

## Case Report

# Extramedullary Plasmacytoma of the Waldeyer's Ring: A Case Report

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Submitted: 20 October 2014

Accepted: 03 November 2014

Published: 04 November 2014

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**OPEN ACCESS****Keywords**

- Extramedullary plasmacytoma
- Hematology
- Otolaryngology
- Radiation therapy

**Abstract**

The Extramedullary Plasmacytomas (EP) are rare tumors originating from plasmacells of the soft tissues outside of the bone. For a correct diagnosis biopsy is mandatory and CT or MRI is necessary.

We report a case of a 62 year-old man, who underwent to otolaryngologist visit for nose bleeding. A peduncolated right palatine tonsillar lesion was diagnosed. The nasopharyngeal examination revealed another mass extending inferiorly from the left fossa of Rosenmuller, confirmed by a facial and neck CT. An excisional biopsy was performed in both sites and the histological examination diagnosed a moderately differentiated extramedullary plasmacytoma.

Patient was referred to hematologist, which requires a 18F-FDG PET-CT that shows tracer accumulation only in the tonsillar region bilaterally. Bone scintigraphy and bone marrow biopsy were negative.

Finally, the patient was referred to our Department and submitted to radiation treatment. Therapy was performed with 3-D conformal technique. The total dose administered was 45 Gy in 25 fractions.

The EP is a radiosensitive disease. Several studies show an excellent local control and a long-term disease-free survival after radiotherapy, but there are still not standardized doses and volumes of radiation.

All retrospective reviews support radiation doses of 40-50 Gy, supporting irradiation of involved lymph nodes. Higher doses are indicated for high dimensions tumors (> 5 cm) or patients with high risk of recurrence. Studies on the role of surgery are conflicting and there is no evidence on the efficacy of adjuvant chemotherapy.

Patient was free from recurrence 30 months after the end of radiation treatment.

**ABBREVIATIONS**

EP: Extramedullary Plasmacytoma; CT: Computed Tomography; MRI: Magnetic Resonance Imaging; 18F-FDG PET-CT: 18 F-Fluoro Deoxy Glucose Positron Emission Tomography - Computed Tomography; 3-D: 3-Dimensional; Gy: Gray; QUANTEC: Quantitative Analysis of Normal Tissue Effects in the Clinic; CTCAE: Common Terminology Criteria for Adverse Events; RT: Radiation Therapy

**INTRODUCTION**

The Extramedullary Plasmacytomas (EP) are rare tumors originating from plasmacells of the soft tissues outside of the bone. The aetiology remains unknown but chronic irritation

(smoke, inhaled irritants) and viral pathogenesis has been suggested.

In the majority of cases patients are male, about 60 years old and the preferred site of disease is the head and neck region, even if it is virtually possible to observe localizations of extramedullary plasmacytoma in every district of body.

Because these tumors are very rare, and can be difficult to distinguish between non-Hodgkin lymphomas, an expert pathologist in the diagnosis of lymphoma is suggested.

Biopsy is mandatory. For a correct diagnosis, CT or MRI are necessary, such as the evaluation of secondary criteria (exclusion of multiple myeloma, including verification of monoclonal

proteins in the serum and / or urine, the bone marrow biopsy and the absence of bone lesions).

## CASE PRESENTATION

We report a case of a 62-year-old man, diabetic, who underwent on october 2011 to otolaringologist visit for nose bleeding. A peduncolated right palatine tonsillar lesion (maximum diameter: 2,3 cm) was diagnosed. The nasopharyngeal examination revealed another mass extending inferiorly from the left fossa of Rosenmuller. No lateral cervical lymphadenopathy was found by palpation.

A facial and neck CT confirmed what was observed during the visit (Figures 1 and 2), reporting an hyperplasia of the right palatine tonsils.

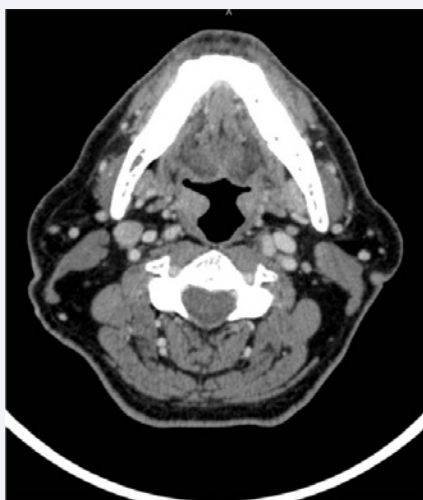
An excisional biopsy was performed in both sites and the histological examination diagnosed a moderately differentiated extramedullary plasmocytoma, with a coherent immunophenotype. The electrophoresis of serum proteins and urine assays for Bence Jones protein were negative.

The patient was referred to the specialist in hematology, which requires a bone marrow biopsy, that was negative and a 18F-FDG PET-CT that shows accumulation of the tracer only in the tonsillar region bilaterally (reported as reactive). Staging was completed with bone scintigraphy (negative).

Finally, the patient was referred to our Department and submitted to radiation treatment. Therapy was performed with 3-D conformal technique (Figure 3). Five convergent fields were used for the Waldeyer's ring region, according with Bellinzona technique and two antero-posterior opposed fields for neck irradiation.

The total dose administered was 45 Gy (1.8 Gy / fr) in 25 fractions.

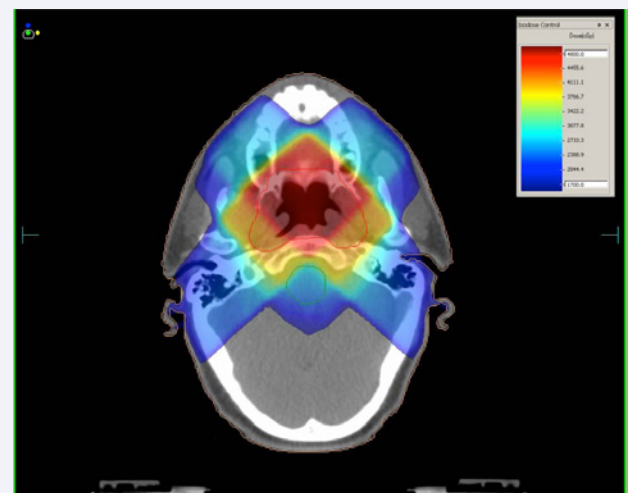
The treatment field included the anatomical structures of Waldeyer's ring, retropharyngeal lymphnodes and the levels II, III, IV and V bilaterally (total neck). All organs at risk's dose



**Figure 1** Facial and neck CT image showing the right palatine tonsillar lesion.



**Figure 2** Facial and neck CT image showing the tumor obliteration of the left Rosenmuller's space.



**Figure 3** Treatment plan and dose distribution (color wash).

constraints were respected according to the QUANTEC table. Visits during treatment were weekly. In the final part of therapy, patient developed grade 1 xerostomia (CTCAE v. 4.03).

The first clinical control took place three months after the end of radiotherapy. The general clinical conditions were good with a skin hyperpigmentation of the neck. Blood tests were normal and xerostomia was gone. Reported a slight change in taste. Patient was free from recurrence 30 months after the end of radiation treatment.

## DISCUSSION

The extramedullary plasmacytoma is a very sensitive disease to radiation therapy.

Although several studies continue to show an excellent local control and a long-term disease-free survival after radiotherapy, there are still not standardized doses and volumes of radiation.

Tournier-Rangard et al [1] compared patients treated with different doses. Five year local control was 90% for patients who received  $\geq 40$  Gy and 40% for those who received  $< 40$  Gy. The

survival rates (5 years) were 87.5% and 37.5% respectively for patients who received  $\geq 45\text{Gy}$  or  $< 45\text{Gy}$ .

This confirms what has already been shown by Chao et al. [2], which obtain 100% of local control in patients treated with a median dose of 45Gy (range 40-50.4Gy).

All retrospective reviews support radiation doses in the range 40-50Gy, supporting irradiation of involved lymph nodes. In particular, for EP of Waldeyer's ring should be planned the elective nodal irradiation to the first echelon cervical nodes (recommendation: grade B, evidence: level III) [3].

Higher doses (50 Gy in 25 fractions) are indicated for high dimensions tumors (EP > 5 cm) and for patients with high risk of recurrence. About 5% of patients undergoing radiation therapy experience local recurrence.

Studies on the role of surgery are conflicting. Some define as acceptable the combination of radiotherapy and surgery [4], while for others it produce a better outcome according to survival rates [5].

Since EP is mainly an head and neck region disease and radical surgery is generally a mutilating procedure, it should be treated with definitive radiotherapy [6].

Complete surgical removal should be considered for patients with localizations in other areas, reserving radiation treatment in case of inadequate surgical margins [7].

There is no evidence on the efficacy of adjuvant chemotherapy. This can be useful in selected patients with high risk of progression (tumor greater than 5 cm, high-grade histology or unresponded patients to RT) [8]. For these patients must be considered the treatment schedules used in multiple myeloma, also considering that approximately 10-25% of patients progress to myeloma at 5 years [1]. The progression of the disease can also occur as solitary bone plasmacytoma, lymphadenopathy or as infiltration of the soft tissues and subcutaneous.

The use of bisphosphonates for EP is not indicated and

thalidomide has been used with highly variable results. Its use is based on case reports only.

Regardless of adopted treatment two-thirds of patients survive for more than ten years.

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## Cite this article

Emanuele Liardo RL, Spatola C, Bevilacqua R, Privitera G (2014) Extramedullary Plasmacytoma of the Waldeyer's Ring: A Case Report. *JSM Clin Case Rep* 2(6): 1069.