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Research Article

UNILATERAL SWELLING IN THE RIGHT PAROTID REGION: MANIFESTATION OF B CELL LYMPHOMA

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ABSTRACT

Approximately 2% of all salivary gland tumors are primary Non-Hodgkin lymphoma (NHL) and 75% of these lymphomas are seen in the parotid gland. Extra-nodal marginal zone B-cell lymphoma of the mucosa-associated lymphoid tissue (MALT), follicular B-cell lymphoma, and diffuse large B-cell lymphoma are the commonest subtypes of the NHL found in the salivary glands. The prognosis of different NHLs of the salivary gland depends on the histological subtypes and clinical stages. We present a case of diffuse large B-cell lymphoma in 65-year-old female patient and presented as a nonspecific unilateral right preauricular swelling.

INTRODUCTION

Malignant lymphoma is a group of diseases which have a wide variety of clinical, histological features, genetic abnormalities and immunophenotypes. Malignant lymphomas can be categorized into two major subtypes, Hodgkin's lymphoma (HL) and Non-Hodgkin's lymphoma (NHL). Lymphomas derived from T-cells, B-cells and NK cells belong to a group of NHL. HL usually appears as a node-type disease including inguinal, axillary and cervical nodes. Whereas, NHL localizes extra nodally in the digestive tract, salivary glands and rarely the jaw. The latter group has the most prevalence of all lymphomas in the head and neck, accounting for 75% of cases. Diffuse large B-cell lymphoma (DLBCL) is the most common NHL type in the head and neck area.[1]

Approximately a quarter of all lymphomas on the extra nodes develop in the head and neck, principally in the parotid glands, tonsils and pharynx. Among tumors of the parotid, the prevalence of lymphoma is rare, representing 1% to 4% of cases.[2]

Regarding therapy, localized low-grade lymphomas can be treated with radiotherapy only, whereas diffuse high-grade types are treated with aggressive chemotherapy. A combination of radiotherapy and chemotherapy is used to treat patients with localized high-grade lymphomas.[3]

Case Report

A 65-year-old female patient came with the complain of swelling on right side of face, in front of ear since 15 days. Patient had pain 15 days ago in lower left back tooth region for which she had applied moist heat application done and then swelling started which was initially smaller in size and gradually increased to present size with no signs and symptoms. No relevant medical and family history.

On extraoral examination there was solitary swelling present on the right pre auricular region, roughly oval in shape, measuring 3X4cm in greatest diameter with well defined borders, skin over the swelling was stretched and shiny, extending antero-posterollarly 3cm in front of ear to 1cm behind the ear and supero-inferiorly from tragus to 1cm below the lower border of the mandibula. On palpation there was local rise in temperature, non-tender, all inspectory findings were confirmed. There was normal mouth opening and salivary flow rate.[Figure 1]

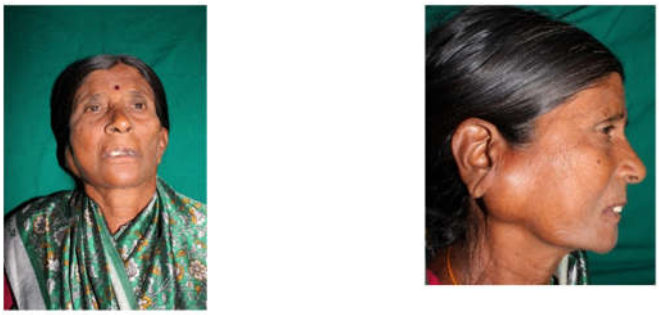

Figure 1 Profile of the patient showing swelling on right side of face at pre auricular region.

On intra oral examination normal complement of teeth present. Grade 3 mobility noted in relation to lower left 2nd and 3rd molar and grade 2 mobility in relation to lower right 2nd and 3rd

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molar and lower left 1st and 2nd molar. The periodontal status of the patient was poor.[Image 2]

Figure 2 There is lobulated focal hypoechoic solid soft tissue lesion in dumbbell shape configuration seen extending form superficial lobe of the right parotid gland towards deep lobe of the right parotid gland measuring 4.4x3 cms.

USG report revealed lobulated focal hypoechoic solid soft tissue lesion in dumbbell shaped configuration seen extending from superficial lobe of right parotid gland towards deep lobe of right parotid gland measuring 4.4X3cm with the impression of Lobulated solid hypoechoic dumbbell configured lesion noted in the superficial lobe of the right parotid gland extending to the deep lobe of the right parotid gland. There is minimal internal vascularity noted. Intraparotid schwannoma to be considered. [Image 3]

Figure 3 Immunohistochemistry study. A – represent the H and E stain, B – represent the KI-67 and C-represent the CD20

Biopsy was done and the tissue was sent for immunohistochemistry where it revealed the diffuse sheets of atypical lymphoid cells of intermediate – to large size having vascular chromatin and scant cytoplasm with prominent nuclei. Tumor cells are positive for CD20, CD30, CD45, CD10, BCL2, BCL6, OCT-2, PAXS and negative for CD3, MUM1, CD21, ALK and the final diagnosis of high gradeB cell lymphoma, germinal center subtype was given.[Image 4]

DISCUSSION

Primary lymphomas of the salivary glands are rare and account for 2–5% of all salivary gland neoplasms. Parotid is most commonly involved in 50-90% cases followed by submandibular gland. Primary Parotid lymphomas account for 0.87% of all NHL cases and 4–5% of all extranodal NHLs [4] Non-Hodgkin's Lymphomas (NHL) are a heterogeneous group of lymphoproliferative malignancies that are much less predictable than Hodgkin's and have a far greater predilection to disseminate to extranodal tissues. In the head and neck region, NHL has been observed in the Waldeyer's ring, oral mucosa, salivary glands, paranasal sinuses, laryngeal tissue, and osseous structures. [5]

These neoplasms may arise from an intraparotid lymph node or in the gland itself. Most primary salivary gland lymphomas are of B cell lineage in which the MALT (Mucosa-associated lymphoid tissue) lymphomas are most common. These arise in a background of benign lymphoepithelial lesions and have known to have an association with Sjögren's disease. Other NHLs like Diffuse Large B Cell (DLBCL) and follicular lymphomas are less commonly reported. The DLBCL is a high-grade infiltrative tumor associated with destruction of salivary gland parenchyma with tumor cells invading between residual gland acini. [6]

Parotid lymphoma most commonly presents as painless mass indistinguishable from other nonmalignant and other more common epithelial tumors. Our case also presented with painless, progressive, unilateral parotid enlargement.[7]

CT and MRI findings of the parotid MALT lymphoma described in previous case reports included a significant swelling of the parotid parenchyma on unilateral or bilateral sides, solid nodules, multiple microcystic changes and multiple calculi in both the intracystic portion and the parenchymal gland. Corr *et al* reported that most solid nodules in MALT lymphoma correspond to hyperplastic lymphoid tissue or lymphoma, and cystic portions correspond to lymphoepithelial cysts arising from compression of terminal parotid ducts by contiguous hyperplastic or neoplastic lymphoid tissue. [8]
On gross examination lymphomas usually are well-circumscribed lesions with greyish white to pink-tan cut surface and soft to firm, rubbery consistency.

On histology the cells are large and atypical and resemble either centroblast or immunoblast.IHC showed diffuse positivity for CD20, focal positivity for Bcl-6 and negative for CD3, CD5, CD10, and MUM1 helped in categorizing it as NHL of diffuse large B cell type. [7]

Most of the lymphomas of the salivary gland are surgically treated because they lack definitive imaging features.Diffuse large B-cell lymphoma is normally treated by CHOP (using cyclophosphamide, doxorubicin hydrochloride, oncovin, and prednisolone) and occasionally by R-CHOP. The RCHOP (rituximab, cyclophosphamide, doxorubicin HCl, vincristine and prednisone) regimen has been approved as gold standard chemotherapy to treat aggressive BNHL. Generally, the prognosis for primary salivary gland lymphoma is reported to be more favorable than that for nodal lymphomas.[8,9]

CONCLUSION

DLBCL of the major salivary glands is a rare yet aggressive malignancy. Head and neck surgery remains a critical component of diagnosis as clinical features may often resemble benign conditions. Despite the rising incidence in recent decades, treatments with chemotherapy and radiation have resulted in increasingly favorable survival outcomes. Female patients and those presenting with late-stage disease may be at higher risk of mortality for this aggressive cancer.

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