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Case Report

Parotid Gland Lymphoma: A Rare Entity

Vipul Bansal[1], Anil K Dhull[2], Rajeev Atri[3], Vivek Kaushal[4], Rakesh Dhankhar[3]

[1]Junior Resident, [2]Senior Medical Officer, [3]Professor, [4]Senior Professor & Head
Department of Radiation Oncology, Pt BD Sharma UHS, Rohtak

Corresponding Author: Vipul Bansal

ABSTRACT

Lymphomas arising from the parotid gland are rare entity and often misdiagnosed and overlooked clinically as well as pathologically. This diagnosis should always be kept in mind when a patient presents with a non-tender mass in this gland, since it may save the patient from undergoing unnecessary diagnostic procedures, thereby prompting the appropriate medical treatment.

Keywords: Parotid, lymphoma, rare

INTRODUCTION

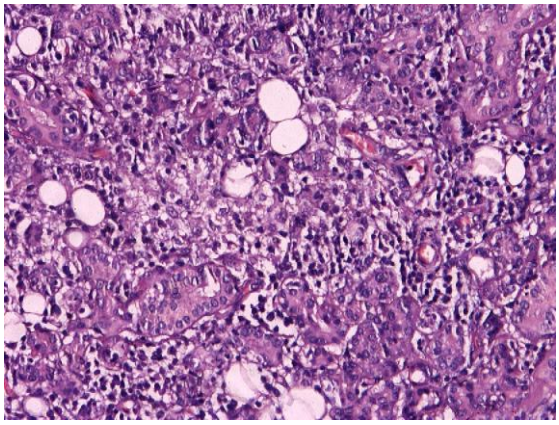
Lymphomas arising from the parotid gland is an uncommon entity and accounts for only 0.2-0.8% of the malignant tumors of the parotid, [1] and is therefore commonly overlooked. The misdiagnosis often leads to unnecessary diagnostic procedures, delaying the initiation of proper treatment.

CASE REPORT

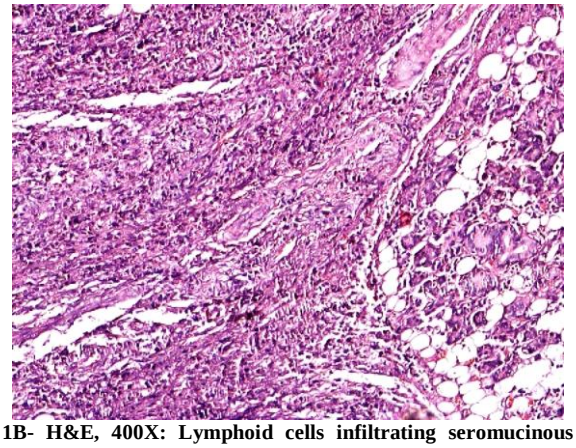
A 64 years old male came with the chief complaints of deviation of mouth to right side with dribbling of saliva for the past 10 days. On examination, patient had a hard swelling with restricted mobility of size approximately 5x4 cm in the parotid region along with a preauricular scar mark. No regional lymphadenopathy was present. According to patient he was apparently asymptomatic 1 year back when he developed a swelling anterior to left ear in the parotid region which was painless and progressively increasing in size for which he underwent parotidectomy at some private clinic. Previous CT scan revealed diffuse enlargement of the left parotid gland with mild heterogeneous enhancement with few enlarged upper deep cervical lymph nodes largest of size 2.2x1.0 cms.

Patient was reinvestigated in the Department of Radiotherapy, PGIMS, Rohtak to find out the cause of facial nerve palsy. MRI of the parotid gland revealed large parotid lesion with indistinct margins infiltrating left sternocleidomastoid muscle with encasement of posterior belly of digastrics, left facial nerve, retromandibular vein, left external carotid artery and compression of external jugular vein. Enlargement of few ipsilateral upper deep cervical lymph node was also evident. The lesion was labelled as malignant and histopathological examination was advised. Aspiration smears were suggestive of a lymph proliferative disorder and comprised of predominant population of immature lymphoid cells with scant to moderate cytoplasm, large nucleus with fine chromatin and conspicuous nucleoli. Histopathology and immunohistochemical (IHC) findings were concordant with cytology report. Biopsy report revealed lymphoid infiltrate of monomorphic variety variably infiltrating the lobules of seromucinous glands. [Fig. 1A & 1B] On, IHC the lymphoid cells were diffusely positive for LCA and CD20. [Fig. 1C & 1D]

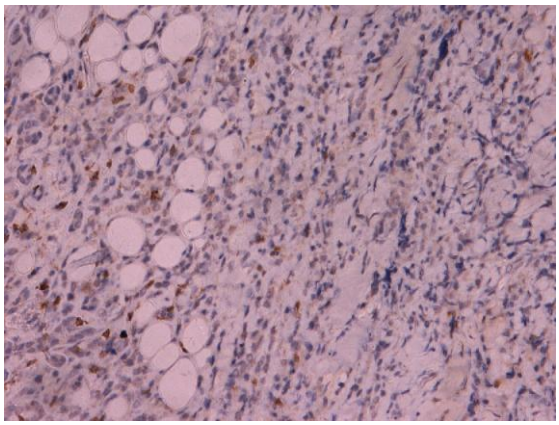
Based on clinical findings, radiological examination and immunohistochemical typing of the tumor, diagnosis of lymphomatous lesion was rendered and treatment was started for Non-Hodgkin's lymphoma. Patient has received 3 cycles of CHOP regimen including cyclophosphamide, doxorubicin, vincristine and prednisolone. The patient responded very well and there is significant clinical improvement.



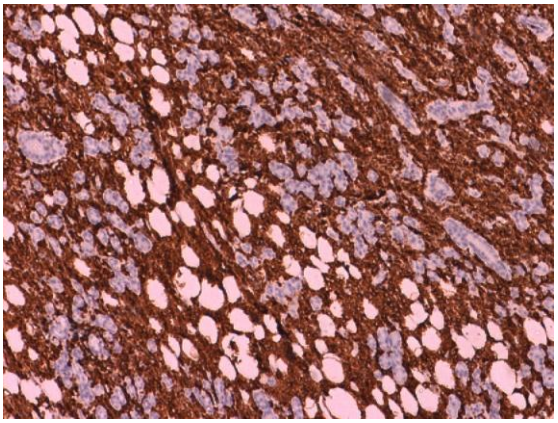
1A- H&E, 40X: Lymphoid cells infiltrating seromucinous glands



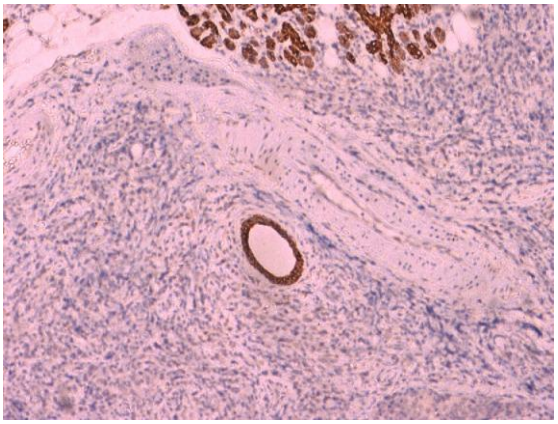
1B- H&E, 400X: Lymphoid cells infiltrating seromucinous glands



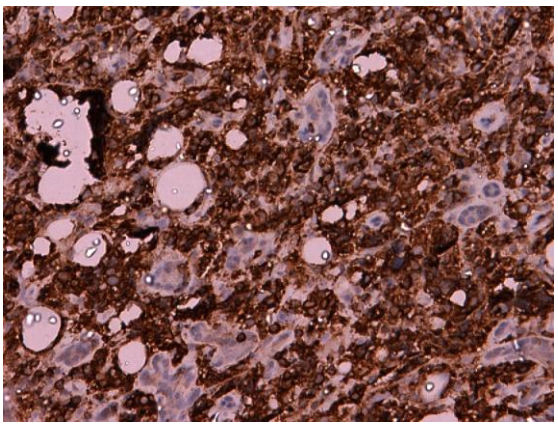
1C- 200X: Lymphoid cells revealing diffuse LCA positivity



1D- 200X: Lymphoid cells revealing diffuse CD 20 positivity



1E- 200X: Lymphoid cells revealing diffuse CD 3 negativity



1F-200X: Lymphoid cells revealing cytokeratin negativity

DISCUSSION

Parotid lymphomas constitute 0.2-0.8% of malignant tumors in the parotid gland although the prevalence of this rare malignancy has risen in recent decades. [2] Most commonly it presents as a painless mass indistinguishable from non-malignant or other more common epithelial tumors. Its prevalence increases with age (peak incidence at 55 years of age) and the male/female ratio is 2:1. Generally surgeons

do not anticipate primary lymphomas in the salivary glands pre-operatively and pathologists too find it difficult to give a definitive and diagnostic report based on either frozen section or fine needle aspiration cytology. This difficulty in pre and intra-operative diagnosis often unnecessarily exposes the patients to radical operations with all its associated risks involved. The pathophysiology of NHL in parotid gland is unknown. Autoimmune disorders like Sjogren's syndrome has been found to be increasingly associated with lymphomatous lesions of the salivary glands as these may predispose to uncontrolled proliferation of lymphoid cells in the glandular tissue. [3] Due to rarity of this entity it is often overlooked, however clinically the presence of regional lymphadenopathy and association of autoimmune disease should make the physician consider lymphoma as possible etiology of the lump. Usually these patients present with unilateral enlargement of the gland and without any associated B-symptoms like fever, weight loss, night sweats etc. Hyman and Wolff have proposed following three criteria for the diagnosis of primary parotid lymphoma: a) involvement of the salivary gland as the first clinical manifestation of disease; b) histologic proof that lymphoma involves the salivary gland parenchyma, rather than being confined to soft tissue or a lymph node in the area; c) architectural and cytologic confirmation of the malignant nature of the infiltrate. [4] Our patient fulfilled all the above said criteria and thus was managed accordingly. Core biopsy has been considered as the procedure of choice for the diagnosis of parotid lymphoma. The diagnostic pathway of an isolated parotid mass must also include imaging and a FNAB that may lead to right diagnostic lines in some case as was in our case. The treatment of primary parotid lymphoma includes chemotherapy and radiotherapy as suggested in several reported series.

Survival rate for parotid lymphoma vary considerably in the literature and is generally better than other extranodal lymphomas in which the survival rate is not over 41%. [5] The majority of authors reported a long-time survival rate ranging from 50% to 75%. [6]

CONCLUSION

Although primary parotid lymphoma is a rare entity, this diagnosis should always be kept in mind when a patient presents with a non-tender mass in this gland, since it may save the patient from undergoing unnecessary diagnostic procedures, thereby prompting the appropriate medical treatment.

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