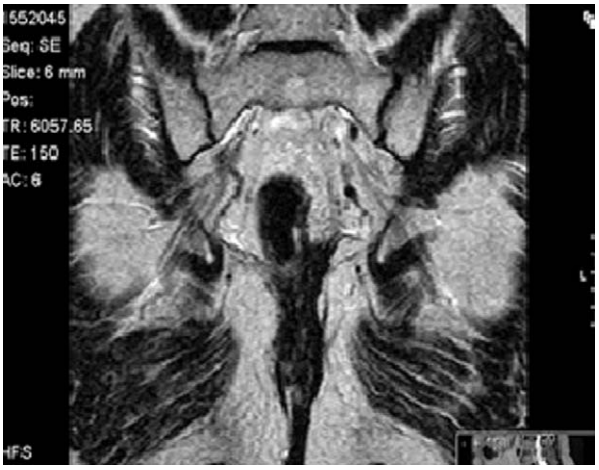


**Fig. 6 – Coronal MRI.**

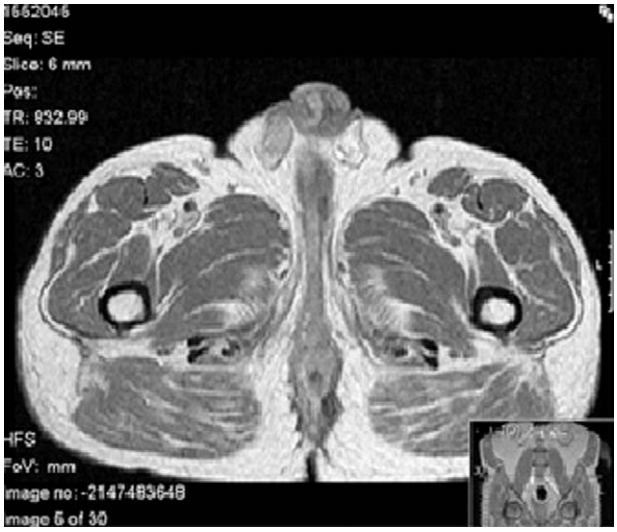


**Fig. 7 – Sagittal MRI.**

anti-inflammatory drug (NSAIDs) and completely resolved at the end of RT. He also mentioned resolution of the initial complaints that had led to the diagnosis.

Six months after RT, the patient was asymptomatic and the lesion was markedly reduced (about 70%). One year later the mass completely disappeared, and only fibrotic changes were identified on pelvic MRI (Figs. 5–7). There was no bone marrow infiltration or other multiple myeloma (MM) features. The patient maintained the response at last follow up, twenty-four months after treatment.

## 3. Discussion

Extramedullary plasmocyoma (EMP) is characterized by the presence of a monoclonal plasma cell tumor at an extramedullary site with no evidence of MM. Some patients may have a small M-protein in the serum or urine, which often disappears after treatment.[4,5]

The lower gastro-intestinal tract is an uncommon site for EMPs, since they are usually located in the stomach or small bowel.

For diagnosis, a positive tissue biopsy is required. MRI of pelvis is recommended to better characterize the lesion and subsequently for RT planning.

Multiple lesions should be excluded in order to select patients eligible for radical RT.[6,7] Positron emission tomography (PET) with $^{18}$F-fluoro-deoxyglicose (FDG) is becoming more relevant in diagnosis and staging of many tumors, including lymphoma, and – as whole body imaging exam – can reveal new sites of disease otherwise ignored by other recommended imaging modalities, such as bone scan, TC or MRI. PET has been proven to be useful in diagnosis and assessing the response to RT in SP.[7,8] It may also be very useful in RT planning, as images can be imported into treatment planning systems (TPS).[7]

Surgery and RT may be used for EMP treatment.[9–12] If the patient has been performed a complete resection during the diagnosis evaluation, adjuvant RT is indicated only if there is a suspicion of residual disease. Otherwise, IFRT is the treatment of choice in a great majority of cases, with a high cure rates. Currently, there is no conclusive data published on the optimal radiation dose for SP. Doses varying from 30 to 60 Gy are described, but most centers use $\geq$50 Gy.[8,13]

Many authors agree that age is a prognostic factor for progression to MM, while others do not. As MM is more common in the elderly, many patients diagnosed with EMP hypothetically harbor subclinical disease which has not been detected. Persistence of M-protein after RT may predict subsequent systemic failure.[3,4,8] Tumor size is relevant for local control. Authors report better outcome in lesions <5 cm.[3] Some other factors, such as local invasiveness, proliferation rate and morphologic grade, usually barely documented, may have an effect on the outcome.[8] Although our patient presented a tumor bigger than 5 cm, M-protein was absent after RT.

In our case, after radical treatment with RT, there was no evidence of disease and our patient remains disease free two years after RT.

The frequency of progression of MM to SP after RT described in the literature suggests that, at least in some cases, the staging work up was insufficient. We can expect that an extended

use of PET may improve patients' staging and more patients could be diagnosed with MM instead of SP, which could optimize treatment.

Theoretically, chemotherapy might have a role in eradicating subclinical disease and enhancing local control.[8]

As it is not yet possible to predict which patients will progress to MM, we recommend that all patients should be followed-up for life with periodical lab exams and imaging.

## 4. Conclusion

This paper is, to our knowledge, one of few reports in the literature of EMP of the anal canal. Further reports of new cases can help establish optimal treatment guidelines.

## Conflict of interest

All the authors deny any conflict of interest.

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