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OFF BEAT PRESENTATION OF EXTRA-NODAL LYMPHOMA – CASE REPORT

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ABSTRACT

Non-Hodgkin's lymphoma (NHLs) is a diverse group of lymphoid neoplasms, prevalence of which has increased since last three decades. It is diverse in the manner of presentation, response to various treatment and prognosis. NHL usually involves lymph nodes, but extra-nodal sites are not uncommon. Usually, salivary gland involvement in NHL is secondary to the widespread involvement throughout the body. Salivary gland NHL presents with clinical features of inflammation. Thus, these diagnostic mirages warrant the necessity of assessing the case in detail. Careful patient evaluation and proper investigations are of paramount importance for accurate and timely diagnosis to ensure no delay in initiating treatment. Here is such an offbeat case report, case of Extra-nodal Marginal lymphoma of Parotid.

KEYWORDS

Lymphoma, Parotid, Non-Hodgkin's lymphoma, biopsy, chemo-radiotherapy

INTRODUCTION

Lymphomas are malignant neoplasms of lymphocyte cell lines, accounting for approximately 14% of all malignant neoplasms of the head and neck. They are divided into two subtypes, Hodgkin's lymphomas (HLs) and non-Hodgkin's lymphomas (NHLs), with differences in histology, clinical features, and prognosis. HL often appears as a localized disease in the lymph nodes, especially in the mediastinum and neck. The extra-nodal onset variant of HL is rare and has an incidence rate of 5%. NHLs occur in extra-nodal sites with much greater frequency, estimated between 10 and 35% [1,2]. NHL includes a heterogeneous group of tumours: 85–90% derive from B lymphocytes and the remaining ones arise from T lymphocytes or Natural Killer (NK) lymphocytes. Both types can have nodal and extra-nodal localization. Isaacson and Wright first described extra-nodal non-Hodgkin lymphoma (ENHL) as a distinct entity in 1984 [3]. Among NHLs in the head and neck area, B-cell NHLs are the most common, while diffuse large B-cell lymphoma (DLBCL) is the most common histological subtype. Immunohistochemistry is essential for a definitive diagnosis [4,5]. The most acknowledged risk factor for the development of ENHLs is congenital or induced immunodeficiency, such as HIV infection or Severe Combined Immunodeficiency Disease [6,7]. Epstein–Barr virus (EBV) infection seemed to be strongly associated with B-cell lymphomas in a variable number of cases [7,8,9]. Other factors implicated in NHL genesis are: genetics (e.g., family history of hematologic malignancies), immune conditions (e.g., Sjogren's syndrome, rheumatoid arthritis), infections (e.g., HCV and Helicobacter pylori), modifiable risk factors (e.g., body mass index, alcohol consumption, and cigarette smoking), toxins and drugs (e.g., phenytoin, digoxin, pesticides), chromosomal translocations, and employment (e.g., agricultural or health workers) [1,2]. NHL occurs quite frequently in extra-nodal sites, and the most frequent localization is the gastrointestinal tract, followed by the head and neck area [1,2,3,6]. Oral involvement is rare, about 2% [6], and sometimes it presents as ulceration with bone destruction.

Case Report

A 66-year-old female, presented to the OPD of Otorhinolaryngology, with complaints of swelling in the right side of face, gradually progressive for the past one year, associated with pain. On examination. 5 x 5 cm swelling noted 1 cm above tragus, posteriorly 2 finger breadths from angle of mandible, anteriorly 3 cm from angle of mandible - firm swelling, with minimal tenderness. MRI done showed superficial and deep lobe, diffusely enlarged in size with altered T1 and T2 signals. Multiple intra-parotid lymph noted largest measuring 20 x 14 mm.FNAC of right parotid taken, showed features suggestive of sialadenitis or parotitis – inflammatory pathology. Patient underwent Right superficial parotidectomy under General Anesthesia. Histopathology was suggestive of Extra-nodal marginal zone lymphoma, positive for CD 45 and CD 20.

Investigation

In the case of the parotid gland, this usually entails a superficial lobectomy. The rationale of this approach is as follows:

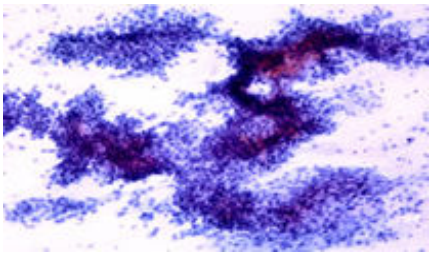
(1) A variety of epithelial salivary tumours either contain or are often associated with an exuberant lymphoid element, such as the Warthin tumour, sebaceous lymphadenoma, lymphoepithelial carcinoma, and acinic cell carcinoma. On small biopsy specimens, these tumours may be confused with malignant lymphomas or result in an uncertain diagnosis.

(2) Malignant lymphomas developing in a background of "benign lymphoepithelial lesion" may initially be focal and subtle and easily missed on a small biopsy specimen.

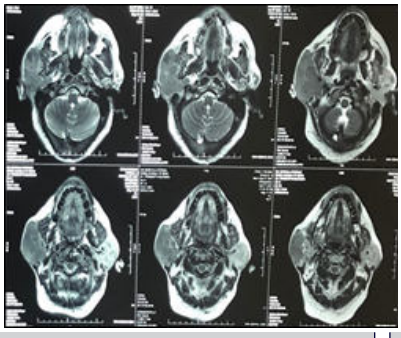
(3) Excisional biopsy also provides enough tissue for thorough routine histological evaluation and for additional specialized studies, such as T- and B-cell markers, flow cytometry, and gene rearrangement.

(4) Parotid masses, even in a patient with known malignant lymphoma, are not always lymphomatous. Some are epithelial in origin and may be even more aggressive than the lymphoma.

Patient underwent FNAC of Parotid tissue which resulted as? Chronic sialadenitis or intra-parotid lymph node.

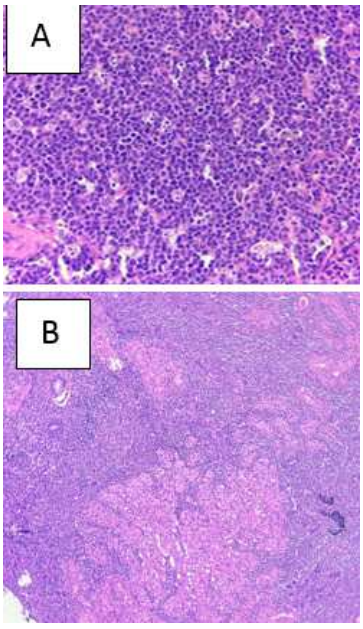


Patient was evaluated with MRI parotids :

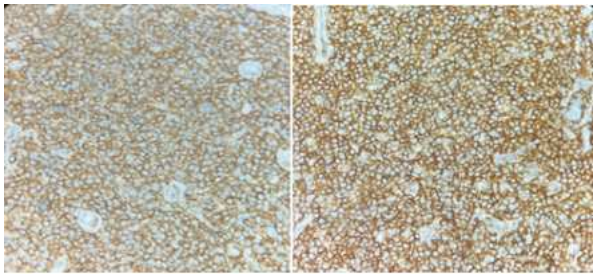


Mri Parotid: Axial cuts showing – superficial and deep lobe, diffusely enlarged in size with altered T1 and T2 signals. Multiple intra-parotid lymph noted largest measuring 20 x 14 mm.

Histopathology



A and **B**, the salivary gland parenchyma is replaced by lymphoma composed of small cells with abundant pale cytoplasm. Note the presence of reactive germinal centres as well as numerous small lymphoid cells invading the epithelial ducts.



Cd 45 Reactive **Cd 20 Reactive**

Immuno- Histochemical panel shows tumor reactivity to CD 20 and CD 45

Treatment

In this case report, patient presented with an unsuspecting benign parotid lesion that was diagnosed to be extra-nodal marginal zone lymphoma of parotid, treated with palliative chemotherapy. The patient was started on 4 cycles of Rituximab and on close follow up.

DISCUSSION

Malignant lymphomas represent approximately 5% of all malignant neoplasms of the head and neck and may involve nodal or extra nodal sites. Nodal head and neck lymphomas are similar to other nodal sites and are not further reviewed here. The head and neck region is the second most frequent anatomical site of extra nodal lymphomas (after the gastrointestinal tract). Most are non-Hodgkin's lymphomas of B-cell lineage, and overall diffuse large B-cell lymphoma is the most common type. Hodgkin's lymphoma rarely occurs in extra nodal sites. Other hematologic neoplasms that commonly involve extra nodal sites of the head and neck are also discussed. In this review, we begin by discussing lymphomas involving the head and neck according to anatomical site. Then we discuss specifically the pathological findings of extra nodal marginal zone lymphoma of mucosa-associated lymphoid tissue, plasmo - blastic lymphoma, extramedullary plasmacytoma, and extra nodal natural killer/T-cell lymphoma of nasal type.

Salivary gland lymphomas represent approximately 3% of all salivary gland tumours [17,18]. The parotid gland is most commonly affected in 80% of cases, followed by the submandibular gland (16%), sublingual gland (2%), and minor salivary glands (2%) [18]. Patients with lymphomas of the salivary glands are most commonly adults with a male to female ratio of approximately 1 to 2 [11]. Patients most often present with a painless mass and a small subset of patients have facial nerve paresis or pain [11]. Twenty percent of patients with salivary gland lymphomas have clinical or laboratory evidence of Sjogren's syndrome [18]. The most common histological types of lymphoma at this site are.

DLBCL and MALT-lymphoma. Myoepithelial sialadenitis (MESA), also commonly referred to as benign lymphoepithelial lesion, is a precursor lesion for non–Hodgkin's lymphoma [12].

Epimyoepithelial islands are nests of ductal epithelial cells that are extensively infiltrated by. small lymphoid cells. Often, abundant hyaline basal lamina material accompanies the epithelial cells, indicating that epimyoepithelial islands originate as ducts that subsequently. collapse. In the minor salivary glands, the changes are similar, although epimyoepithelial islands may be smaller or absent. Over time, the lymphoid infiltrate progressively replaces. acinar tissue, resulting in atrophy, and is associated with reactive lymphoid follicles. Myoepithelial sialadenitis and closely related lymphoepithelial cysts can also occur in HIV-positive patients [24]. Excluding the cystic component, the histological findings in lymphoepithelial cysts are similar to those in MESA. Human immunodeficiency virus has been identified within the follicular dendritic cells of these lesions [16]. Patients with MESA are at increased risk of developing non–Hodgkin's lymphoma [17]. The risk of malignant lymphoma in patients with MESA and Sjogren's syndrome has been estimated to be 43.8 times greater than that of general population [18].

Most lymphomas arising in the salivary gland are lymphomas of MALT origin and can be divided into 2 groups: low-grade and DLBCL [13,15]. Historically, DLBCLs arising in the setting of MESA were the first to be recognized because of their cytological atypia. The increased risk of MALT-lymphomas was recognized later, with the availability. of immunohistochemical and molecular methods [18]. Histologically, MALT-lymphoma forms part of a spectrum with MESA, as has been described by others [13,16,17]. The presence of atypical lymphoid cells with abundant pale cytoplasm (centrocyte-like or monocytoid B cells) forming wide zones surrounding epi-myo-epithelial islands is useful. histological finding to suggest that MALT-lymphoma is evolving from MESA. Lymphoepithelial lesions are not helpful in distinguishing MESA from MALT-lymphoma as they occur in both conditions. As the neoplasm progresses, epimyoepithelial islands are destroyed, reactive follicles are infiltrated and replaced, and the process extends outside the salivary gland and may exhibit perineural invasion [15,16]

Diffuse large B-cell lymphoma arises at a later stage in this sequence of events, with accrual of sheets of large cells and accompanying mitotic figures.

Not all lymphomas that involve the salivary glands are truly of extra nodal origin. The parotid gland is closely associated with many lymph nodes surrounding the gland. and within the parenchyma. These lymph nodes may be replaced by nodal lymphomas that secondarily replace the parotid gland. At the time of histological analysis, it may not be possible to determine the site of origin of the neoplasm without immunophenotypic or molecular studies. Some investigators consider these nodal lymphomas to be primary. lymphomas of the parotid gland, further confusing the issue [18].

Lymphomas also are rarely associated with Warthin's tumour, with approximately 20 cases reported in the literature. Most of these neoplasms have been follicular lymphoma, DLBCL, or small lymphocytic lymphoma, neoplasms that typically arise in nodal sites. Extra nodal marginal zone B-cell lymphoma of MALT is rare. Others have suggested. that Warthin's tumour arises from salivary gland inclusions within lymph node, explaining the types of lymphoma that most often occur with Warthin's tumours [18]

Sarris et al suggested radiotherapy for early-stage lesion and chemotherapy for late onset disease [17]. Marioni et al emphasized that incomplete surgical removal or disseminated cases should be treated with radiotherapy and chemotherapy [18].

In our case patient was started on 4 cycles of Rituximab and on close follow up.

CONCLUSION

- It's imperative to keep in mind, the unusual sites and patterns of presentations of hematological malignancies.
- Histopathological examination of strong and suspicious lesions is essential for early diagnosis and treatment.
- Ensuring early initiation of treatment is vital for better.
- A complete assessment and staging is mandatory before starting any therapy. The patients who present at an early stage or with low-grade NHL

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Conflicts Of Interest
There are no conflicts of interest.

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