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Original research

Bone and soft tissue non-Hodgkin lymphoma of the maxillofacial area: Report of two cases, literature review and new therapeutic strategies

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1. Introduction

Extra nodal non-Hodgkin lymphoma (NHL) represent 20%–30% of all the NHL the second one occur in extra-nodal sites [1,2]. Among the NHL that occur in the oral cavity, 15–45% arise in the maxilla and mandible [3] being the upper jaw maxilla (11%), the mandible (8%), the palatal soft tissue (8%) and gum (7%) the most common locations. Isolated mandibular NHL accounts for only 0.6% of all the NHL. Soft tissues NHL of the maxillofacial area are even more rare representing a minimal percentage of the head and neck tumors [4]. The unique mandibular and soft localization is very rare for the NHL and in some cases, differential diagnosis with the most common dental lesions and other soft tissue pathologies may be difficult. Corresponding patients have to be treated with

ABSTRACT

Primary mandibular non-Hodgkin lymphoma (NHL) and soft tissues NHL of the maxillofacial are extremely rare representing a minimal percentage of the head and neck tumors. Two cases of bone and soft tissue maxillofacial NHL are reported. Clinical, radiological and pathological features are described and the therapeutic procedures are discussed accordingly. Mandibular radiologic features have been carefully analyzed and discussed to achieve an early and accurate diagnosis avoiding improper dental therapies.

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chemotherapy, radiation therapy or with the combination of both [5]. It is essential to diagnose the lesion at an early stage, paying attention to the differential diagnosis, particularly for unique mandibular localization because clinical signs of most common dental lesions may lead to misunderstanding of non-Hodgkin lymphoma diagnosis [6]. There have been some reports describing NHL [7–9] involving the mandible and soft tissue, but not all of them clarify all clinical and imaging aspects for early differential diagnosis. Our goal is to evaluate the clinical findings and radiographic images of NHL with mandibular and soft tissue localization with emphasis on differential diagnosis, in order to achieve immediately a right diagnosis avoiding improper therapy detrimental for prognosis.

2. Cases report

2.1. Case 1

A 53-year-old woman, without any history for other pathology, consulted our Department for left facial tumefaction with one year's evolution, with associated dental pain without any local signs of





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Abbreviations: NHL, non-Hodgkin lymphoma; B-NHL, B-cell non-Hodgkin lymphoma; CT, computerized tomography; FNC, fine needle cytology.

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Fig. 1. CT in coronal projection showing mandibular tumor involvement.

inflammation. Clinical examination revealed a pain-free hard mass involving the horizontal branch of the mandible, without any cutaneous involvement. At oral examination obstruction of the entire inferior vestibule was found. The patient didn't suffer from human immunodeficiency virus infection. She did not complain of bleeding, and no significant clinical data were present in her clinical history. Computerized tomography (CT) was available (Figs. 1 and 2). On panoramic Rx, the area affected by the lesion showed osteolysis with almost indistinct borders, root resorption of the 33, destructive tooth decay with bone resorption at the furcation of 36; these signs in association with the absence of inflammation, purulent discharge and pain were inclined to the diagnosis of a malignant tumor. Therefore it was decided to perform a CT scan and biopsy



Fig. 2. CT in axial projection showing mandibular tumor involvement.

immediately. On CT images, tumor involvement of the bone marrow cortex and of the surrounding soft tissue was reported. Subsequently to careful evaluation of these radiological findings, a fine needle cytology (FNC) was performed, in order to assess the proper diagnosis of the lesion. Based on the final diagnosis of B-cell non-Hodgkin lymphoma (B-NHL), she started chemotherapy according to the R-CHOP-21 regimen that was based on the original doses used by the GELA group and consisted of intravenous cyclophosphamide 750 mg/m, doxorubicin 50 mg/m, vincristine 1.4 mg/m (maximum dose 2 mg), rituximab 375 mg/m on day 1, and oral prednisolone 40 mg/m on days 1–5, administered every 21 days for a total of eight cycles [10]. The patient at the end of the eight cycles will be visited every two months in very close follow-up. To date, she has completed 3 cycles of therapy and clinical response was good with the disappearance of the mandibular swelling. If a recurrence or persistence of the disease will be detected, the patient will undergo IEV (Ifosfamide, Epirubicin, Etoposide) or DHAP (Cisplatin, Ara-C, Dexamethasone) therapy, with subsequent mobilization and autologous stem cell [11]. Before the second scheme of chemotherapy in case of recurrence of the tumor, a new procedure for testing tumor stem cells sensibility to the chemotherapy will be performed. Because of about 21 days need for test response and necessity to start for immediate chemotherapy at the diagnosis, this method was not used at the first cycle.

2.2. Case 2

A 76-year-old woman presented with a left parotid swelling in the left parotid area and a palpable ipsilateral submental lymph node. The patient reported that she had noticed the left side of her face growing progressively larger over the past 4 months. The patient denied any history of fever, night sweats, or significant weight loss. Medical and social history were not significant. Computerized tomography (CT) was available (Fig. 3). A fine needle cytology (FNC), performed on the periparotideal mass diagnosed an NHL, and the diagnosis was confirmed by the following surgical biopsy. Based on the final diagnosis of B-cell NHL the patient was treated



Fig. 3. CT in axial projection showing the soft tissue mass in the periparotideal area.

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