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**CASE STUDY**

# NON – HODGKIN'S LYMPHOMA OF MAXILLOFACIAL REGION: A DIAGNOSTIC CHALLENGE

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**ABSTRACT**

Malignant lymphomas of lymph nodes are rare entities in maxillofacial region. The extranodal Hodgkin's and Non- Hodgkin's lymphomas account for 10 – 20 % of all lymphomas; with an incidence of 0.1 – 5% in oral cavity. Those involving salivary glands are uncommon, and account for less than 5% of lymphomas with parotid gland being most frequently involved. These neoplasms pose a great challenge for clinicians and their prognosis is based on early clinical diagnosis and immunohistochemistry. This is a rare case report of Non – Hodgkin's lymphoma in a 40 years old male which was initially misdiagnosed as an inflammatory lesion.



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## INTRODUCTION

Lymphomas are malignant neoplasms of lymphocyte cell lines; which include Hodgkin's and Non - Hodgkin's type (NHL). NHL comprises a heterogeneous group of lymphoid neoplasms with a spectrum of behavior ranging from relatively indolent to highly aggressive and potentially fatal. (Shashidara *et al*., 2014) They are segregated into B – cell and T – cell types based on the cell of origin, where the former accounts approximately 90% and latter around 10% of all cases. Non-Hodgkin's lymphomas usually manifests outside the lymphoid system involving skin, abdomen, lungs and CNS. (Nagalaxmi *et al*., 2013) They are the third most common neoplasms of oral and maxillofacial region and account for 1.7% of all reported salivary neoplasms. (Revanappa *et al*., 2013) Majority of these develop in parotid glands (76%), but may develop in submandibular (20%), sublingual (3%) and palatal glands (1%). In 10% of the cases, involvement of more than one gland is seen. (Soni *et al*., 2014) Though the exact etiology of lymphomas is unknown; genetic predisposition, immunodeficiency states, and chromosomal translocation have been implicated as predisposing factors. (Nagalaxmi *et al*., 2013) Non – Hodgkin's lymphomas usually present as a painless, progressive enlarging mass with superficial ulceration; which is often indistinguishable from other non-malignant or more common epithelial tumors. (Soni *et al*., 2014; Singh *et al*., 2015) Hence diagnosis of lymphoma poses a great challenge for the clinicians. The prognosis is usually favorable; however, it depends upon the clinical stage and histological subtype of the tumor. Here we present a rare case of Non – Hodgkin's lymphoma involving submandibular region in a 40 year old male which was initially misdiagnosed as an inflammatory lesion on the basis of clinical, radiological and histological findings; but finally proved to be a B cell lymphoma based on immunohistochemistry.

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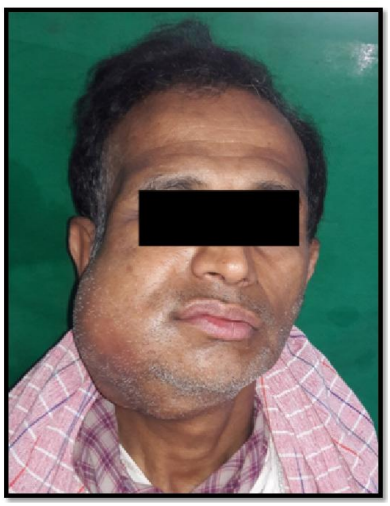
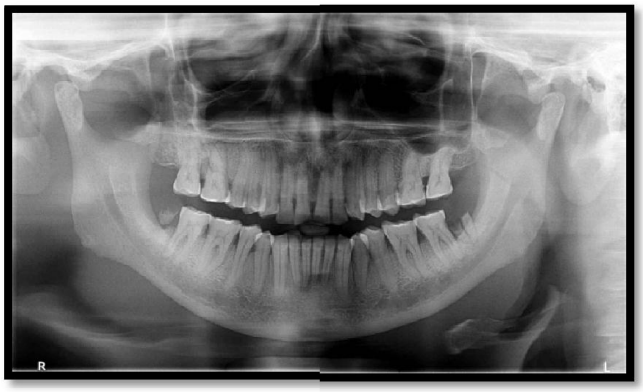
## CASE REPORT

A 40 years old male visited Oral Medicine clinic with a painful swelling of right side of face. In the history of presenting illness, the swelling started one and a half month ago with an insidious onset; and was associated with fever, weight loss with gradual progression of the swelling. There is a history of laparotomy for the abdomen mass 15 years back, details of which are not available. On general physical examination, the patient is an adult male, apparently healthy with normal built. Extraoral examination revealed a diffuse large swelling on the right side of face which was more bulky in parotid and submandibular gland region and extended anteroposteriorly from lateral aspect of nasal fossa to right ear lobule and superoinferiorly from right infraorbital region to the level of hyoid bone, measuring roughly 12 x 8 cm in size. The swelling was highly tender with elevated surface temperature and

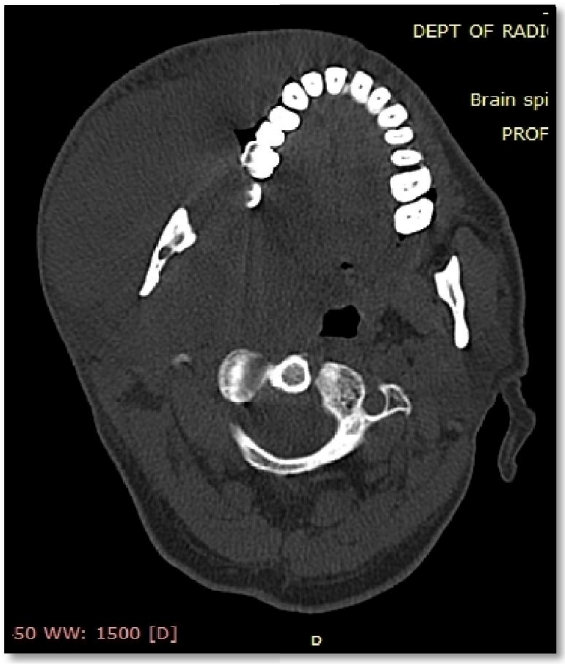
restricted mouth opening. It was uniformly firm in consistency with no significant enlargement of submandibular and cervical lymph nodes. On intraoral examination, a full complement of teeth was present except for the root stump irt 48 and a soft tissue swelling with obliteration of buccal vestibule irt 44 – 48 region. Based on the above findings, a provisional diagnosis of an acute dentoalveolar abscess irt 48 was considered with a differential diagnosis of space infection involving submandibular, buccal and sub – masseteric space, parotid gland abscess and malignancy arising from salivary glands. All the blood parameters were within normal limits and the antibodies for HIV I and II were non-reactive. Panoramic radiograph revealed presence of root stump irt 48 with no evident periapical pathology. The diagnosis of acute dentoalveolar abscess was considered and the patient was administered a course of Tab. Augmentin 625 mg, twice a day for 5 days. On evaluation after 3 days of antibiotic therapy, the patient did not show any response. Following this, ultrasonography of right neck and parotid region was done using 12 – MHz linear probe and a 5 – 7 – MHz convex probe; which revealed lobulated, hypoechoic solid extensive mass lesion with echogenic septations in the right parotid and submandibular gland region, which was more suggestive of a malignant lesion. To rule out the possibility of malignancy, FNAC was done wherein bimodal population of small round cells with scant cytoplasm was observed, with features suggestive of small round cell tumour. Further, CT scan with contrast was carried out to assess the extent of the lesion wherein a fairly defined heterogeneously enhancing mass in right masticator space with extension into surrounding tissues was observed, favoring the diagnosis of neoplastic lesion. As the lesion was a heterogeneously enhancing mass, MRI of neck was planned which reported an ill – defined peripherally enhancing T1 isointense and T2 hypointense lesion in right side of neck anterior to sternocleidomastoid muscle and carotid vessels involving right submandibular region, suggestive of an abscess involving sub – masseteric space. In view of the above findings, culture and sensitivity test of the smear was done; following which parenteral Cefotaxime (1g q8hrs) was administered. As the condition worsened with increase in size of the swelling and the clinical, radiological and histological findings were conflicting, incisional biopsy of the lesion was planned. The cut surface of the swelling showed greyish white areas intermixed with areas of necrosis.



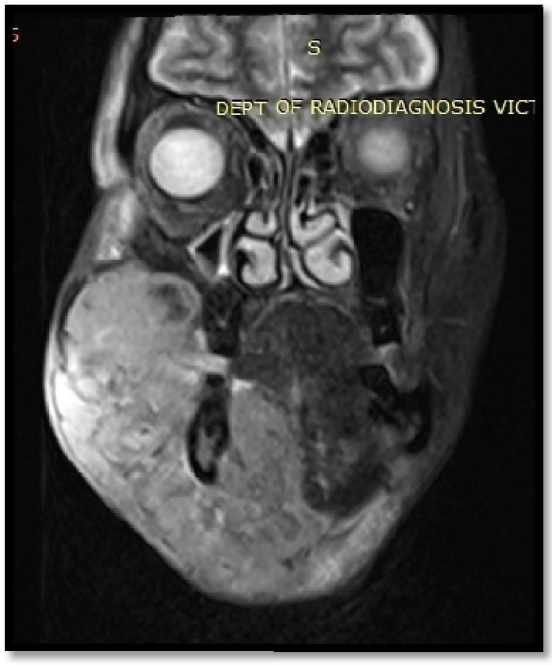
**Photograph 1. Front profile of the patient showing a diffuse swelling of right side of face which is more prominent in parotid and submandibular gland region**



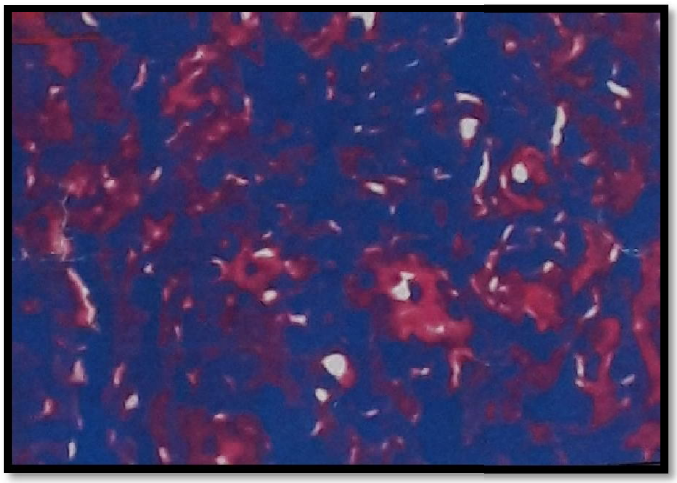
**Photograph 2. Panoramic radiograph showing root stump irt 48 with no evident periapical pathology**




**Photograph 3. CT showing a fairly defined heterogeneously enhancing mass in right masticator space**




**Photograph 4. MRI of neck showing an ill – defined peripherally enhancing T1 isointense and T2 hypointense lesion in right side of neck**



**Photograph 5. H/E stain of the biopsy specimen showing lymphocytic infiltrates, consisting of small and large lymphocytes, mixed with few macrophages**

Surprisingly, the microscopic examination of the specimen revealed large atypical lymphoid cells with areas of tumour necrosis, suggestive of Non – Hodgkin's lymphoma. Immunohistochemistry was performed to confirm the diagnosis wherein the neoplastic lymphoid cells expressed CD – 20, CD – 79a, CD – 10 and were immunonegative for Mum – 1, Bcl – 6, and Bcl – 2. The Ki – 67 index was found to be approximately 80%. Diagnosis of B – cell lymphoma, NOS, germinal centre type was concluded based on radiological and histological investigations, supported by immunohistochemical positivity for CD20 marker. The patient was referred to Department of Oncology for further management.

## DISCUSSION

Non – Hodgkin's lymphomas include a diverse and complex group of malignancies of lymphoreticular histogenesis and differentiation. The expression Non - Hodgkin's lymphoma covers a wide group of lymphoid neoplasias having tendency to affect organs and tissues that do not contain lymphoid cells. (Inchingolo *et al*., 2011) About 25-40% of NHLs are extranodal in origin and usually manifest in the gastrointestinal tract, followed by head and neck region. (Samanta *et al*., 2014) Only 3 – 5% of cases occur intraorally, whereas salivary gland involvement is seen in less than 5%. Non – Hodgkin's lymphoma occurs primarily in adults, such as in the present case of a 40 year old male. Although the exact etiology is unclear; they are frequently seen in patients with acquired immune deficiency syndrome (AIDS) and might serve as the first presentation of the syndrome. (Revanappa *et al*., 2013) However the present case was seronegative for HIV antibodies. Lymphomas present as firm to hard swelling of the affected gland which is usually of short duration. In this case the patient presented with a rapidly progressive, painful, firm swelling of the face involving mainly the parotid and submandibular gland region. Since there were no signs and symptoms suggestive of involvement of salivary glands, a diagnosis of dentoalveolar abscess was considered and patient was administered a course of antibiotics. As the response to antibiotics was not appreciated, the possibility of a malignancy arising from salivary glands was considered. To rule out malignancy, the patient was subjected to investigations like FNAC, CT scan and MRI. Because the radiological and cytological reports were conflicting, an incisional biopsy of the lesion was planned. On gross examination lymphomas are usually well-circumscribed with grayish white to pink-tan cut surface and soft to firm, rubbery consistency. (Agale *et al*., 2010) The biopsy specimen we received was solid, nodular, showed grayish white areas intermixed with yellowish necrotic areas.

The high-grade B-cell lymphoma must be differentiated from other poorly differentiated malignant tumors, particularly lymphoepithelioma like carcinoma and melanoma. Immunohistochemistry plays an important role in this regard. The positive expression of CD – 20, CD – 79a, CD – 10 and negativity for cytokeratin provided confirmation in our case. The diagnosis of NHL in maxillofacial region is a challenge as there is divergence in clinical, radiographic and histological patterns. One should suspect this neoplasm when there is a mismatch between radiological and histological findings. (Singh *et al*., 2015) Based on the morphology, cell lineage, and immunohistochemical findings, the present case was categorized as Diffuse large B – cell lymphoma, NOS, germinal centre type. The diagnosis of these tumours can be made precisely by applying clinicopathologic criteria as well as a panel of IHC markers or genetic studies so as to facilitate prompt and accurate therapy. Lymphomas of the salivary glands are highly chemo-radiosensitive, therefore, an early and timely diagnosis is important. (Dey *et al*., 2016) Therapeutically, complete tumor resection with addition of chemotherapy is the treatment of choice. Recently, the use of rituximab; an anti CD 20 antibody has emerged as a treatment option in patients with lymphomas. (Soni *et al*., 2014) The prognosis of the disease is usually good with an estimated 5 year survival rate of 30 % following therapy. Survival is excellent in localized diseases, whereas less favorable in disseminated cases. Moreover, patients older than 60 years, with stage 3 and 4 and severe extranodal involvement carry an unfavorable prognosis. (Nagalaxmi *et al*., 2013) Lymphomas should always be considered in differential diagnosis of lesions in maxillofacial region though their incidence is low in seronegative patients. The patients with oral manifestations of NHL might present at the dental clinic in the first instance. (Dave *et al*., 2015) Thus, with the rising incidence of extranodal lymphomas, it has become important for today's dentists to properly investigate the pathology and treat it judiciously. (Parihar *et al*., 2013) Hence, oral physicians can play an important role in early detection of the disease, thus improving the prognosis. (Dave *et al*., 2015)

### Conclusion

Although salivary gland lymphomas are rare; their possibility should be kept in mind when dealing with the swellings of parotid and submandibular glands. The present case emphasizes the importance of imaging and prognostic markers, which is essential for diagnosis and management. Awareness and scrupulous knowledge of dentist is essential in such cases for prompt diagnosis and immaculate treatment, all of which enhance the probability of existence.

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