Case Report

An aggressivte umor in parotid gland

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Abstract

Non Hodgkin Lymphoma (NHL) of salivary gland (SG) is rare with an incidence of 2-5%. We describe a stage III, high grade NHL of parotid gland in a background of benign lymphoepithelial lesions. A 54 year female presented with a right side facial mass since 6 months. Fine needle aspiration cytology (FNAC) suggested a lymphoproliferative disorder and superficial parotidectomy was done. Histopathology and immunohistochemistry (IHC) revealed an extranodal marginal zone NHL with transformation to high grade diffuse large B cell lymphoma. SG lymphoma should be considered in elderly and IHC helps in prognostication and treatment protocols.

Key words: Parotid gland, Lymphoepithelial lesions, Non Hodgkin Lymphoma, Immunohistochemistry

Introduction

Non Hodgkin Lymphoma (NHL) of salivary gland is extremely rare constituting about 2–5% of all salivary gland neoplasms and the parotid gland (50-93%) being the most commonly affected major salivary gland [1, 2].

NHL can arise from an intraparotidlymphnode or the gland per se, however their distinction on the basis of morphology and prognosis is often difficult [1,3]. Most of them are of B cell lineage the more common being low-grade B-cell lymphomas of mucosa-associated lymphoid tissue (MALT), diffuse large B-cell lymphomas and follicular lymphomas [1]. Benign lymphoepithelial lesion (BLL) proves a suitable precursor lesion for an Extranodal marginal zone B-cell lymphoma (EMZBCL) which can further transform into high grade diffuse large B cell lymphoma (DLBCL) [4].

EMZBCL of salivary gland has to be differentiated from the non-MALT lymphoma and the epithelial tumors as their management and prognosis is completely different [1]. Immunohistochemical analysis of NHL helps in the diagnostic classification, to guide a therapy, and to predict the clinical outcome [5].

Case Report

A 64 years female presented with swelling and pain in right parotid region since 6 months. A firm mass with restricted mobility of 6x4cm size with intact right facial nerve was noted. Magnetic Resonance Imaging (MRI) neck also showed three nodular high intense solid lesions with ill defined margins suggestive of malignant behaviour (Fig-1A).

Fine needle aspiration cytology (FNAC) revealed a cellular smear which were pleomorphic with increased nuclear: cytoplasmic ratio and scanty cytoplasm (Fig-1B). A possibility of a malignant lymphoproliferative disorder was considered. Conservative superficial paroditectomy with facial nerve preservation was done, which measured 7x4.5x2cm. Cut section was fleshy with three ill defined nodular grey brown lesions varying in sizes from 2 to 2.5 cm (Fig-1C).

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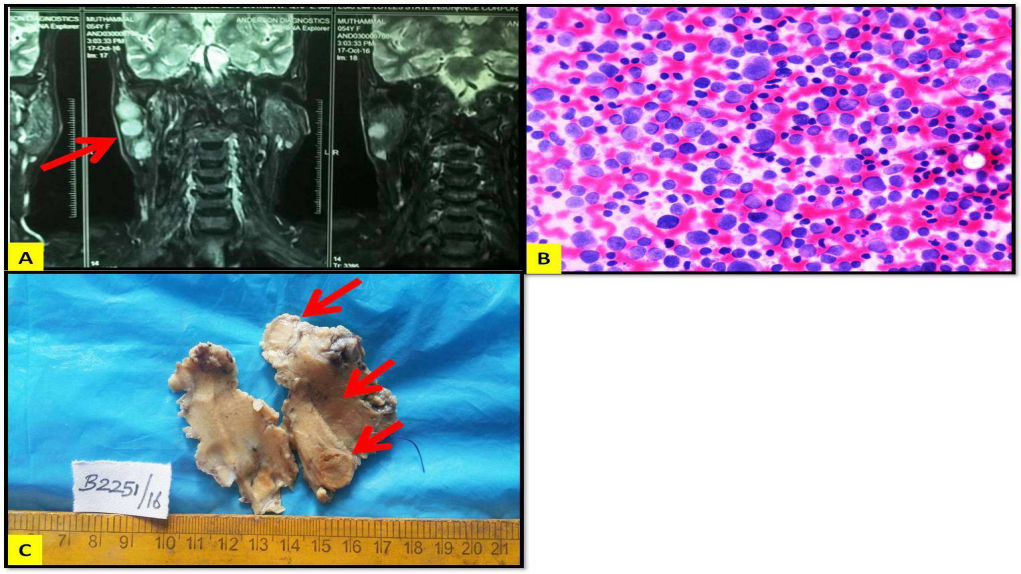

Figure 1:A) MRI neck showed 3 nodular high intense solid lesions with ill defined margins suggestive of malignant lesions (Arrow). B) FNAC revealed a cellular smear with pleomorphic cells, increased N:C ratio and scanty cytoplasm. (H&E, X400). C) Cut section was fleshy with 3 ill defined nodular grey brown lesions varying in sizes from 2 to 2.5 cm. (Arrows)

Histopathological examination exposed a neoplasm arranged in sheets consisting of large cells, scanty cytoplasm with round to oval pleomorphic nuclei with vesicular chromatin and prominent nucleoli (Fig-2A,2B). Brisk mitoses and necrosis were noted along with areas of benign lymphoepithelial lesions (Fig-2B,2C,2D).

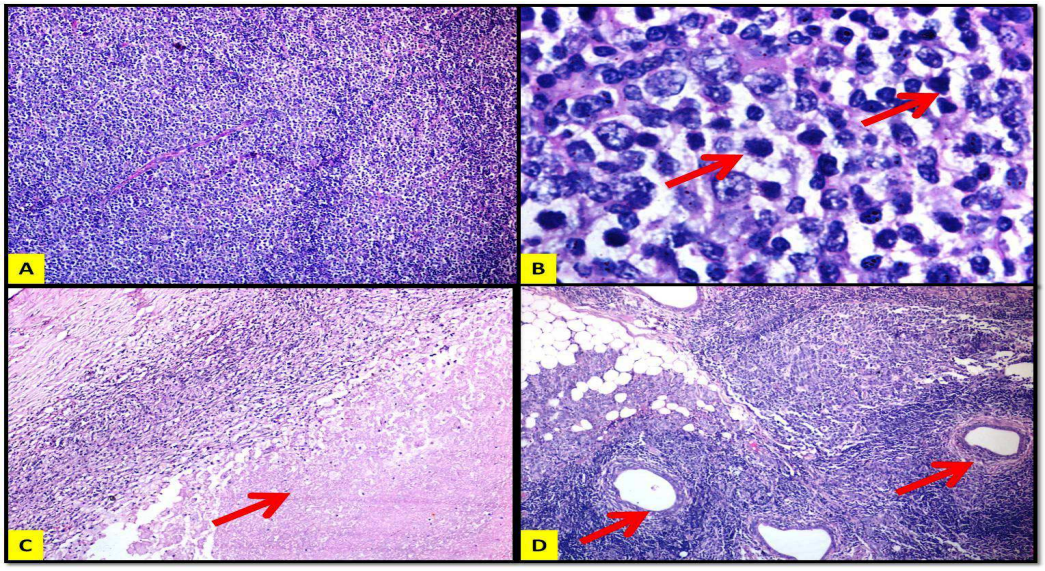

Figure-2: A) Diffuse sheets of lymphoid cells were seen (H&E, X 40). B) Cells are large cells, scanty cytoplasm with round to oval pleomorphic nuclei with vesicular chromatin and prominent nucleoli with brisk mitoses (Arrows) (H&E, X 40). C) Necrosis was noted. (Arrow) (H&E, X 40) D) Areas of benign lymphoepithelial lesions were seen (Arrows) (H&E, X 40)

A diagnosis of extranodal marginal zone lymphoma of parotid gland with transformation into high grade NHL was made. Immunohistochemistry showed positivity of CD20 and BCL2 with high Ki67 index of 40 to 50% (Fig-3A, 3B, 3C). CD10, BCL6, MUM1 (Fig-3D, 3E, 3F), CD5 and Cyclin D1 were negative confirming the transformation.

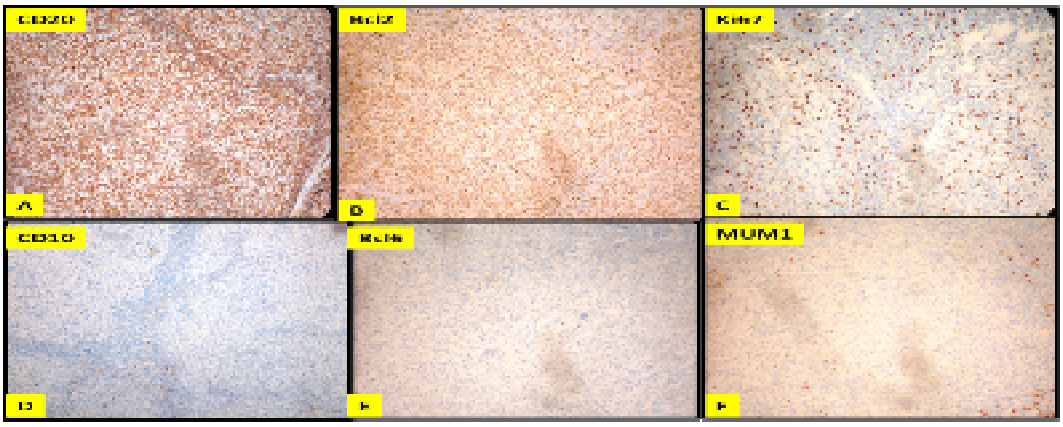

Figure- 3: Microphotograph showing tumor cells A) positive for CD20 (IHC, x200), B) positive for Bcl2 (IHC, x200), C) Ki 67- high index 40% - 50% (IHC, x200), D) negative for CD10 (IHC, x200), E) negative for Bcl6 (IHC, x200), F) negative for MUM1 (IHC, x200)

A whole body PET-CT scan showed metabolic activity in portocaval, peripancreatic and submandibular lymph nodes confirming a clinical stage III disease (Fig-4A, 4B).

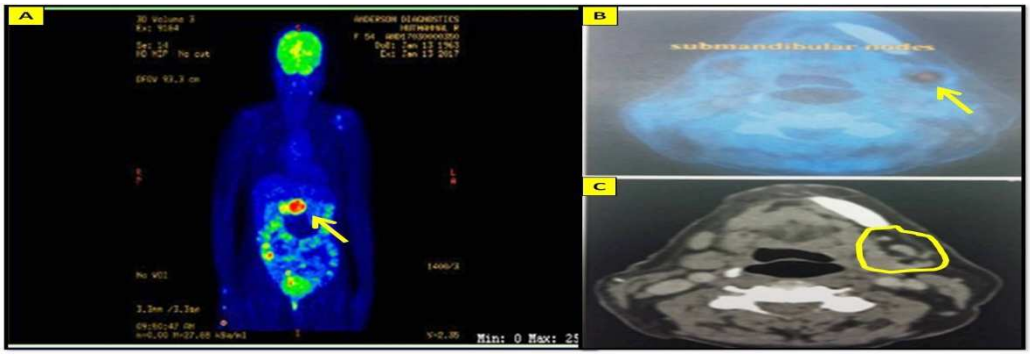

Figure- 4: A) Whole body PET-CT scan showed metabolic activity in portocaval, peripancreatic. (Arrow) B) & C) Metabolic activity in submandibular lymph nodes confirming a clinical stage III disease (Arrow and marking)

Post operative recovery was uneventful with intact facial nerve function. Patient was started on R-CHOP regimen.

Discussion

EMZBL of MALT- type is the predominant type of lymphoma in salivary gland [6]. In the mucosal sites they are referred as mucosa- associated lymphoid tumors (or "maltomas"), first described by Isaacson and Wright in 1983 [7]. They can present as a slow growing, less painful mass creating diagnostic dilemmas [4].

Majority develop in parotid glands (76%), and the rest in submandibular (20%), sublingual (3%) and palatal glands (1%) [8]. Kalpadakis et al has reported increased incidence in females in a series of 76 patients with non-gastric EMZBL for unknown reasons [9]. NHLs such as marginal zone lymphomas, follicular and DLBCL are common and marginal zone lymphomas are heterogeneous B-cell tumors arising in lymph nodes, spleen, or extranodal tissues [1]. Chronic lympho-proliferations such as BLL can act as fertile ground for lymphoma development of which EMZBL of MALT type is the commonest to occur [1,4]. BLL are clusters of B cells inter-digitating with ductal epithelial cells and may contain clonal B cells [4]. Many such lesions were identified in our case.

Though FNAC has its reservations in diagnostic utility of salivary gland lymphomas, along with clinical and radiological evaluation, it helps in preoperative assessment [4].

FNAC in our case showed a high grade malignant lymphoproliferative lesion. Surgery helps in treatment, reaching a definitive histological diagnosis and planning of follow up [4]. De novo transformation into DLBCL can occur in low grade lymphomas such as follicular lymphoma, CLL/ SLL or marginal zone lymphoma making up to 30%–40% of NHL in adults [10].

DLBCL has medium to large cells arranged in a diffuse pattern often distorting normal architecture and the neoplastic cells are positive for CD20, CD19, CD79a, CD22 and PAX5 markers and proliferation index is usually more than 40% [5].

Hans et al. divided DLBCL cases into germinal centre B cell like (GCB) subtype with good prognosis and non GCB cell like subtype with poor prognosis, based on the expressions of CD10, Bcl- 6, and MUM 1[10].

Positive expression of CD10, Bcl-6 and negative expression of MUM1 suggest GCB cell like subtype, whereas negative expression of CD10, Bcl-6 with MUM1 positivity suggest non GCB cell like subtype [10].

CD10, Bcl6 and MUM1 are independent markers for prognostication [10].

BCL-2 positivity is seen in lymphomas but not in hyperplasia of monocytoid B cells and is associated with poor prognosis [10].

Our case was positive for CD20 and BCL2 with negative staining for Bcl6, CD10, MUM1, CD5, Cyclin D1, and EBV.

Ki 67 proliferation index was more than 40%, all suggesting a non GCB subtype carrying a poor prognosis. IHC markers therefore immensely help in predicting prognosis and directing treatment options thereby help in improving the survival of patients with DLBCL [5].

Extra gastric MALT lymphoma in head and neck especially salivary gland region behaves more aggressively and can recur in other organ sites when compared to the gastric type and therefore regular follow up planning is essential [4].

Myeloid cell Nuclear Differentiation Antigen (MNDA) is an emerging marker for Marginal Zone Lymphoma and can differentiate from follicular lymphoma [5]. Complete evaluation for disseminated disease is essential before the commencement of the treatment [4].

PET-CT scan can be used for staging and follow up [4]. Our patient had clinical stage III at the time of presentation with metabolically active large portocaval and peripancreatic lymphnodes.

Radiotherapy or surgery has been advised for localised disease while chemotherapy along with surgery and or radiotherapy is needed for disseminated diseases [4].

CD20 positivity of lymphoma helps in the usage of rituximab (monoclonal anti CD20 antibody) in chemotherapy therapy regimens [5].

Conclusions

Parotid gland lymphomas can mimic benign lesions and should be considered in the differential diagnosis especially in elderly patients.

A high index of suspicion is required for an early diagnosis.

IHC expression of CD20, BCL2, CD10, BCL-6 and MUM1 helps in prognostication and treatment protocols.

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