

# Multifocal Extramedullary Plasmocytoma of the Sinonasal Region: a Case Report and Literature Review

Plasmocitoma Multifocal Extramedular de la Región Sinonasal:  
Reporte de un Caso y Revisión de la Literatura

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**ABSTRACT:** We report on a patient who presented to ENT services with right side epistaxis, frontal lobe headache, right infraorbital pain and the feeling of having a stuffy nose. CT and MRI were performed and later a biopsy confirmed the presence of sinonasal plasmocytoma. The Head and Neck oncology committee recommended radiotherapy as the choice of treatment. At the present time the patient is being followed on scheduled medical visits.

**KEY WORDS:** CT, MRI, plasmocytoma, sinonasal, extramedullary, tumor.

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## INTRODUCTION

Multiple Myeloma (MM), Solitary Bone Plasmocytoma (SBP) and Extramedullary Plasmocytoma (EMP) are plasmatic cell tumors that can be grouped in one category. Only 30% of extramedullary plasmocytomas evolve into multiple myelomas, while most bone plasmocytomas do (Gross *et al.*, 2004; Seoane *et al.*, 2003) Schridde described the first extramedullary plasmocytoma in 1905 (Schridde, 1905). Extramedullary plasmocytomas represent less than 1% of all head and neck neoplasms and 4% of all non-epithelial nasal tumors (Kanoetra & Lateef, 2010). Extramedullary plasmocytomas are usually confined to soft tissues without bone involvement (Mcafee *et al.*, 2004) and are produced by monoclonal plasmatic cell proliferation. Occurrence can happen at any age, but the majority appear more frequently in the fifth and seventh decades (Jaswal *et al.*, 2008; Megat Shiraz *et al.*, 2008; Lomeo *et al.*, 2007). Extramedullary plasmocytoma affects more males than females and studies have reported 4:1 and 2:1 ratios.

Lesions are predominantly seen in the upper nasal and GI tract affecting the submucosal tissue of the nasal cavity and paranasal sinuses 43.8% of the time, nasopharynx 18.3%, oropharynx 17.8% and larynx 11.1% (Megat Shiraz *et al.*; Lomeo *et al.*; Alexiou *et al.*, 1999) Affected regions could present as solitary or multiple masses but present more commonly as solitary lesions.

Plasmocytomas can be characterized by the degree of atypia as follows: Low or grade 1 intermediate or grade 2 and advanced or grade 3. Also, based on laboratory tests such as blood, urine, bone scans, bone marrow test and radiographic studies, it can be staged as, 1 or localized; stage 2 when there is local invasion and lymph node involvement and stage 3 when metastatic invasion is present.

There are no specific clinical manifestations or symptomatology since it varies depending on tumor

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location. The most common symptom is the presence of a lump or a sessile or pedunculated soft tissue mass. These lesions are circumscribed or infiltrating in 80% of the cases. Nasal obstruction and pressure is present in 35% of the cases, epistaxis 35%, pain 20%, ocular proptosis and blurry vision 15%, rhinorrhea 10%, regional lymphadenopathy 10%, facial nerve paralysis 5% with symptoms often lasting from 4 to 5 months (Miller *et al.*, 1998).

Diagnosis of Extramedullary Plasmocytoma is achieved from a biopsy of the lesion and from histologically and immunohistochemical analysis. It is based on the presence of plasmatic cells showing birefractation with Congo red stain and presenting cytologically as benign tracts however, immunohistochemical tests demonstrate its monoclonality pointing to its neoplastic nature (González *et al.*, 2011). Neoplastic cell usually produce a large chain of monoclonal immunoglobulin that can be detected in both blood and urine (Pinto *et al.*, 2007). Radiologic imaging is crucial in the evaluation and localization of the size of the lesion. Magnetic Resonance Imaging (MRI) helps to determine the extent of soft tissue involvement often showing up as hyperintense lesions in T1 relaxation and homogeneously enhancing after Gadolinium contrast administration (Waldro & Mitchell, 1988; Cakir *et al.*, 2003). In Computed Tomography (CT), plasmocytomas are seen as radiopaque lesions, with homogeneous contrast enhancement.

Due to the high radiosensitivity of the tumor the recommended standard treatment is Radiation therapy. Depending on the size of the tumor, the therapeutic dose ranges from 40 to 50 Gys, for 4 to 6 weeks. Surgery could be used as the primary treatment if the size and location are favorable. In some cases, a combination of surgery and radiotherapy could be used depending on the possibility of resecting the lesion (Liebross *et al.*, 1999). Surgery is the best choice in cases of recurrence, while chemotherapy is indicated only when there are multiple and refractory lesions or when recurrence cannot be treated surgically (Soutar *et al.*, 2004).

It has been reported that 22% of the cases that have been adequately treated for multiple extramedullary plasmocytoma in the superior gastro intestinal tract have resulted in a local recurrence (Kanotra & Lateef). Survival rate has been reported to be 10 years for 70% of those afflicted with the disease (Knowling *et al.*, 1983). The best prognostic indicator

for multiple extramedullary plasmocytoma is whether the lesion progresses into multiple myeloma. Progression into multiple myeloma occurs in 15% to 20% of the cases and this is often associated with a poor prognosis reducing the survival rate from 10 years to 20 months (Fu & Perzin, 1978; Shreif *et al.*, 2001). The prognosis is even worse when there are multiple lesions present (Mochimatsu *et al.*, 1993; Wanebo *et al.*, 1966).

## CASE REPORT

We report on a 52-year-old male patient who presented to ENT services without a significant clinical history. However, he complained of having the following symptoms; right side epistaxis, frontal lobe headache, right infraorbital pain and the feeling of having a stuffy nose. CT and MRI were performed with the CT revealing a solid and expansive mass occupying the right nasal fossa and left maxillary sinus. The mass was associated with inflammatory changes and dense secretions emerging from the right maxillary sinus (Fig. 1). The mass demonstrated a moderate enhancement after intravenous contrast was given (Fig. 2). The mass revealed erosion of the cribriform plate producing dehiscence without a clear extension into the anterior cranial fossa. Destruction of the nasal conchae, medial orbital wall and right maxillary sinus can be seen (Fig. 3).

MRI revealed an expansive process of low intensity in the right nasal fossa suggestive of high cellular activity (Figs. 4 and 5), and heterogeneous enhancement after gadolinium intravenous injection suggestive of necrosis in the central area. Also, a Focal

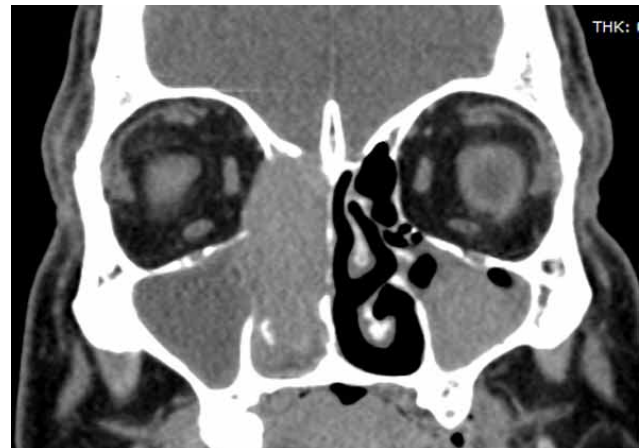


Fig. 1 Invasion of the mass is seen affecting the soft tissue in the right nasal fossa and right maxillary sinus.

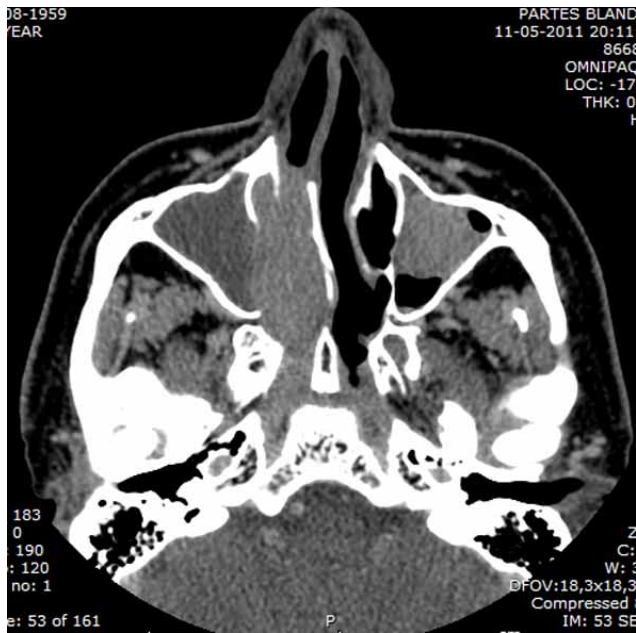


Fig. 2 Image following contrast media injection shows poor to moderate enhancement of the region.

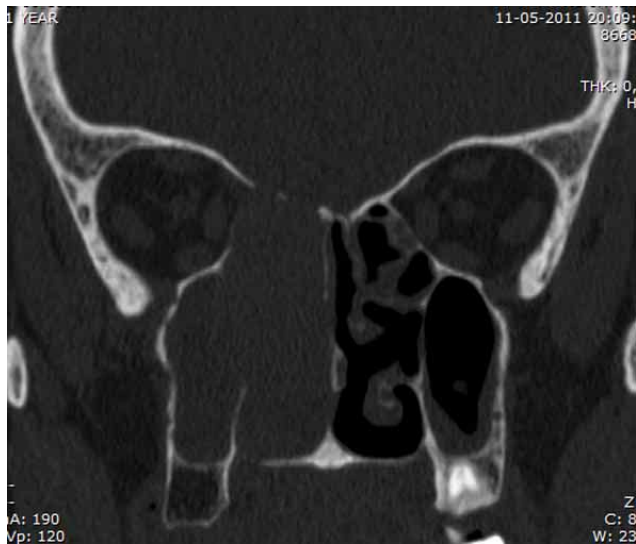


Fig. 3 Erosion of the cribriform plate is seen affecting the right nasal conchae, lateral wall of the orbit and right maxillary sinus.

lesion with rounded contours in the left maxillary sinus was seen with moderate enhancement after contrast injection. Mucosal thickening in the right maxillary sinus and the appearance of secretions were seen in the central (Figs. 6 and 7).

A biopsy was performed and the immunohistochemical analysis revealed positive results for CD 138 and Vimentine; negative results for

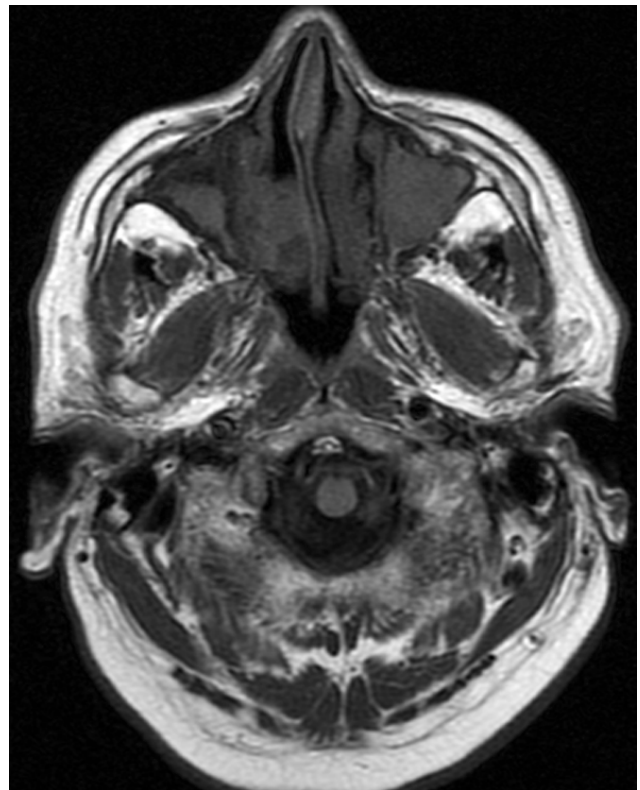


Fig. 4. T1 weighted image reveal masses in the right nasal and maxillary sinus areas.

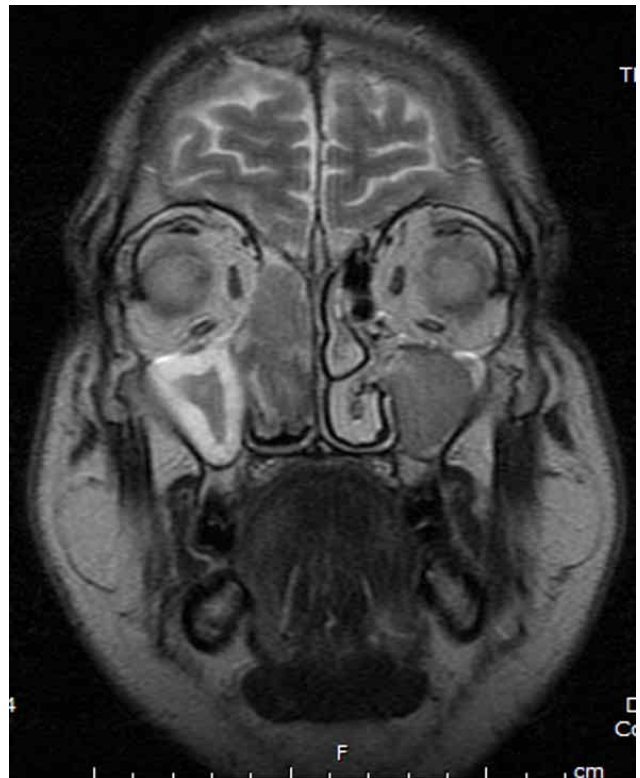


Fig. 5. T2 weighted image shows the presence of mucus occupying the right maxillary sinus.

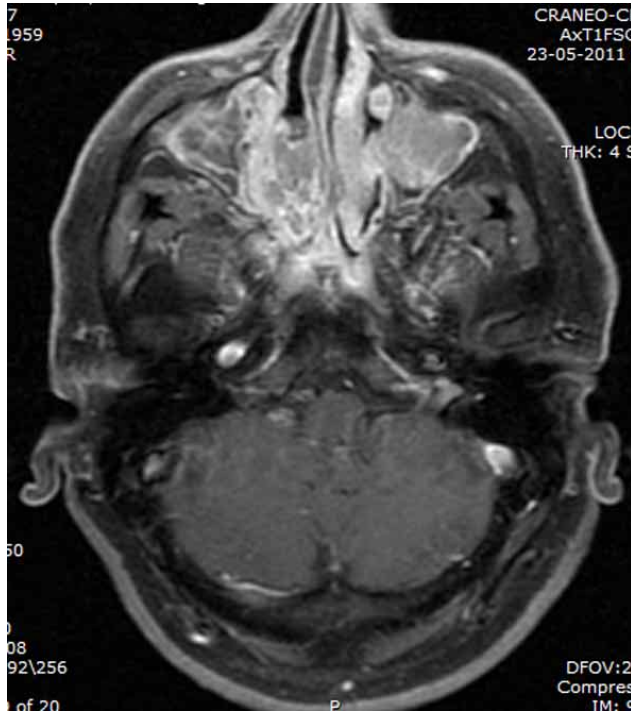


Fig. 6. T1 weighted image demonstrate heterogeneous areas of the mass following injection of contrast media.

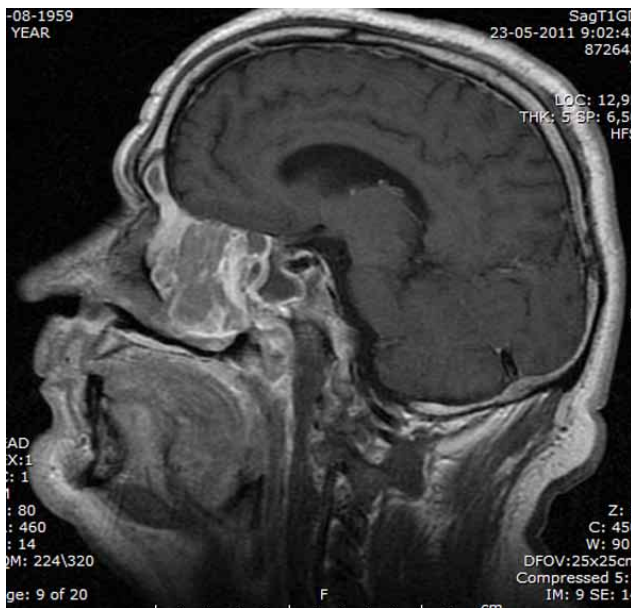


Fig. 7. Post gadolinium contrast media injection image fails to reveal anterior cranial fossae involvement.

S-100 and Synaptophysin. Ultimately the histopathology revealed “Maxillary and Nasal Plasmocytoma” (Fig 8)

A consultation with the Hematology service to rule out Multiple Myeloma was carried out and

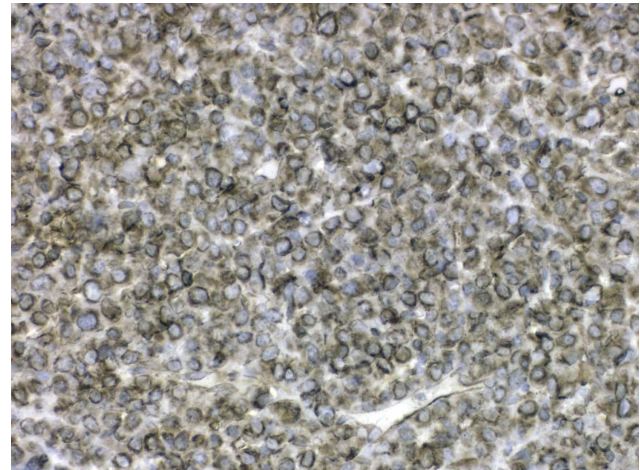


Fig. 8. Positive results for Vimentine.

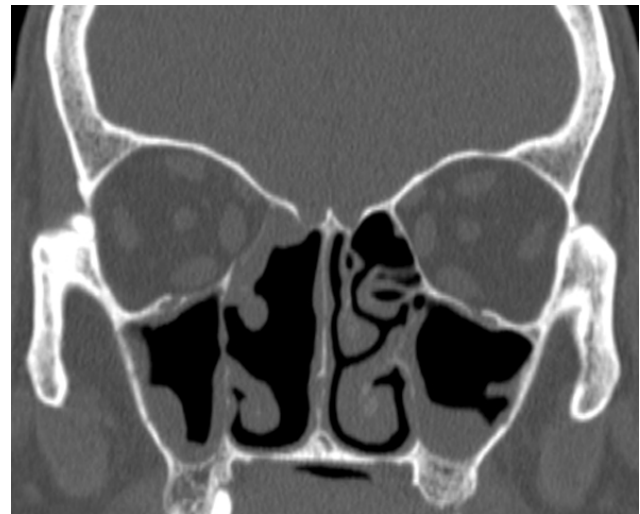


Fig. 9. Follow up CT shows resolution of the mass and thickening of the mucous membrane in maxillary sinus regions.

consequently The Head and Neck Cancer Committee recommended radiation therapy resulting in excellent results as seen in Figure 9. Currently the patient is evaluated on a regular basis to assess his progress.

## CONCLUSION

Extramedullary Plasmocytoma are tumors that present predominantly in the upper respiratory-digestive tract. Clinical signs and symptoms are non-specific and could vary due to tumor location. The most common symptom is the presence of tumefaction or soft tissue mass. The final Diagnosis is based on immunohistochemical and histological studies showing the presence of plasmatic cells. Imaging is crucial for

evaluating the location and size of the lesion differentiating it from adjacent inflammatory processes Standard treatment is radiation therapy due to the radiosensitivity of the lesions. The 10-year survival rate

is 70%. Progression of the lesion to Multiple Myeloma is associated with a poor prognosis reducing the survival rate approximately to 20 months from the time it changes.

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**RESUMEN:** Se reporta el caso de un paciente que se presentó en el servicio de Otorrinolaringología con epistaxis, cefalea del lóbulo frontal derecho, dolor infraorbitario derecho y la sensación de tener la nariz tapada. Se realizaron TC y RNM, y luego una biopsia confirmó la presencia de un plasmocitoma nasosinusal. El comité de oncología de cabeza y cuello recomendó radioterapia como tratamiento de elección. En la actualidad, el paciente está en seguimiento y control en las visitas médicas regulares.

**PALABRAS CLAVE:** tomografía computarizada, resonancia nuclear magnética, plasmocitoma, nasosinusal, extramedular, tumor.

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