



## Diffuse Large B-Cell Lymphoma Involving of Face - A Case Report and a Brief Review

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### Abstract

The most common subtype of Non-Hodgkin's lymphoma is Diffuse Large B-Cell Lymphoma. Presentation of the extra-nodal type of Diffuse Large B-Cell Lymphoma is common in stomach, bone, CNS and liver but in a very rare condition, it can happen in the maxillofacial muscles accompanying neurologic symptoms. An 83 years old man with nasolabial area swelling was referred to Oral and Maxillofacial Surgery Department. After a thorough examination, soft tissue benign and malignant tumors were proposed. An incisional biopsy was done for the patient under local anesthesia. Diffuse Large B-Cell Lymphoma was reported by pathology department. Due to non-surgical approach for lymphoma masses, the patient was referred to the oncologist and was treated with R-CHOP chemotherapy protocol for 8 sessions. The mass was disappeared after 3 sessions of chemotherapy. After 4 years follow-up, no recurrence was seen. A thorough examination of pathologic lesions considering the rarities especially in head and neck region is the key factor for providing the patients appropriate treatment modalities and also preventing misdiagnosis and treatment.

**Keywords:** Non-Hodgkins's lymphoma; Extranodal lymphoma; B-Cell lymphoma; Face

### Introduction

Diffuse large B-cell lymphoma (DLBCL) is the most common type of the non-Hodgkin lymphomas (NHLs) and according to CDC statistics it accounts for 31% of lymphomas. It is an aggressive clinical situation that would be fatal if left undiagnosed or untreated. DLBCL can occur at any age but generally, develop in middle-aged and older adults. Most patients with DLBCL are diagnosed during the 7 and 8 decades of life, with a median age of 63 years. There is a slight tendency to the male gender. However, DLBCL affects females more often than males. Clinically, patients with this type of lymphoma usually present with advanced, often extranodal disease. Presentation of the extranodal type of DLBCL is common in stomach, bone, CNS and liver but in a very rare condition, it can happen in facial muscles. As described by Muralee Mohan C et al. [1] only five cases of NHL arising in the muscles of mastication and two cases involving the muscles of facial expression have been reported up to 2011. We could not find newly reported cases since then. Here, we report clinical, radiological and histopathological features of a rare extranodal DLBCL infiltrated in facial muscle while affecting facial and infraorbital nerves.

### Case Presentation

An 83 years Old man with chief complaint of swelling in the left nasolabial area was referred to Oral and Maxillofacial Surgery Department. The swelling was begun suddenly about 2 months ago with hemifacial paralysis and grows increasingly. The hemifacial paralysis was slightly improved spontaneously. The patient had been visited by a general practitioner for two times before referral and had received antibiotic (Cephalexin 500 mg, QID, for 10 days) but the swelling still has been increasing. Due to persistent swelling and unsuccessful treatment, the patient referred to our department. Patient's history showed mild hypertension and smoking for more than 50 y (25 pack/y). Extra and intraoral examination showed a firm mass in left nasolabial area and following characteristics were observed: approximate size 3 cm × 3 cm, not clearly defined borders, semi-mobile, no tenderness on palpation, normal skin and mucosa on the mass, no bleeding or any discharge history from mass, no bruit or murmur on auscultation, no lymphadenopathy, paresthesia of the area innervated by left infraorbital nerve and no organomegaly in abdominal Exam (Figure 1). In panoramic view, an ill-defined hazy radiopacity was seen in left anterior maxilla. A plain CT scan of the face showed a well-defined soft tissue density mass in left anterior maxilla, without bony

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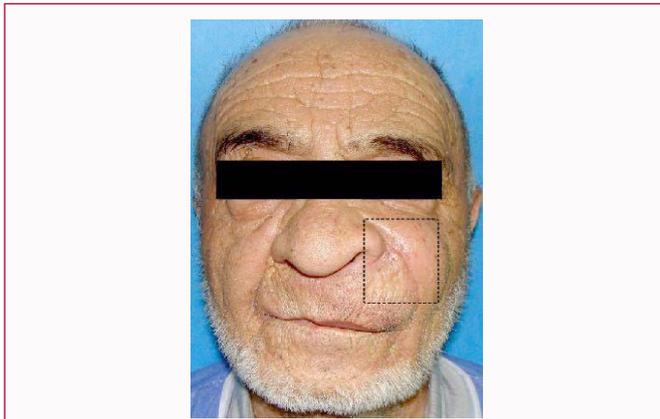
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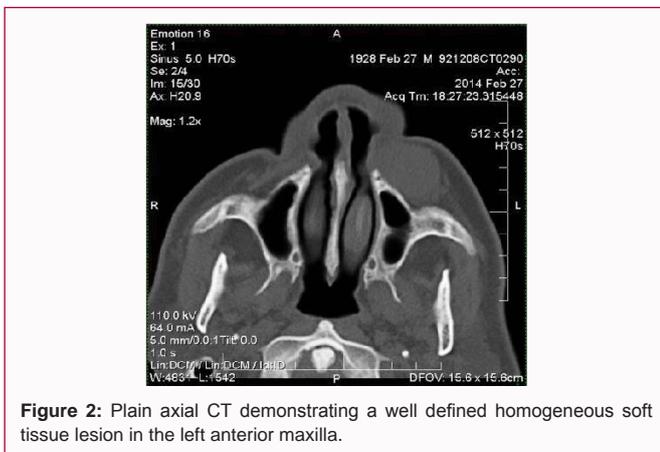
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**Figure 1:** Frontal view showing the location and extent of swelling in left nasolabial area.



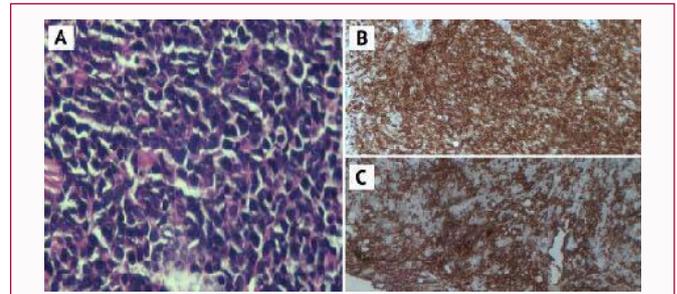
**Figure 2:** Plain axial CT demonstrating a well defined homogeneous soft tissue lesion in the left anterior maxilla.

erosion (Figure 2). Aspiration was negative for this lesion. Blood, serological and liver function tests were all within normal range. LDH and CRP were normal.

A provisional differential diagnosis list consisting benign and malignant soft tissue tumor was considered. This list consists: Peripheral Benign Mesenchymal Tumors (Schwannoma, Rhabdomyoma, Leiomyoma, Lymphangioma), Peripheral Metastatic Tumors, Peripheral Malignant Tumors (SCC, Malignant salivary gland tumor, Melanoma, Metastatic Carcinoma, Lymphoma, and Leukemia) Fine needle aspiration cytology of the lesion revealed lymphocytes and plasma cells suggestive of lymphadenopathy. A transoral incisional biopsy under local anesthesia was done. Histopathological examination of the specimen showed neoplastic infiltration in submucous, skeletal muscle and nerve fascicles. The neoplastic cells had multiple nuclei and vesicular, clefted nucleoli membrane with numerous mitotic and apoptotic activities (Figure 3).

The final diagnosis was malignant lymphoproliferative disorder, diffuse type, and high-grade oral cavity lymphoma. The further immunohistochemical analysis confirmed Diffuse Large B-Cell Lymphoma type, involving left anterior facial muscles and nerves. According to the Ann Arbour staging technique, it was stage I. As complimentary workup, chest and abdomen CT scans were done that indicated no other lesions.

Following the diagnosis and non-surgical approach for Lymphoma's lesions, the patient was referred to an oncologist. Before treatment, bone marrow aspiration that was done by the oncologist,



**Figure 3:** (A) Hematoxylin and eosin stain under high power field (x400) revealing neoplastic lymphocytes containing large nuclei with irregular nuclear membrane with prominent nucleoli and high mitotic index. (B) LCA marker strongly positive in neoplastic cells. (C) CD20 marker strongly positive in neoplastic cells showing diffused arranged B lymphocytes suggesting B-Cell lymphoma.

showed no bone marrow involvement by disease. He was treated with R-CHOP (Rituximab, Cyclophosphamide, Doxorubicin, Vincristine and Prednisolone) protocol for 8 sessions with 3-4 weeks interval according to the patient's compliance. Intraoral mass regressed completely after 3 sessions. At 4 years follow-up, the patient still remains disease free.

### Discussion

The diagnosis of the head and neck masses is complicated because of the variety of possible differential diagnosis list. It is critical for a physician to have insight about rarities such as extra-nodal lymphoma as a cause of maxillofacial swellings. Because of the aggressive nature of the lymphoma, the early diagnosis will lead to early appropriate treatment plan and an improved prognosis. Extranodal NHL especially involving the facial muscles and accompanying facial nerve paralysis is a very rare condition. As described by Muralee Mohan C et al. [1] up to 2011 in maxillofacial area only seven cases of primary NHL arising in the muscles have been reported. We could not find newly reported cases after 2011. Ceysens et al. [2] and Chong et al. [3] and Connor et al. [4] reported patients with extranodal non-Hodgkin lymphoma primarily infiltrating muscles of mastication. Harnsberger et al. [5] described masseter and medial pterygoid muscle lymphomatous involvement. Set et al. [6] described unilateral pterygomasseteric muscle complex involvement by primary non-Hodgkin lymphoma of the acute lymphoblastic type in a non-AIDS 6-year-old patient. Liapi et al. [7] reported a case of primary extranodal non-Hodgkin lymphoma involving only the muscles of facial expression.

Interestingly, Amo et al. [8] reported a 75-year-old patient with non-Hodgkin lymphoma affecting the facial muscles. Their case also had primary neurologic symptoms of trigeminal and facial nerves. Their patient as our patient presented with facial hemiplegia and paraesthesia.

One of the other important risk factors that can contribute in Hodgkin and non-Hodgkin lymphoma development, is Epstein-Barr Virus. According to the WHO, IVR department report, about 95% of the world adult populations are seropositive for EBV. As described by Heslop, EBV is associated with aggressive types of non-Hodgkin lymphoma, especially makes immunocompromised patients susceptible to B cell lymphoproliferative type due to outgrowth of EBV infected B cells, also she explained that lymphomas including NK and T malignancies, most occurs in individuals who do not have a known immunodeficiency [9].

The best approach in rare cases is multidisciplinary team approach including clinician, radiologist and pathologist. Clinical and paraclinical examinations of a patient have the most important role in preparing differential diagnosis list, and lesion's properties in imagings will be so helpful. As described by Beggs in ultrasound assessment, a solid, heterogeneous, hypoechoic mass with irregular or ill-defined margin can be seen [10]. In CT, a soft tissue density mass would be expected and in MRI you can see a hyperintense or isointense lesion relative to normal muscle on the T1-weighted sequence and hyperintense on the T2-weighted and fat suppression sequences [1]. Still, the biopsy is mandatory in order to diagnose exact type of the lesion.

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