

## Extramedullary Plasmacytoma of Aero-Digestive Tract: About 3 Cases

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### Abstract

### Original Research Article

Extra-medullary plasmacytoma is a rare tumor, resulting from the malignant proliferation of particular B lymphocytes, the plasma cells. However, there are localized forms, divided into solitary bone plasmacytoma (POS) and extramedullary plasmacytoma (EMP). The incidence is about 3-4% of all plasmacytic neoplasms. The median age of EMP presentation is 55 years with male predominance (75% of cases). The majority of cases of EMP (80% of cases) reported in the literature are in the head and neck [3], particularly in the upper aero-digestive tract and nasosinus. His treatment is based on chemotherapy, radiotherapy or surgery for some locations. His prognosis depends essentially on the risk of progression to multiple myeloma.

**Keywords:** Extra-medullary, tumor, plasmacytoma, chemotherapy.

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## INTRODUCTION

Extra-medullary plasmacytoma is a rare tumor, resulting from the malignant proliferation of particular B lymphocytes, the plasma cells. These most often infiltrate the bone marrow and generate a disseminated tumor: multiple myeloma (MM) [1]. However, there are localized forms, divided into solitary bone plasmacytoma (POS) and extramedullary plasmacytoma (EMP), the otorhinolaryngologist must be particularly attentive to 90% of the level of the upper aero-digestive tract.

Three cases of EMP have been diagnosed in the last 5 years at Rabat University Hospital. All three cases had an EMP diagnosed according to recognized criteria in the literature.

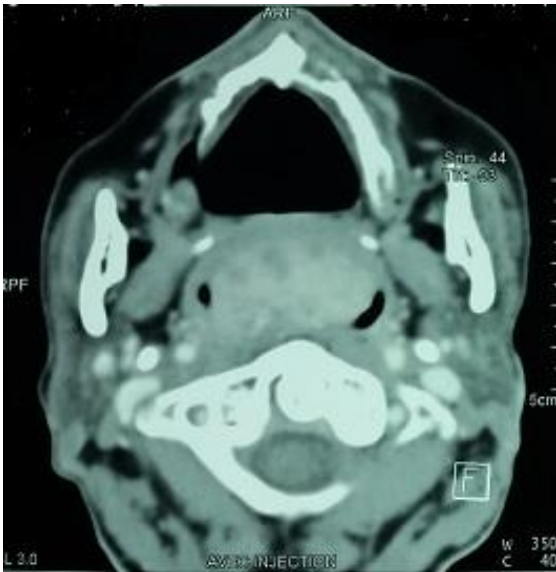
## MATERIALS AND METHODS

We report a case of patient 61-year-old diabetic present a progressive nasal obstruction associated with a nasal voice for one year. The evolution was marked by the appearance of a dysphagia, with deterioration of the general state. Physical examination was revealed an erythematous mass, in the oropharynx (Figure-1), and obstructing the oral cavity. Cervical examination didn't found a lymphadenopathy. A CT scan showed a lesion process attached to the posterior wall of the oropharynx obstructing the latter, and rising in the nasopharynx

(Figure-2), enhancing the injection of the contrast medium. This process was moderately vascularized with arteriography. A transoral biopsy was performed.

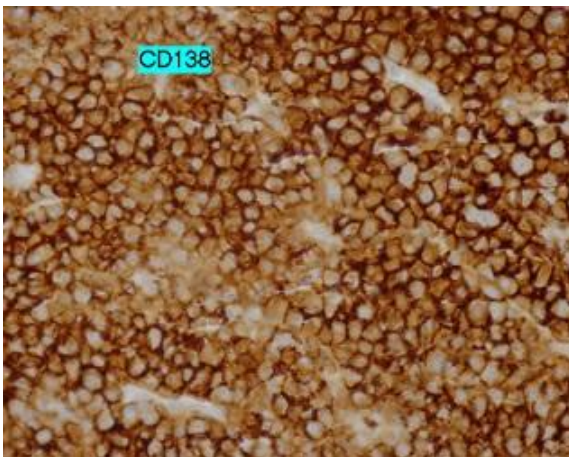


Fig-1: Oropharyngeal Plasmacytoma



**Fig-2: CT scan showed a lesion process attached to the posterior wall of the oropharynx**

Anatomo-pathologic examination with immunohistochemical study (Figure-3) revealed cells with plasmocyte cell differentiation, intensely expressing and diffusing the anti-CD138 antibody (MI15-Dako). Thus, the diagnosis of extramedullary plasmocytoma was made.



**Fig-3: Immunohistochemical Study of Plasmocytoma**

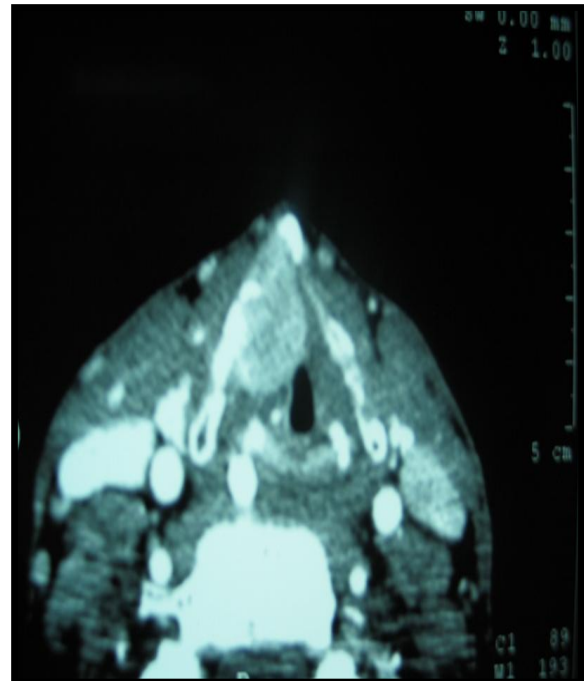
The medullary biopsy is normal (medullary plasmocytosis of less than 10%), the biological assessment (blood count, blood ionogram, serum calcium, creatinemia, electrophoresis of blood and urine proteins), and radiology (radiographs of the skeleton), have confirmed the extraosseous localization.

The patient underwent tumor reduction and then referred to chemotherapy and radiotherapy. Local recurrence or signs of progression to multiple myeloma wasn't noted at 6 months after treatment.

The other cases are 2 men aged 50 and 51 respectively, who were admitted to ENT emergencies for severe laryngeal dyspnea with dysphonia that required tracheotomy. Nasofibroscope showed the

presence of a whitish, glottic budding tumor in the first patient and subglottic in the second.

On cervical CT (Figure-4): it was an heterogeneous laryngeal process. Laryngoscopy with biopsy and anatomo-pathological examination with immunohistochemical studies was revealed a laryngeal plasmocytoma. The patients are referred to chemotherapy and radiotherapy.



**Fig-4: Cervical CT revealed a heterogeneous laryngeal process**

## DISCUSSION

Extramedullary plasmocytomas (EMP) are a rare solitary tumors, including proliferations of neoplastic plasma cells that occur in locations other than bone. They must be differentiated from multiple myeloma. This can be difficult because they can be associated later with the development of multiple myeloma [2]. EMP is rare with an average incidence of 3 per 100 000 habitants per year in France [3], representing less than 1% of head and neck tumors [14], but 4% of non-epithelial tumors in this anatomical region [15]. It is in 80 to 90% of the cases of a submucosal tumor of the upper aerodigestive tract [3-12].

The majority of cases of EMP (80% of cases) reported in the literature are in the head and neck [3], particularly in the upper aerodigestive tract and nasosinus cavities. They represent only 1% of all tumors in this region [13]. Oropharyngeal localization is extremely rare and usually occurs in the tonsils [14]. Thus one of our patients has an oropharyngeal location.

The incidence is about 3-4% of all plasmocytic neoplasms. The median age of EMP presentation is 55 years with male predominance (75% of cases) [15].

The diagnosis of the plasmocytoma is difficult to pose clinically and radiologically, only histopathological examination, associated with an immunohistochemical study confirms the diagnosis by highlighting plasmocyt cells that express the marker CD138 with concomitant cytoplasmic expression of kappa or lambda light chains [16].

In our case, the patient presented the kappa light chain, but there was no evidence of progression to multiple myeloma. Holland *et al.*, [17] states that lesion size, total protein levels in the serum, and the presence of a monoclonal peak at electrophoresis of serum proteins may have a prognostic value for identifying solitary lesions that would have evolved into multiple myeloma.

The most widely accepted treatment for EMP is chemotherapy with cyclophosphamide and melphalan, combined with steroids. Because these tumors are radiosensitive, radiotherapy is the treatment of choice for extrasosseous plasmocytomas of the head and neck [18]. Sometimes surgical resection can be considered [19].

The role of adjuvant chemotherapy is uncertain. This is recommended for tumors with a high risk of progression (size greater than 5 cm and high grade) [18].

Since extra-bone plasmocytomas are often born in the head and neck, curative radical surgery for this location is often mutilating and not indicated.

The prognosis of extramedullary solitary plasmocytoma depends essentially on the risk of transformation into multiple myeloma.

Dimopoulos *et al.*, affirm that the prognosis for patients with solitary extramedullary plasmocytoma appears to be better than for those with solitary bone plasmocytoma, because 70% of patients with solitary extramedullary plasmocytoma will not have disease recurrence after 10-years.

## CONCLUSION

Extramedullary plasmocytoma is a rare affection that can affect all areas of the body. He is especially located in the head and neck. His treatment is based on chemotherapy, radiotherapy or surgery for some locations. His prognosis depends essentially on the risk of transformation into multiple myeloma.

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