CASE REPORT

Follicular lymphoma of the submandibular salivary gland

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ABSTRACT

Lymphomas are neoplastic diseases of lymph nodes. Lymphoma of the salivary gland is rare accounting for less than 5% of lymphomas overall. Furthermore, lymphomas arising in the submandibular gland are reported to comprise 916% of all salivary gland lymphomas. Among lymphomas originating from salivary glands, the ratio of follicular lymphoma is very low. They can also be seen in the lymph nodes of the salivary glands which is an uncommon presentation. Here, we present a case follicular lymphoma which presented as a salivary gland tumour.

Key words: Follicular lymphoma, Non Hodgkin`s lymphoma, submandibular salivary gland

INTRODUCTION

Salivary gland neoplasms constitute an important area in the field of pathology of head and neck region. The most common site for salivary gland tumour is the parotid gland. Majority of salivary gland tumours are parenchymal in origin. But, tumours of the mesenchymal origin have also been reported. One such neoplasm is lymphomas.

Lymphomas are malignant neoplasms of the lymphocyte cell lines. They are mainly classified as either Hodgkin's or Non-Hodgkin's lymphoma (NHL). NHL comprises a heterogeneous group of lymphoid neoplasms with a spectrum of behaviour ranging from relatively indolent to highly aggressive and potentially fatal.[1]

The vast majority of NHL represent B cell and less commonly T cell lineage neoplasm.[2] NHLs of B cell origin are known to be relatively common in extra nodal sites and many are thought to originate from mucosa associated lymphoid tissue (MALT).[3]

Follicular lymphomas accounts for one third of NHL.[4] This is a low to intermediate grade lymphoma that show a follicular architecture and represents the neoplastic counterpart of germinal center B lymphocytes. Follicular lymphoma is very uncommon under the age-group of 40 years and is extremely rare in children. Male is to female ratio is 1.3:1.[2] Somatic hyper mutations responsible for high intraclonal diversity, is characteristic of this malignancy.[5]

The t (14;18) (q32;q21) chromosome translocation has been purported to be the cytogenetic hallmark of follicular lymphoma. The molecular consequence is deregulation of Bcl2 expression leading to over expression of Bcl2 proteins in neoplastic follicles.[6]

Primary NHL of the salivary glands are relatively uncommon.[3,7,8] Here we present a case of Primary NHL involving submadibular salivary gland.

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CASE REPORT

A 40-year-old female patient reported to our institute with a complaint of swelling in the left side of lower jaw since 2 months [Figure 1]. Post its onset, the swelling gradually increased in size over a period of 2 months. The swelling was associated with dull pain. The patient is a known diabetic since 5 years and is under oral anti diabetic medication. Concurrent with the history of swelling, the patient gives a history of weight loss in past 2 months. No history of cold, cough or fever.

Examination revealed a swelling of 9 × 4 cm in size which extended anteriorly 1 cm in close proximity to chin on left side and posteriorly in line with the ear lobe [Figure 2]. Skin

over the lesion was pinchable, normal in color and texture. Margins were well-defined and the swelling was firm, fixed to underlying tissues and non tender on palpation. There were no signs of paresthesia. No local rise of temperature was seen in the concerned area. Left submandibular lymph node was 0.5 cm in size and mildly tender on palpation. A provisional diagnosis of benign tumour of left submandibular salivary gland was given. Few differential diagnoses such as Sarcoidosis, Sjogren`s syndrome, Lymphomas and Mikulicz disease were considered. Sarcoidosis was ruled out because only one of the submandibular glands was involved. Generally, all the salivary glands are involved. Sjogren`s syndrome was ruled out as there was no history of dryness. In both Sarcoidosis and Sjogren`s syndrome, the patient did not give any relevant history. Mikulicz disease is an autoimmune disease and thus was ruled out.

The following investigations were done:

- FNAC gave an impression of chronic non specific lymphadenitis. No evidence of cytological atypia seen
- Ultra sonography report showed a circumscribed mixed area of 3 × 1.5 cm in the anterior region of left submandibular gland. Moderate blood flow was seen and the region showed two enlarged lymph nodes. The impression was that of a questionable mass of submandibular gland.

The patient was further subjected to CT scan of head and neck and the impression was of

- Submandibular salivary gland tumour
- Multiple submental and submandibular lymphadenopthy.

Based on all these findings, a diagnosis of benign tumour of the left submandibular gland was given. During excision of the tumour, it was found that whole of the gland was involved. So, total submandibular gland excision was done and the specimen was sent for histopathological analysis.

Gross finding revealed a single bit of soft tissue measuring 4 × 6 ×2 cm, yellowish red in color, firm in consistency. Two lymph nodes were seen attached to the soft tissue bit. [Figure 3] Cut sections revealed yellowish to whitish central core surrounded by whitish brown to brownish areas [Figure 4].

Histopathological examination revealed Hematoxylin and Eosin (H and E) stained sections revealed destruction of salivary gland architecture. No acinar cell/ducts were seen. The entire mass was infiltrated by proliferation of lymphocytes which were arranged in sheets. Occasional epimyoepithelial islands were seen. Intraglandular lymph nodes showed follicular proliferation with destruction of nodal architecture. [Figures 5 and 6]. No lymphoepithelial proliferation was seen.

Based on the histopathological appearances, a provisional diagnosis of low grade lymphoma was made and immunohistochemical profiling was done to confirm the type of lymphoma.

The tissue was stained for bcl2 an anti apoptotic marker, CD20 B lymphocyte marker, CD3 which is exclusively a T lymphocyte marker, Cytokeratins and EMA.

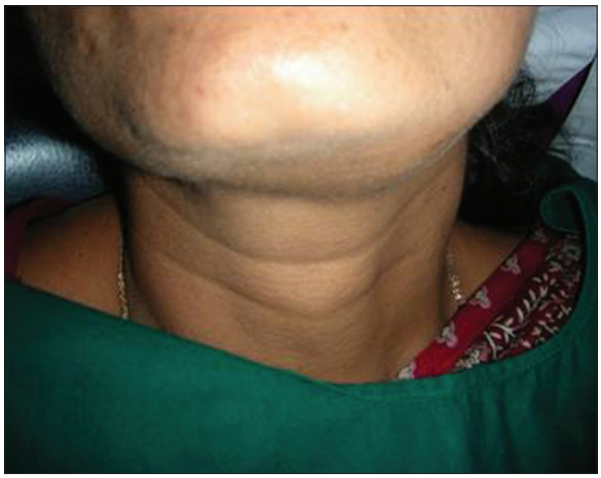

Figure 1: Clinical picture showing the swelling in the left submandibular region

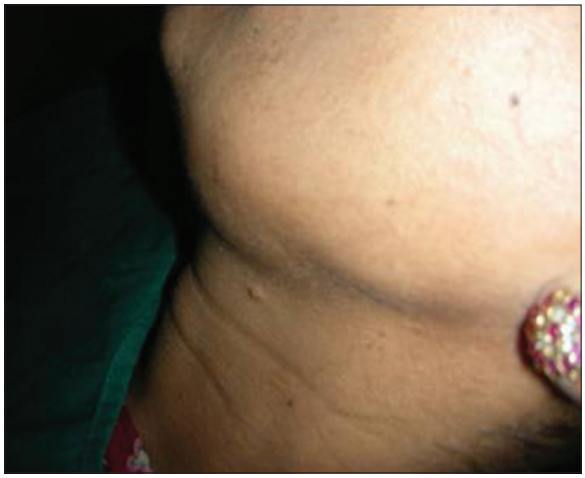

Figure 2: Swelling seen 2 cm below the ear lobe

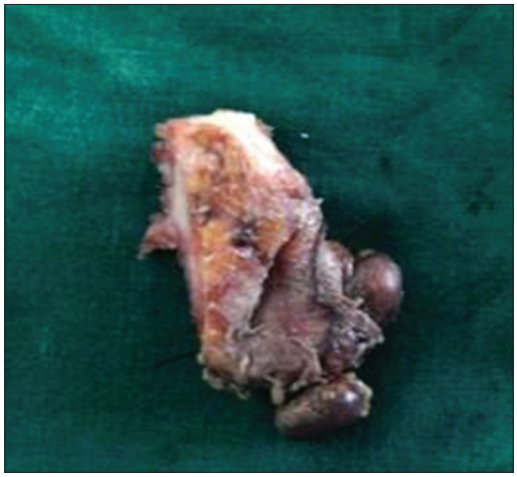

Figure 3: Specimen showing 2 lymph nodes attached to the soft tissue specimen

Immunohistochemistry revealed that the cells were strongly positive for Bcl-2 [Figure 7] and CD 20 [Figure 8] which are both cytoplasmic markers and negative for CD3 [Figure 9].

Based on the results of immunohistochemical profiling, a diagnosis of Follicular B cell lymphoma was reached. Patient was referred to higher center where full body MRI did not reveal any foci of disease. Nevertheless, considering the indolent course of this disease, she has been on regular follow up and has not reported with any clinically suspicious changes.

DISCUSSION

Lymphoma of salivary glands are uncommon and constitute 1.7–5% of all salivary gland neoplasms.[8-10] Parotid gland is by far, most commonly involved salivary gland (70%). Lymphomas also involve in the descending order of frequency, the submandibular gland (30%), minor salivary glands and the sub lingual glands.[11]

The finding of a lymphoma in a major salivary gland could mean one of two things: Either it is part of a disseminated process, or it is the first clinicopathologic evidence of lymphoma. In the latter instance, whether the disease originated in the glandular stroma itself or in a para glandular lymph node and then invaded the stroma, the lymphoma is defined as primary, as long as there is no detectable disease outside the salivary gland.[12]

Hyman and wolf have given few criterions for diagnosis of primary lymphoma[13] which are:

- Involvement of gland should be the first clinical manifestation of the disease
- Histologically, the disease should involve the gland parenchyma and not the adjacent node or soft tissue alone
- There should be confirmation of malignant nature of lymphoid infiltrate.

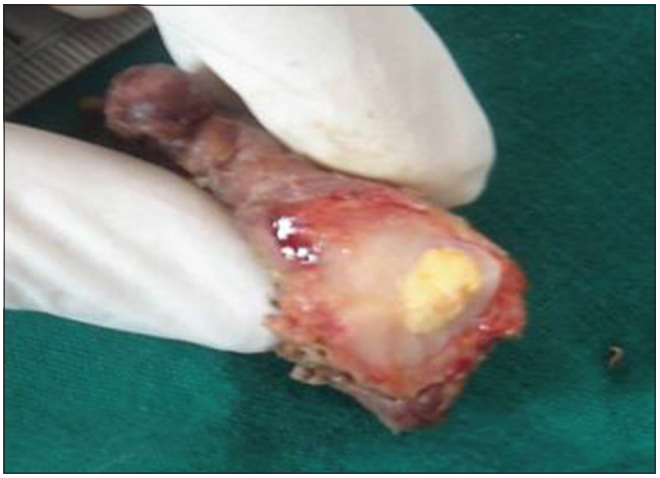

Figure 4: Grossed specimen showing yellowish to whitish central core surrounded by whitish brown to brownish areas

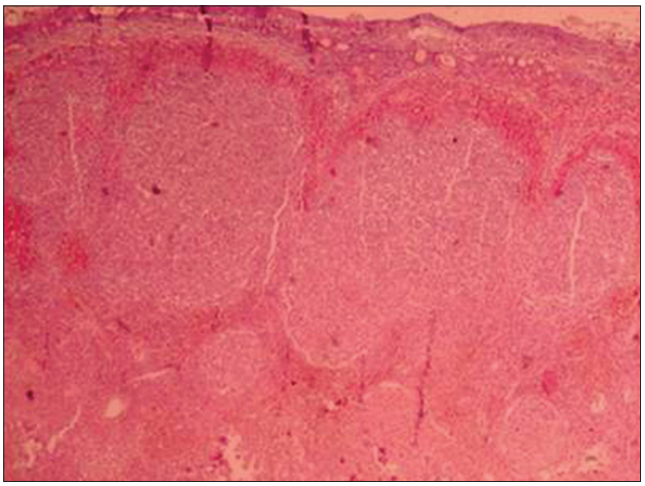

Figure 5: Photomicrograph showing neoplastic lymphocytes in a follicular pattern.(H&E stain, ×100)

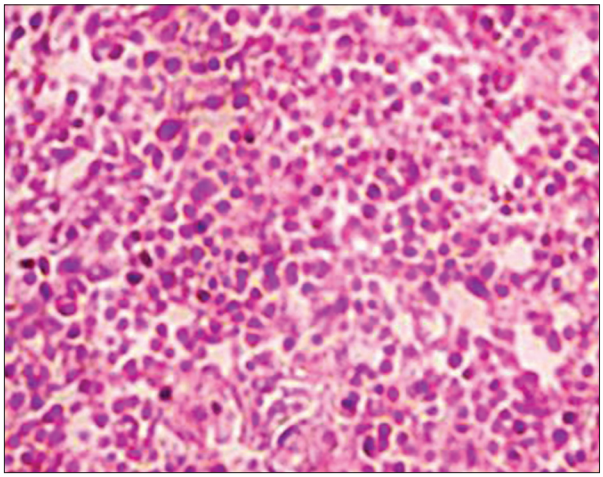

Figure 6: Photomicrograph showing neoplastic lymphocytes with hyperchromatic nuclei and scanty cytoplasm. Cellular and nuclear pleomorphism can also be seen. (H&E stain, ×200)

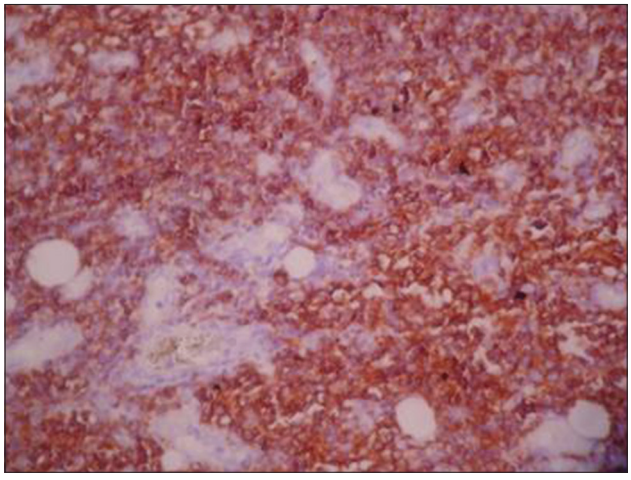

Figure 7: Photomicrograph showing neoplastic cells expressing strong positivity for Bcl-2. (IHC stain, ×200)

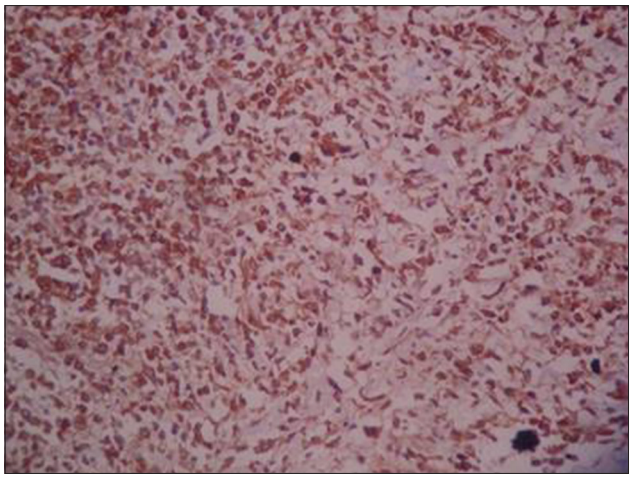

Figure 8: Photomicrograph showing neoplastic cells expressing strong positivity for CD20. (IHC stain, ×100)

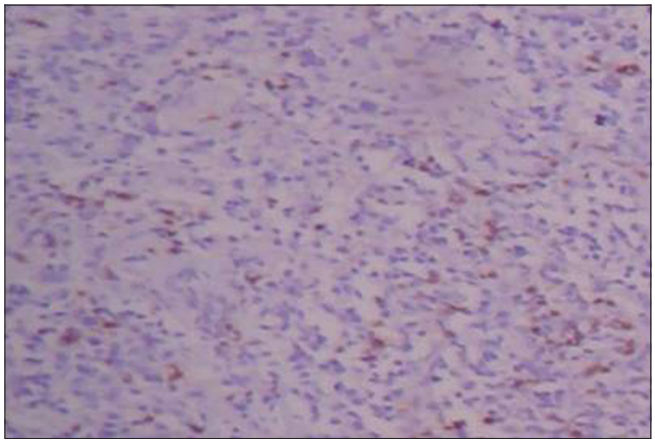

Figure 9: Photomicrograph showing neoplastic cells with negative expression for CD3. (IHC stain, ×100)

Lichenfield *et al*., reserved the diagnosis of primary salivary gland lymphoma, when lymphoma has been ruled out elsewhere in the body.

The diagnosis of lymphoma in case of a salivary gland swelling is rarely done preoperatively. Even though the diagnosis of lymphomas, in the algorithm of diagnosis of salivary gland swelling is rare, nevertheless, it should be considered.

Follicular lymphoma, although unpredictable and often associated with a prolonged course shows indolent behavior.[14] Overtime, there is a tendency to transform to a higher grade of lymphoma, usually diffuse large cell lymphoma.[2] Although various treatment modalities exist, none are curative and very few have been shown to prolong survival.[14] Follow up of the patient is necessary.

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