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EXTRAMEDULLARY PLASMACYTOMA OF THE TONSIL: A NEW MANAGEMENT

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ABSTRACT

Introduction*: Extramedullary plasmacytoma (EMP) is a rare tumor of all plasma cell neoplasms. The tumor is mainly localized in the head and neck region, but rarely involving the tonsil.*

Case presentation*: The authors report the 5th case of EMP of the tonsil in the literature occurred in a 57-year-old Caucasian male.*

Conclusions*: Through a review of the relevant literature, we consider adjuvant radiotherapy not necessary for EMP of tonsil because of an adequate resection achieved by surgery.*

Key words*: plasmacytoma, extramedullary plasmacytoma, tonsil.*

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Introduction

Plasmacytoma is malignant proliferation of a monoclonal population of plasma cells all producing the same immunoglobulin. This plasma cell malignancy accounts for <1% of all cancers and for 10% of the hematological malignancies[(1)]. Usually this tumor is located in bone marrow as solitary bone plasmacytoma (SBP), although sometimes it can be found in soft tissue as extramedullary plasmacytoma (EMP) or be a part of the multifocal disseminated disease multiple myeloma (MM)[(2)]. Particularly EMP accounts for less than 3-4% of all plasma cell neoplasms and for 4% of all non-epithelial tumors of the upper airways[(3)].

The median age of EMP onset is 55 years and 75% of affected patients are men. In the 80% of cases it is localized submucosally, mainly in the head and neck region, particularly in the nasal cavity and/or paranasal sinuses (43%), nasopharynx (18.3%), oropharynx (17.8%) and tonsils (<10%). There are no identifiable risk factors[(4,5,6,7,8,9,10)].

Symptoms and signs depend on the EMP's location, but the most frequent are pain, swelling and obstruction of the respiratory tract or oral cavity.

The authors describe the 5th case of EMP of the tonsil reported by literature making a review with respect to evaluation and management of this tumor.

Case presentation

A 57-year-old Caucasian male worker was admitted to our section on May 2009. He was lifelong nonsmoker and denied alcohol consumption. He complained a painless right-upper neck mass rapidly increased in size during the last month. The

patient did not report other disorders; in particular his family and medical history was unremarkable, he denied loss of weight, other masses, swallowing or breathing difficulties. Physical examination revealed only a palpable, freely mobile and non-tender right level II lymph node comprise between 2 and 4 cm. Oropharyngeal examination showed a moderately enlarged right tonsil with mucosal telangiectasias and elevation of the lateral floor. No pulsations were detectable; flexible fiberoptic nasopharyngoscopy was normal.

CT (computerized tomography) of the neck evidenced ovoid nonspecific soft tissue masses, likely a lymph nodes (Figure 1a), and a mass in the right tonsillar fossa suspicious for a primary tumor (Figure 1b).

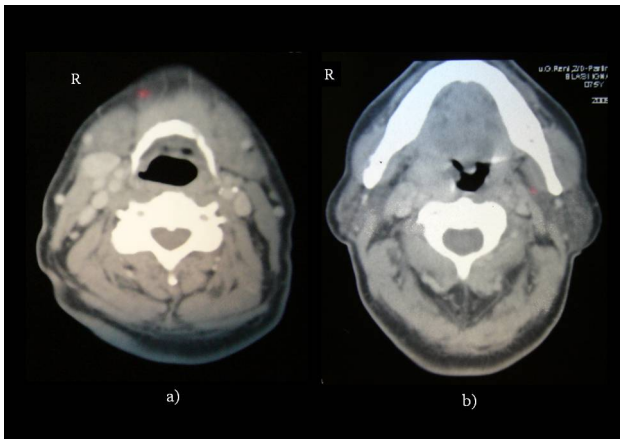

Figure 1. **a**) Axial Computerized Tomography, with contrast media, showing enlarged right level II lymph node that push the submandibular gland; **b**) Axial Computerized Tomography, with contrast media, showing a mass in the right tonsillar fossa suspicious for a primary tumor.

So CT imaging supported the clinical diagnosis of a tonsillar primary lesion with a regional lymph nodal involvement. Suspecting a malign lesion an excision biopsy (tonsillectomy) of right tonsil was performed (Figure 2a) and, after extemporaneous examination predicting for plasmacytoma, a neck dissection was carried out (Figure 2b).

Histological examination of the tonsillar mass showed dense plasmacells with a monotone aspect. They possessed a round to oval nucleus with a vesicular nuclear chromatin pattern. The nucleus is often located eccentrically in the cytoplasm (perinuclear halo). The tonsillar lesion was fairly circumscribed and the epithelium was not infiltrated.

The lymph nodes showed alteration of normal architecture and extensive infiltration of neoplastic cells possessing plasmacytoid morphology.

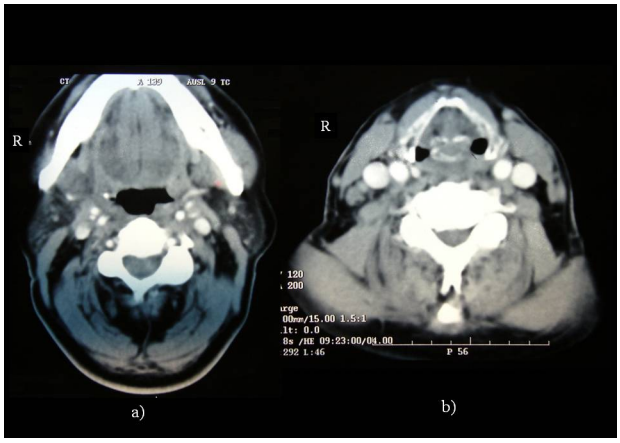

Figure 2. **a**) Axial Computerized Tomography, with contrast media, showing right tonsillar fossa free from primary tumor three years after tonsillectomy; **b**) Axial Computerized Tomography, with contrast media, showing the absence of lymph nodes three years after neck dissection.

Further immunocytochemistric analysis on the specimens showed CD138 plasma cell marker positivity with concomitant cytoplasmic expression of kappa light chains, while CD20 and CD3 were negative. This profile confirmed the diagnosis of plasmacytoma (Figure 3 a and b). Serum protein electrophoresis showed the presence of monoclonal IgG with kappa light chain with normal levels of residual immunoglobulins. Lumbar puncture, skeletal survey and Bence-Jones protein in urine were performed with anything relevant. After surgery, at 3 monthly intervals for the first year, the patient was controlled. Monoclonal IgG and kappa light chains plasma levels were normalized. Bone marrow biopsy resulted negative for multiple myeloma and skeletal survey was negative for coexisting lesions. The patient underwent new examination at 6 monthly intervals for the second year, and then annually; to date he remains free of disease.

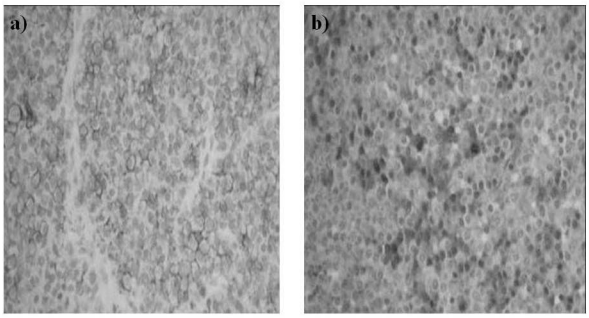

Figure 3. Tissue sampling of Extramedullary plasmacytoma (EMP) and immunohistochemical staining. **a**) Positive staining of CD138; **b**) positive staining for kappa light chains.

Discussion

The exact incidence of extramedullary plasmacytomas (EMP) is unknown, but it represents only 3-4% of all plasma cell malignancies[8]. In the 80% of patients, EMP occurs in the upperairways, with an involvement of the tonsil in 10.5% of cases[8].

So far, only 5 cases of tonsillar EMP have been described in the literature (Table I).

Cases	Years	Age	Sex	Side	Neck node	Light chain	Management	Follow-up
Kalan et al	2000	51	M	Right	No	Kappa	Radiotherapy	At 5 months
Bazaadut et al	2010	58	M	Right	Yes	Kappa	Tonsillectomy	At 1 year
Bhat et al	2010	43	M	Right	No	Kappa	Tonsillectomy	At 2 years
Huoh et al	2011	52	M	Left	No	Lambda	Tonsillectomy	At 6 month
Martines et al	2012	57	M	Right	Yes	Kappa	Tonsillectomy neck dissection	At 3 years

Table 1: EMPs of tonsil described in the literature.

Kalan et al reported a EMP of tonsil treated with radiotherapy alone (4500 cGY in 20 fractions)[11].

Bazaadut et al reported EMP arose from the right tonsil with involved ipsilateral lymph node as in our case. The lesion was removed by tonsillectomy but neck dissection was not performed because these authors make it when extemporaneous examination shows a squamous cell carcinoma[12].

The other authors reported cases of the same tumor, treated by tonsillectomy and/or neck dissection when lymph nodes were involved, without performing the examination extemporaneous[13,14].

As suggested by "guidelines on the diagnosis and management of solitary bone plasmacytoma and extramedullary plasmacytoma" (2004) the management of our case included full history of patient, clinical examination of nose, throat, neck region and nasopharyngoscopy[15]. Fine-needle aspiration biopsy (FNA) of the neck mass was not performed because it doesn't provide conclusive results because it doesn't distinguish with absolute certainty between inflammatory condition and malignant lesion[16]. In fact it is universally accepted that only pathology exam of primary lesion and/or lymph nodes associated, especially if accompanied by immunocytochemistric analysis[16], is able to confirm the diagnosis especially through positive plasma cells that express CD138 with concomitant cytoplasmic expression of kappa or lambda light chains[16].

Imaging studies such as CT, Magnetic resonance imaging (MRI) and Positron Emission Tomography (PET) can help evaluating the local extension of disease, involvement of lymph nodes and bone structures and the choose of adjuvant radiotherapy[4].

Baseline full blood examination, serum calcium, urea, and electrolytes should be performed in addition to serum protein electrophoresis and immunofixation, urine Bence-Jones proteins, skeletal survey, and bone marrow examination. In particular serum protein electrophoresis can show the presence of monoclonal IgG with kappa or lambda light chain with normal levels of residual immunoglobulins. As in this subject the commonest immunoglobulin evidenced is IgG with kappa light chain restriction. This serological profile is associated with the lowest rate of progression to MM, while cases with prevalence of lambda light chain are more likely to progress to MM. Solitary EMP management includes radiotherapy, surgery, or both techniques. Literature reveal no advantages of chemotherapy administration. EMP respond well to radiation therapy and some advocate use of radiation as primary treatment[2,11]. In contrast to Kalan et al, the authors suggest that if disease is localized in the head and neck and amenable to complete resection, surgery is advocated also because radiation therapy may involve major organs such as the thyroid and salivary glands with sequelae. In contrast to Bazaadut and to other authors, if extemporaneous examination predict for plasmacytoma and lymph nodal involvement is localized, the authors prefer to perform neck dissection because this tumor can metastasize; therefore complete surgical resection must be preferred to radiotherapy[15] even if some authors affirm that approximately 10-30% of treated patients distant failure seem to occur[15]. This fact usually occurs within 2 years after diagnosis[4]. For this reason, we recommend a complete reassessment every three months in the first year, every six month in the second year, and annually thereafter for life.

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