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# Acta Otorrinolaringológica Gallega

## Caso Clínico

### Systemic disease presenting as frontal forehead lump

### Enfermedad sistémica presentada como bulto en la frente

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### Abstract

We describe in this report a case of a Solitary Extramedullary Plasmacytoma presenting as a forehead lump in a 74-year-old woman with previous history of Multiple Myeloma. Forehead lumps are an uncommon presentation in otorhinolaryngology patients and a significant proportion are manifestations of malignancies.

We have found extremely few publications in the literature indicating locations of Extramedullary Plasmacytoma in the frontal sinus.

Besides that, a recurrent Multiple Myeloma presenting as an Extramedullary Plasmacytoma despite concomitant bone marrow response to therapy is a rare presentation of relapsed disease.

This clinical report highlights the fact that systemic diseases may manifest in our clinical practice and there should be a high index of suspicion, in order to the patient could be treated in the best way.

**Keywords:** Multiple Myeloma, Solitary Extramedullary Plasmacytoma, Frontal sinus.

### Resumen

Describimos un caso de un Plasmacitoma Extramedular Solitario que se presenta como un bulto en la frente en una mujer de 74 años con an-

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tecedentes de Mieloma Múltiple. Los bultos en la frente son una presentación poco común en pacientes otorrinolaringológicos y en una proporción significativa son manifestaciones de tumores malignos.

Hemos encontrado muy pocas publicaciones en la literatura que indiquen ubicaciones de Plasmacitoma Extramedular en el seno frontal. Además de eso, un mieloma múltiple recidivante que se presente como un Plasmacitoma extramedular a pesar de la respuesta concomitante de la médula ósea al tratamiento, es una presentación rara de la enfermedad recurrente.

Este caso clínico destaca el hecho de que las enfermedades sistémicas pueden manifestarse en nuestra práctica clínica y debe haber un alto índice de sospecha, para que el paciente pueda ser tratado de la mejor manera.

**Palabras Clave:** Mieloma múltiple, Plasmacitoma extramedular solitario, seno frontal.

### **Introduction**

Multiple Myeloma (MM) is characterized by malignant proliferation of clonal plasma cells, usually restricted to the bone marrow. MM comprises approximately 1% of all cancers and is associated with very poor clinical outcomes<sup>1</sup>. Extramedullary Plasmacytoma (EMP) is characterized by a mass of neoplastic monoclonal plasma cells occurring outside the bone marrow, in soft tissue and organs, without evidence of systemic disease attributing to myeloma<sup>2</sup>. It is an uncommon manifestation in MM and can either accompany newly diagnosed disease or develop with disease progression or relapse. It can appear in any soft tissue, but 80% are located in the upper airway<sup>3</sup>. The incidence of EMP is 7% to 18% at MM diagnosis and up to 20% at relapse<sup>4,5</sup>. We present a case referred to our hospital of a Solitary Extramedullary Plasmacytoma presenting as a forehead lump in a patient with recurrent MM.

### **Case Report**

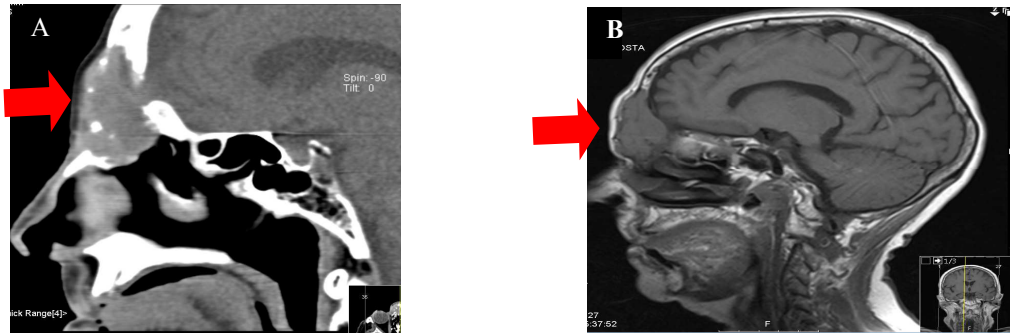
We present the case of a 74-year-old woman who presented to our Hospital with a 2-month history of a painless, progressively enlarging mass on the right side of the forehead and bridge of the nose, without other symptoms. Her past history revealed diagnosis of Multiple Myeloma (MM) 11 years ago and she received several chemotherapy treatments. She had remained in remission for more than 2 years.

Local examination revealed a 3 cm frontal tender mass. Computed Tomography (CT) scan and Magnetic Resonance Imaging (MRI) (figure 1) showed a 3,6 × 3,8 cm lesion, in the frontal region, with bone erosion, involving the frontal sinus, the nasal pyramid, the anterior ethmoid, the cribriform plate and the maxillary sinus walls. It was suspected that the lesion might represent recurrent MM.

Using a transnasal endoscopic approach and an external approach, the tumour was partially excised and a skin reconstruction with a flap was made (figure 2). The lesion extended from frontal sinus bilaterally, to

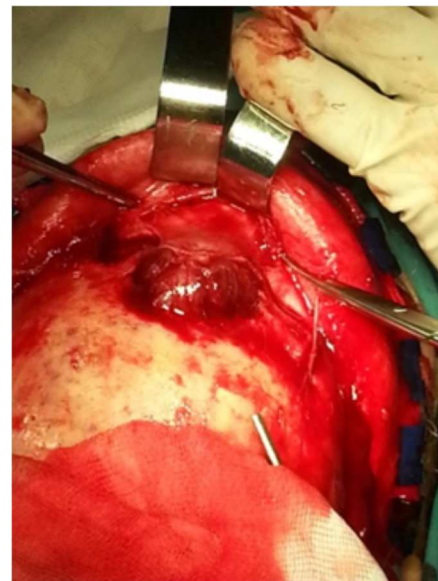
nasal bones, right lamina papyracea and ethmoid posteriorly. The postsurgical histopathological analysis revealed proliferation of plasma cells, consistent with Extramedullary Plasmacytoma (EMP).

The patient was referred to the Haematology Service, which carried out the appropriate extension study, concluding no evidence of intramedullary disease. The patient was diagnosed with recurrent Multiple Myeloma presenting as a Solitary Extramedullary Plasmacytoma. Radiotherapy plus chemotherapy treatments were instituted and serial follow-ups were scheduled.



**Figure 1:** Sagittal CT scan (A) and MRI (B) before the surgery, showing the lesion (red arrows).

**Figure 2:** The lesion seen through external approach. It is possible the macroscopic appearance of the mass.



### **Discussion**

EMP is a rare plasma cell neoplasm which involves soft tissues, without systemic involvement<sup>6</sup>. From those plasma cell tumors localized in the upper aero-digestive tract, approximately 80% occur within the sinonasal cavity<sup>6</sup>. Only a few cases of EMP (15-20%) progress to MM and it is impossible to identify which cases will do it<sup>6</sup>.

Clinical manifestations of EMP vary according to its location. Nasal obstruction, soft-tissue swelling, headache, epistaxis and anosmia are just some examples of the symptoms that can be found if the lesion

is found in the nasal cavity or surroundings. In the case of orbital involvement, it can cause diplopia, ptosis, decreased visual acuity or impaired ocular motility<sup>7</sup>.

CT and MRI are complementary techniques in assessing the size and location of a tumour and the involvement of the neighbouring structures. The images of CT and MRI are nonspecific, showing lesions of soft parts or infiltrating lesions. Bony destruction is displayed depending on the expansion of the tumour. In MRI assessment, isointense images on T1-weighted and iso to hyperintense images on T2-weighted sequences are well demonstrated in MRI assessment. It should be pointed out that MRI and CT could not distinguish EMP from other possible differential diagnosis<sup>8</sup>. The diagnosis is given by the histological examination that consists of a monoclonal proliferation of plasma cells.

To reach a diagnosis, an extension study is also required. This study consists of blood and urine protein electrophoresis, analysis of renal and hepatic function, biopsy of bone marrow and complete skeletal study with CT or PET scan. The presence of anaemia, hypercalcaemia and renal insufficiency should also be discarded<sup>3,9</sup>.

Regarding treatment, surgery is needed to biopsy the lesion and to attain a diagnosis. However, it should be noted that depending on the nature of the lesion, radiotherapy or chemotherapy treatments are needed<sup>3</sup>.

These patients require a rigorous follow-up since EMP has a risk of recurrence of 22% and the possibility of causing distance metastasis of 30%, which can occur several years after the initial diagnosis<sup>10</sup>.

We have found extremely few publications indicating locations of EMP in the frontal sinus. Besides that, a recurrent MM presenting as a EMP despite concomitant bone marrow response to therapy is an uncommon presentation of relapsed disease.

Subsequently, this is a very rare case, which at the outset led us to consider a wide differential diagnosis.

In fact, forehead lumps are an uncommon presentation in otorhinolaryngology patients and a significant proportion are manifestations of malignancies.

Concluding, this clinical report highlights the fact that systemic diseases may manifest in our clinical practice and there should be a high index of suspicion. In this case, once the diagnosis was established, the patient could be treated in the best way.

**Conflict of interest:** The authors declare that they have no conflict of interest.

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