

## Short Case Report

# Extramedullar plasmocytoma of the oral cavity

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(Received: 30 December 2019, accepted: 9 June 2020)

**Keywords:**  
Oral cavity /  
Extramedullar  
plasmocytoma /  
Multiple myeloma

**Abstract – Introduction:** Extramedullar plasmocytoma (EMP) is a plasma cell disease, such as solitary plasmocytoma of the bone and multiple myeloma. It is developed in the soft tissues. **Observation:** A 79-year-old patient consulted for a painful tumefaction of the gum associated to a radiolucent area on the right side of the mandible on the panoramic x-ray. After surgical enucleation, histological examination showed that the diagnosis was either an IgA extramedullar plasmocytoma or a multiple myeloma. The spreading assessment eliminated the diagnosis of multiple myeloma. The patient was then treated with radiotherapy. **Discussion:** EMP is a rare tumoral disease. Few cases described oral localisation. The main differential diagnosis, that must be excluded is multiple myeloma. Transformation in multiple myeloma exists. The first intention treatment is radiotherapy of the lesion. **Conclusion:** EMP of the oral cavity is very rare. After treatment, follow-up is required to detect second localisation or progression in multiple myeloma.

## Observation

A 79-year-old man presented to a unit of plastic, oral and maxillofacial surgery. His dental surgeon referred him after discovering a radiolucent area on the right side of the mandible on the orthopantomography. He had a history of persistent atrial fibrillation and was treated from March 11th 2016 to April 18th 2016 for an extramedullar plasmocytoma (EMP) of the right maxillar sinus. He received 50 Gy, 2 Gy per day, 5 times a week. Since then, he has been checked up twice a year by an oncologist. His last results were normal, except for a medium peak of IgG kappa, which stayed stable for months.

Facial examination showed no particularity. However intraoral examination revealed a painful tumefaction of the gum of tooth 47, without suppuration. The mouth mucosa was normal in appearance. The molar was repaired by a porcelain-fused-to-metal crown and had no mobility. The rest of the oral cavity was normal.

The panoramic X-ray (Fig. 1) revealed a large and well-delimited unilocular radiolucent area, next to the roots of tooth 47 which had received an endodontic treatment. That's why, a radicular cyst was initially suspected.

A cone beam CT (Fig. 2) was prescribed after the first consultation. It confirmed the presence of an unilocular radiolucent area on the right mandibular angle, in which the roots of the tooth were included. The bone cortical was partially

destroyed and the inferior alveolar nerve was not visible. Ameloblastoma or a radicular cyst was then suspected.

The enucleation of the lesion and the tooth extraction were performed under general anaesthesia. The lesion, which was located just under the gum, appeared to be very fibrous, grey and extra-alveolar.

The histological examinations' conclusion was leaning towards an IgA EMP or a multiple myeloma (MM).

Because of the former existence of a plasmocytoma of the maxillar sinus, a systemic disorder such as MM was suspected. The patient was referred to his oncologist for a spreading assessment to eliminate the diagnosis of myeloma. After several exams, EMP without systemic affection was confirmed. The multidisciplinary tumor board meeting concluded that a radiotherapy treatment of the lesions was necessary.

The patient received 50 Gy, 2 Gy per day, 5 times a week, on both, maxillar and mandibular areas, in April 2019. A recurrence of the mandibular plasmocytoma was found, In August 2019, but, the medical team decided to not proceed with a new surgery of the patient. Since then, he is followed by his oncologist, each 3 months, and is treated with corticosteroid (Dexamethasone<sup>®</sup>) and an immunomodulatory drug (Revlimid<sup>®</sup>).

## Comments

Plasma cell diseases are characterised by a tumoral proliferation of monoclonal plasma cells. These one could be either unique, inside the bone in case of solitary plasmocytoma

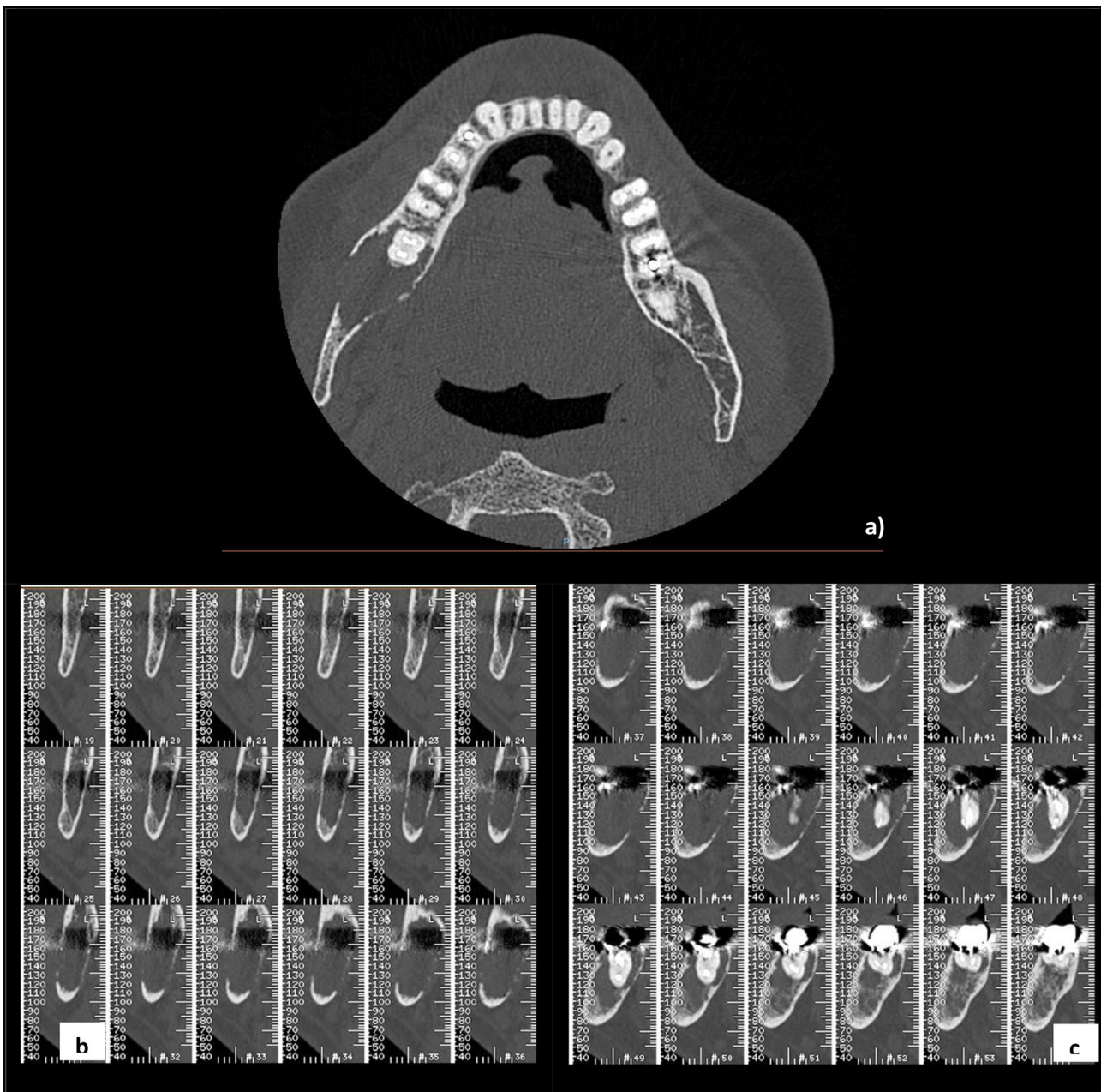
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**Fig. 1.** Intial panoramic X-ray showed an unilocular radiolucent area in the right side of the mandible.

of bone (SPB) or inside soft tissues, for the EMP, or associated with a bone marrow disorder in MM [1,2].

EMP is a rare tumoral disease. It represents 3% of plasma cell diseases and 0.4% of the head and neck tumours. It has a low incidence, lower than the SPB with 0.10/100000 against 0.15/100000 new cases per year [2]. It affects more frequently men during the sixth decade of life [1,2]. Plasmocytomas are located in almost 80% of the cases in the head and neck areas, particularly in the upper aerodigestive track; mostly in the nasal cavity, in the nasopharynx and in the maxillar sinus [1]. The abundance of lymphatic tissues in these areas explains



**Fig. 2.** (a-c) Cone beam CT. (b, c) Centered on the tooth 47.

these specific localisations. Tumoral proliferation is also found in the gastro-intestinal track, lungs or skin. Some are also described in the oral cavity.

An extension to an adjacent bone or to the cervical lymph nodes is also possible and it does not impact the diagnosis made of a solitary lesion [3].

The etiology of the disease is still misunderstood but some authors described contributing factors such as chronic stimulations, irradiations overdose, viral or genetic interactions inside endoplasmic reticulum [4]. The irradiation received by the patient in 2016 could have contributed to this second localisation.

The diagnosis must confirm the EMP with the help of biopsy and excludes the systemic affection, according to the spreading assessment which includes blood count, serum proteins electrophoresis, proteinuria, myelogram and X-ray examination.

Histological examinations must confirm monoclonal plasma cell infiltrations that express CD 138 and/or CD38 [5].

<sup>18</sup>F-FDG-PET-CT is the referral radiographic exam for the detection of medullar or extramedullar diseases if a MRI of the full body is not available. It detects bone lesion even at early stages [5]. Hypermetabolic activity of lymph nodes can also be revealed by this exam. In 7.6% of the cases, they are affected by the plasmacytoma [1]. However, there is no higher risk to develop a MM later. In first intention, the treatment is radiotherapy of the tumour with irradiation doses between 40 and 50 Gy and 2 cm margins [5]. When a surgery is possible, it must be followed by a radiotherapy. The association of those 2 therapies could decrease the risk of recurrence [5]. Secondary plasmacytomas are less responsive to radiotherapy but increasing the dose is not recommended. There is no consensus for the use of chemotherapy but its use in first intention is not recommended. Treatment response is assessed with a radiographic follow-up of the lesion, and serum and urine monoclonal protein rate assay. They must be stable over time [5].

A yearly follow-up is necessary. Although the transformation rate of EMPs is lower than the SPB's one, a study has showed that they have a worst 2-year-survival rate [5]. Several hypotheses have been suggested to assess the risk of MM development. However, only the persistence of elevated serum monoclonal protein levels after radiotherapy has been proven to be a prognostic factor for progression to MM, provided that it was detectable prior to treatment [5]. Persistence of moderate serum monoclonal protein rates after initial radiation therapy could explain the recurrence of plasmacytoma of our patient, even if there is no sign of MM for now.

**Conflicts of interests:** The authors declare that they have no conflicts of interest in relation to the publication of this article.

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