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Extramedullary plasmacytoma in the maxillofacial region: A review of the literature and case report

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A B S T R A C T

Extramedullary Plasmacytoma is an uncommon cell dyscrasia, representing 1% of all head and neck tumours. Characteristically, EMP is a slowly growing neoplasm with pattern of growth that delays its diagnosis. Secondary to this evolution, most patients present symptoms related to the location of the mass.

We present a case of EMP in the maxillofacial region in a 59 years old male, who was treated by our department.

Treatment approach is generally personalized to each case. These tumours often appear in complicated anatomic locations. EMPs are radiosensitive, and radiotherapy is often used as a first-line treatment to avoid disfiguring results and potentially eliminate the need for further invasive surgical procedures, but surgery combined with Radiotherapy has demonstrated good results in preventing local recurrence.

1. Introduction

Extramedullary Plasmacytoma (EMP) is a rare tumor who constitutes 3% of all cell plasma neoplasms [1].

Plasma cell neoplasms can be present as a single lesion (Solitary Plasmacytoma) or as multiples lesions in different locations (Multiple Myeloma). Solitary Plasmacytomas most frequently occur in bone but can also be found in other locations (EMP). Multiple myeloma (MM) is the most common neoplasm within this group [2].

The aim of this article is to present a case of EMP in the maxillary area treated by our department and make a review of this pathology in the literature.

2. Case report

We present a 69 years old male patient, with no other medical history, who was referred to our department with clinical history of right maxillary tumefaction.

The patient reported a 2-month history of a progressive mass in the upper maxillary ridge, with an aggressive pattern of growth (Fig. 1). He had no history of nasal obstruction, orbital symptoms, or facial pain. The patient denied any history of fever or any systematic disease (weight loss, loss of appetite …) during this period. Otherwise, his history was unremarkable.

The patient presented a bulk in the right upper maxillary which affected hard palate and oral mucosa with mobility of several teeth. He didn't present proptosis or any limitation of extraocular movements and no neurological alterations (Fig. 2). There was no clinical adenopathy.

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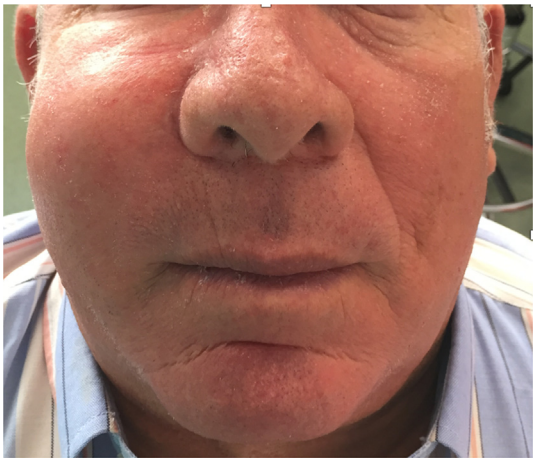

Fig. 1. Right maxillary tumefaction.

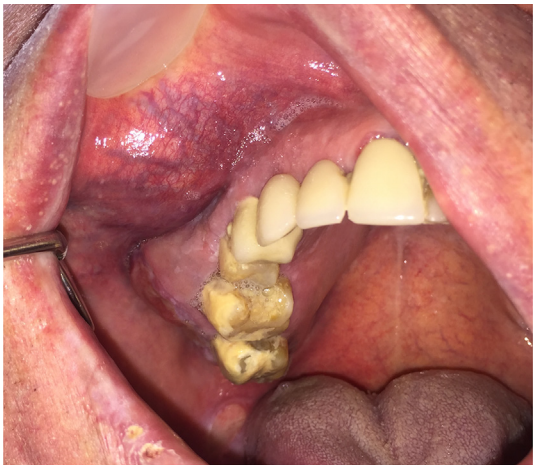

Fig. 2. Bulk in the right upper maxillary who affected hard palate and oral mucosa.

On computerized tomography (CT) and magnetic resonance imaging (MRI) examination a 5, 3 × 3 cm expansive mass lesion located in the right maxillary sinus, who invaded bone and infiltrated adjacent facial structures, was observed (Fig. 3).

A biopsy was performed. Microscopically, a tumor formed by spheric-ovoid neoplastic plasma cells with hyperchromatic nucleus some of which included marked nucleolus and with occasional paranuclear Golgi zones was observed (Fig. 4). Histopathological examination of the specimen showed an Extramedullary Plasmacytoma.

The blood results showed changes in free kappa chains (533mg/l) and free lambda chains (9,1 mg/l), with a Kappa/Lambda index of 58,53.

Staging investigations for a possible multiple myeloma included a bone marrow biopsy and a Positron Emission Tomography (PET) scan. Bone marrow punction showed 0,51% of plasmatic cells, with no clonal plasma cells. On PET-CT the radiologist reported a hypermetabolic mass in right maxillary sinus, with bone destruction of the adjacent areas and cervical lymph nodes with doubtful affectation, there was no evidence of disseminated disease.

The case was investigated by the hematologists and discussed at the multidisciplinary hematology meeting. Due to the high local aggressiveness and the radiological possibility of incipient medullar invasion, the hematologists decided to begin an initial cycle of Chemotherapy with Thalidomide, Bortezomib and Dexamethasone, trying to slow the growth of the mass, prior to surgery or radiotherapy.

During Chemotherapy, the patient presented several complications including: peripheral neuropathy in both legs and loss in high of vertebral bones. After the fourth cycle the patient presented an episode of dyspnoea who was diagnosed as Pulmonary Embolism. The Doppler confirmed the diagnosis of acute popliteal vein occlusion. The pause in the treatment induced a new regrowth of the mass after partial response. The committee decided radical radiotherapy, due to the physical situation of the patient after chemotherapy and Anaesthesiologist's contraindication for surgery.