

# Diffuse Swelling of Salivary and Lacrimal Glands as a First and only Manifestation of Non-Hodgkin B-Cell Lymphoma: A Case Report.

Amel Salah Eltayeb<sup>1</sup>, Abbas Abraham Alfadul<sup>2</sup>, Safa Abass Agaimy<sup>3</sup>, Ahmed M. Suleiman<sup>4</sup>

<sup>1</sup>Department of Oral and Maxillofacial Surgery, Khartoum teaching dental hospital, Khartoum, Sudan.

<sup>2</sup>Department of Oral and Maxillofacial surgery, Military Hospital, Khartoum, Sudan.

<sup>3</sup>Department of Pathology, Faculty of Dentistry, University of Gezira, Khartoum, Sudan.

<sup>4</sup>Department of Oral and Maxillofacial Surgery, Faculty of Dentistry, University of Khartoum Khartoum, Sudan.

Received: November 2016

Accepted: December 2016

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## ABSTRACT

Lymphoma is the third most common cancer worldwide. Oral cavity Lymphoma forms 3% of all lymphomas in the general population. This is a case report of non Hodgkin B-cell lymphoma in 60-year-old black male who was diagnosed earlier as having benign lympho-epithelial lesions of salivary and lacrimal glands. The patient was complaining of painless symmetrical facial swelling of the parotid, submandibular and lacrimal glands. Serological findings were negative for antinuclear antibody (ANA), anti-SS-A and anti-SS-B antibodies. Bilateral surgical excision of disfiguring buccal swollen salivary gland revealed a B-cell lymphoma. Chemotherapy started and dramatic response was noticed with total disappearance of all lacrimal and salivary gland swelling.

**Keywords:** lacrimal glands, lymphoma, lymph-epithelial lesions, salivary glands.

## INTRODUCTION

Lymphoma is the third most common cancer worldwide. It constitutes 3% of all malignant tumors and 2.2% of head and neck malignancies<sup>[1]</sup>. Lymphoma has two forms: Hodgkin's lymphoma (HL) and non-Hodgkin's lymphoma (NHL). Head and neck lymphomas are mainly non-Hodgkin's lymphoma with an incidence of 11–33%<sup>[2, 3]</sup>. The most common sites of growth are salivary glands, eyes, nasopharynx, maxillary sinuses and rarely the floor of the mouth and lips<sup>[4]</sup>.

### Name & Address of Corresponding Author

Dr. Amel Salah Eltayeb  
Dept of Department of Oral and Maxillofacial Surgery,  
Khartoum teaching dental hospital,  
Khartoum, Sudan.

Oral cavity Lymphoma forms 3% of all lymphomas in the general population and 4% in AIDS patients. It represents the third most common malignancy in the oral cavity<sup>[5]</sup>. Oral manifestations usually include asymptomatic soft swelling<sup>[6]</sup> that primarily affect the tonsils, palate, buccal mucosa, gingiva, tongue, floor of the mouth, salivary glands, and retromolar region.<sup>[7]</sup> Most cases of non-Hodgkin's lymphoma

arising in salivary glands are of B-cell origin including low-grade B-cell lymphomas of mucosa-associated lymphoid tissue (MALT), diffuse large B-cell lymphomas and follicular lymphomas.<sup>[8]</sup>

We report a case of a non-Hodgkin B-cell lymphoma (MALT) in a patient who was diagnosed earlier as having benign lympho-epithelial lesions of salivary and lacrimal glands.

## CASE REPORT

A 60-year-old black male presented to our clinic with painless symmetrical facial swelling of the parotid, submandibular, minor salivary glands and lacrimal glands. There was no history of dry eyes, conjunctivitis or dry mouth. His past medical history revealed tuberculosis from which he was treated three years ago. The patient was diagnosed earlier as having benign lymphoepithelial lesions of salivary and lacrimal glands based on lower lip salivary gland incisional biopsy and negative serological finding for antinuclear antibody (ANA), anti-SS-A and anti-SS-B antibodies. He was under treatment with steroids for a period of time with no noticeable response.

On examination, bilateral diffuse swellings of the lacrimal, parotid, submandibular and cheek regions were identified with normal overlying skin. Intraorally, all minor salivary glands of the palate,

buccolabial mucosa and the floor of the mouth were swollen. All the swellings were non tender and soft to firm in consistency. Conjunctivitis of the right eye was noticed. Cranial nerve assessment revealed no impairment of facial nerve function. Computed tomography (CT) demonstrated generalized enlargement of salivary and lacrimal glands with homogenous enhancement. Bilateral excisional biopsy of the enlarged buccal salivary glands that were interfering with eating revealed non-Hodgkin B- cell lymphoma [Figure 1]. This type of low-grade B-cell lymphoma has been recognized for many years, although it has only recently been categorized as a distinct clinico-pathologic entity in the Revised European -American Lymphoma (REAL) classification of lymphoid neoplasms where they called it 'Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphatic tissue (MALT) type'<sup>19</sup>. Immuno-histochemical staining was positive for CD20 and negative for CD3 and CD4 [Figure 2] which supported the diagnosis of non Hodgkin B-cell lymphoma.

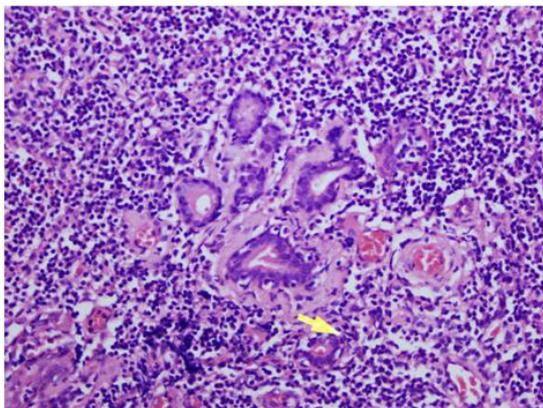


Figure 1: High-power photomicrograph shows lesional cells of lymphoma infiltrating an epimyoepithelial islands, the tumor cells consist of irregular shape of lymphocytes with minimal cytoplasm. ( hematoxylin-eosin stain).

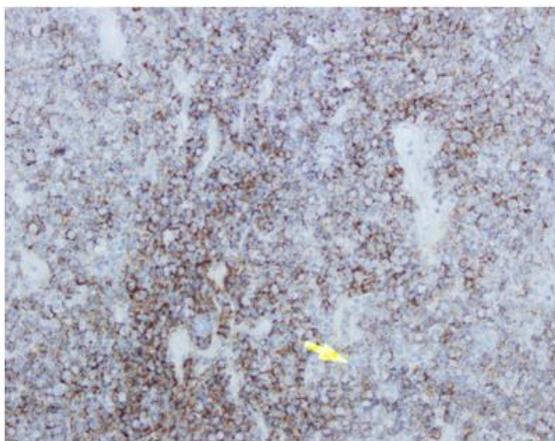


Figure 2: Immunohistochemistry staining for CD20 highlights diffuse infiltrate of CD20+ B cell lymphocytes.

The patient underwent chemotherapy with cyclophosphamide, vincristine and doxorubicin, along with prednisolone. There response immediately after the first cycle of chemotherapy was dramatic with complete disappearance of the lacrimal glands swelling [Figure 3], and all salivary glands swellings after 2 months [Figure 4]. Patient completed 6 cycles of chemotherapy. The patient did not have recurrent problems over a two year follow-up period [Figure 5].



Figure 3: (a) A bilateral lacrimal glands enlargement. (b) the same patient after chemotherapy with total disappearance of the lacrimal glands enlargement.



Figure 4: (a) patient presenting with bilateral enlargement of parotid and minor salivary glands of the cheek.(b) the same patient with normal appearance after chemotherapy.

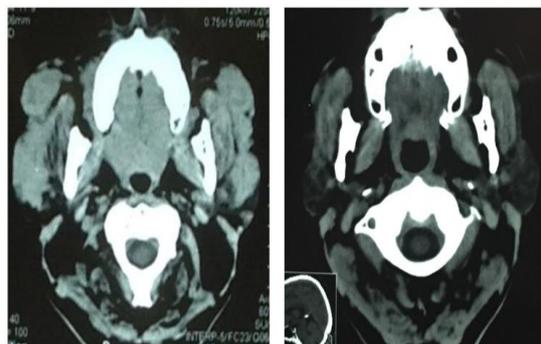


Figure 5: (a) Axial view CT-scan showing the enlarged salivary glands.(b) The same view after chemotherapy treatment with the lesions disappearance.

## DISCUSSION

Non-Hodgkin's lymphoma constitute 4–5% of all cancers.<sup>[10]</sup> It presents in extranodal sites in 40% of cases where the gastrointestinal tract is the most common site occurring in over 50% of patients, followed by the head and neck regions (11–33%).<sup>[11]</sup> The most recent World Health Organization (WHO) classification of lymphoma subdivides lymphomas based on cellular origin: B cell lymphomas, T cell lymphomas, natural killer lymphomas, and Hodgkin's lymphoma.<sup>[12]</sup> MALT lymphomas represent approximately 8% of all non-Hodgkin's lymphomas. The association of MALT lymphoma of the salivary gland, Sjogren's syndrome, and myoepithelial sialadenitis (MESA) has been studied, but some aspects are still controversial.<sup>[13]</sup> Primary salivary gland lymphoma is an uncommon neoplasm that usually occurs in the parotid gland and often shows bilateral involvement. The most common subtype is a low-grade B-cell lymphoma<sup>[14]</sup>. Lymphoma of the oral region has different clinical characteristics; it depends on the site involved, the lymph node involvement and/or the presence of metastasis. It presents as local mass, pain or discomfort, dysphagia or sensation of a foreign body in the throat.<sup>[15]</sup> In the present case, there were diffuse painless facial swellings. The enlarged minor salivary glands of the cheek were interfering with the patient's eating ability and involvement of the lacrimal glands lead to mild discomfort and conjunctivitis of the right eye. When a clinician is faced with diffuse salivary gland swelling, they do not anticipate primary Non-Hodgkin's Lymphoma (NHL) from the beginning since other inflammatory diseases with the same clinical presentation must be considered<sup>[16]</sup>. A good medical history, detailed clinical and imaging evaluations, and attention to the patient's signs and symptoms are crucial for the correct diagnosis and appropriate treatment.<sup>[2]</sup> In the present case, a diagnosis of B-cell lymphoma was suspected when a sudden increase in the facial swellings was noticed which indicated the possibility of emergence of a pathology other than the initial benign lymphoepithelial lesion. It is difficult to give a definitive diagnosis pre-operatively based on fine needle aspiration biopsy (FNAB)<sup>[17]</sup>, therefore surgical excision is preferable in such cases. A definitive diagnosis was obtained following surgical excision of the cheek enlarged salivary gland in the present study after which chemotherapy treatment was started. The main method of treatment of lymphomas, regardless of their primary location, is chemotherapy and radiotherapy.<sup>[16]</sup> Fortunately, the majority of MALT lymphomas are low-grade tumors that have a propensity to remain localized with good survival rates. Conversely, occasional tumors transform to higher-grade lymphomas with more aggressive behavior. Extra-

nodal B-cell lymphomas usually have a good prognosis with an 80% 5-year survival rate.<sup>[18]</sup>

## CONCLUSION

Clinicians should also be aware of the possibility of lymphoma in patients with benign lymphoepithelial lesions. Any changes in the nature and behavior of the swelling should alert the clinician to the possibility of lymphoma development. The difficulty in pre and intra-operative diagnosis often results in unnecessary radical operations and delay in treatment.

### Ethics approval and consent for publication:

Ethical approval was obtained from the research ethical committee of Khartoum teaching dental hospital.

### Consent for publication

Written consent was obtained from the patient for publication.

## REFERENCES

1. Epstein JB, JD E, M LN. Characteristics of oral and paraoral malignant lymphoma: a population based review of 361 cases. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod.* 2001; 92(5): 519-25.
2. Silva TDB, Ferreira CBT, Leite GB, Pontes JRdM, Antunes HS. Oral manifestations of lymphoma: a systematic review. *Ecancer.* 2016, 10:665.
3. Inchingolo F, al e. Non-Hodgkin lymphoma affecting the tongue: unusual intra-oral location *Head Neck. Oncol.* 2011; 3(1):1-5.
4. Nadendla KL, Meduri V, G. P. Imaging characteristics of diffuse large cell extranodal non-Hodgkin's lymphoma involving the palate and maxillary sinus: a case report. *Imaging Science in Dentistry.* 2012;42:111-4.
5. Levine AM. AIDS-related lymphoma: clinical aspects. *Semin Oncol.* 2000;27(4):442–53
6. Richards A, al e. Oral mucosal non-Hodgkin's lymphoma ± a dangerous mimic. *Oral Oncology.* 2000;36(6):556–8.
7. Ardekian L, al e. Burkitt's Lymphoma of the oral cavity in Israel. *J Cranio-Maxillofac Surg.* 1999;27(5):294–7.
8. Borovecki A KP, Ventura RA, Perisa MM, Banham AH, et al. MALT1, BCL10 and FOXP1 in salivary gland mucosa associated lymphoid tissue lymphomas. *Pathol Int* 2007; 57(1): 47-51.
9. Khari Lewis, Clive Vandervelde, Richard Grace, Keith Ramesar, Michael Williams, David C. Howlett. Salivary Gland Mucosa-Associated Lymphoid Tissue Lymphoma in 2 Patients with Sjogren's Syndrome: Clinical and Sonographic Features with Pathological Correlation. *J Clin Ultrasound* 2007; 35(2):97-101.
10. Frei M, Dubach P, Reichart PA, Schmitt AM, Muller-Garamvolgyi E, MM. B. diffuse swelling of the buccal mucosa and palate as first and only manifestation of an extranodal non-Hodgkin, double-hit, lymphoma; report of a case. *Oral Maxillofac Surg.* 2012;16:69-74.
11. Wulfrank D, al e. Extranodal non-Hodgkin's lymphoma of the head and neck. *Radiother Oncol.* 1987;8(3):199-207.
12. Harris NL, ES J, H S. A revised European-American classification of lymphoid neoplasms: a proposal from the International Lymphoma Study Group. *Blood.* 1994; 84(5): 1361–92.

13. Susan L. Abbondanzo. Extranodal Marginal-Zone B-Cell Lymphoma of the Salivary Gland. *Annals Diagnostic Pathology*. 2001;5(4):246-54.
14. Royer B, Cazals-Hatem D, Sibilia J, et al. Lymphomas in patients with Sjögren's syndrome are marginal zone B-cell neoplasms, arise in diverse extranodal and nodal sites, and are not associated with viruses. *Blood*. 1997;90:766-75.
15. M. Malaguarnera, M. Giordano, C. Russo, L. Puzzo, M. Trainiti, A.S. Consoli, et al. Lymphoma of cheek: a case report. *European Review for Medical and Pharmacological Sciences*. 2012;16(4):4-7.
16. Lewandowski L, MT. N. Non-Hodgkin's Lymphomas oral Cavity and Neck-diagnosis, treatment. *Poznańska Stom*. 2002; 24:15-8.
17. Francesco D GC, Gianluca M, Donatella M, Gautham K Primary Non-Hodgkins lymphoma of the parotid gland. *Braz J Otolaryngol*. 2011;77(5):639-44.
18. Bhattacharyya I, Chehal HK, Cohen DM, et al. Primary diffuse large B-cell lymphoma of the oral cavity: germinal center classification. *Head and Neck Pathol*. 2010;4(181-191)

**How to cite this article:** Eltayeb AS, Alfadul AA, Agaimy SA, Suleiman AM. Diffuse Swelling of Salivary and Lacrimal Glands as a First and only Manifestation of Non-Hodgkin B-Cell Lymphoma : A Case Report. *Ann. Int. Med. Den. Res*. 2017; 3(1):DE01-DE04.

**Source of Support:** Nil, **Conflict of Interest:** None declared