Primary aggressive non-Hodgkin lymphoma of the parotid gland in a young individual: A case report

Devika Gupta, GPS Gahlot, Vandana Rana, Rajat Jagani, Davendra Swarup

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Introduction: Extranodal non-Hodgkin lymphomas constitute 25–40% of all lymphomas. The most common site is gastrointestinal tract followed by head and neck area. Salivary gland is involved in 2–5% of all cases.

Case Report: We herein report a case of a 24-year-old male who presented with painless, gradually increasing mass in the region of left parotid. Imaging studies, both ultrasonography and magnetic resonance imaging (MRI), were suggestive of infiltrative mass lesion involving the left parotid gland along with evidence of suspected bony metastasis. Patient underwent total parotidectomy with excision of level II&III cervical lymph nodes. Histopathological examination supported by immunohistochemistry helped us to clinch the diagnosis of primary diffuse large B cell lymphoma of the parotid gland.

Conclusion: Non-Hodgkin lymphoma can mimic a large number of both benign and malignant disease entities of the salivary gland. Hence a high index of suspicion is required for an early diagnosis of non-Hodgkin lymphoma in a rapidly growing mass of salivary gland.
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Keywords: Non-Hodgkin lymphoma, Salivary glands, Parotid gland, Diffuse large B cell (DLBCL)

INTRODUCTION

Non-Hodgkin lymphoma (NHL) is a heterogenous disease of lymphoid/extra lymphoid tissues presenting clinically in indolent as well as aggressive forms. Lymphoma constitute 2–5% of all salivary gland neoplasms with the parotid gland being involved most frequently followed by submandibular and then minor salivary glands [1]. 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. Our case is primary DLBCL arising in the parotid which was diagnosed on histology following total parotidectomy.
CASE REPORT

A 24-year-old male, non-smoker with no known co-morbidities presented with rapidly progressive, painless swelling of left parotid region which gradually increased in size from about 2x1 cm to 5x4 cm over a short span of four months. There was no history suggestive of fever or any connective tissue disorder. There was no other significant personal/family history. General physical examination revealed a well-nourished individual with no lymphadenopathy. On local examination there was a firm, nontender diffuse swelling measuring approximately 5x4 cm in the left parotid region associated with puckering and discoloration of overlying skin. There were no overlying dilated veins, visible pulsation, sinus fistula seen. Oral examination and laryngoscopy was normal. Systemic examination of the respiratory, cardiac and central nervous systems were within normal limits. All his routine hematological and biochemical parameters were within normal limits except for serum lactate dehydrogenase (LDH) which was raised to 522 IU/L. Fine-needle aspiration cytology (FNAC) attempted twice from the lesion was inconclusive. Ultrasonography of neck revealed a lesion heterogenous in echotexture involving the left parotid gland with multiple enlarged level II, III cervical lymph nodes, largest measuring 1.3x1.7x2 cm at level II (left). The lymph node in level II appeared necrotic. Magnetic resonance imaging (MRI) scan showed irregularly contoured lesion in left parotid gland involving both the superficial lobe and tail region with extension in to the deep lobe and measuring approximately 32.6x20.4x22 mm (CCxAPxTR). There was focus of T1 and T2 hyperintensity within this lesion in the tail suggestive of hemorrhage. No cystic components were noted within this lesion. Multiple left necrotic lymph nodes were noted in the level I, II, and level III.

Patient was taken up for left extended parotidectomy under general anesthesia. Peroperative findings showed a growth involving anterior one-third upper border of sternocleidomastoid, level II and part of level III lymph nodes. A firm mass measuring 3.5x2x4x2 cm was also present in deep lobe below cervical and zygomatic temporal divisions of facial trunk in a dumb bell fashion. External carotid artery, internal jugular vein and facial nerve were preserved. Excision of the superficial lobe, deep lobe, part of sternocleidomastoid muscle, level IIb and level III lymph nodes with skin over parotid was done. The procedure was uneventful without any evidence of facial nerve damage. Histopathology of the mass revealed atypical lymphoid cells involving both the superficial and deep parotid lobes. These cells were seen infiltrating through the parenchyma of both lobes and infiltrating the sternocleidomastoid muscle. The cells were large, oval to round had vesicular, hyperchromatic nuclei with conspicuous nucleoli. Mitosis was brisk. Areas of necrosis with numerous scattered apoptotic bodies were seen. No lymphoepithelial lesions were identified. The left juxtraglandular lymph nodes, level III and level IIb lymph nodes showed involvement by similar malignant cells. Immunohistochemistry (IHC) study was performed and the tumor cells were strongly positive for LCA, CD20, CD79a and negative for CD3, CD5, CK, EMA, Tdt. The Ki-67 labeling index was 100%.

Based on histomorphology and IHC a diagnosis of Diffuse large B cell lymphoma of the left parotid was made. After this diagnosis patient underwent systemic imaging studies which showed multiple, ill-defined lytic lesions in the body of C7, DV4-DV6, DV10, LV2, LV4, SV1 and bilateral iliac bones. Bone marrow study was unremarkable. A clinical stage IV disease was established by oncologist and patient started on RCHOP
chemotherapy. As part of RCHOP therapy patient received eight cycles of injection rituximab (375 mg/m²), injection cyclophosphamide (750 mg/m²), injection vincristine (1.4 mg/m²), injection adriamycin (50 mg/m²) and Tablet Prednisolone 100 mg per day for five days. Eight cycles of chemotherapy were given each at 21 days intervals. He responded well and six months after treatment he is on follow-up at our hospital.

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. In the largest study of 40 salivary gland lymphomas, only three cases were seen arising from submandibular gland [2]. Primary parotid lymphomas account for 0.87% of all NHL cases and 4–5% of all extranodal NHLs [3]. Malignant lymphomas of the parotid are uncommon in patients younger than 50 years with peak age at 55 years [4]. Our patient is young individual of 24 years of age. The lymphomas may arise from intraparotid lymph nodes or in the gland itself. If only the intraglandular lymph nodes are involved with no parenchymal infiltration then it should be considered of nodal origin. The distinction is usually difficult as there is extensive, diffuse parenchymal involvement of intraparotid lymph nodes. So primary salivary gland lymphoma is considered when parenchyma of the gland is involved [5]. Most NHLs arising in the salivary glands are B cell lineage including low grade B cell lymphoma of MALT, follicular lymphoma and DLBCL [6]. The DLBCL is a high grade infiltrative tumor associated with destruction of salivary gland parenchyma with tumor cells invading between residual gland acini. On histology the cells are large and atypical and resemble either centroblast or immunoblast as was seen in our case. On the other hand, low grade MALT lymphomas of parotid gland usually arise in setting of benign lymphoepithelial lesion [7].

Most of the lymphoma cases present with painless, firm swelling in the region of salivary gland mimicking other salivary gland epithelial neoplasms. Hence the patient are subjected to imaging studies and other investigations like FNAC which are not very helpful and cause delay in diagnosis. Most of the lymphomas of the salivary gland are surgically treated because they lack definitive imaging features as happened in our case.

Histological examination shows infiltration of monoclonal B cells into the ductal epithelium cells and destroying them. Association between MALT lymphoma and autoimmune diseases like SLE and Sjögren is known [8].

Once the diagnosis of NHL is established it is important to evaluate the patient for any other synchronous systemic involvement or dissemination to decide on the therapy. An Ann Arbor or International Prognostic Index (IPI) scoring system is used to stage the disease. Ours is an aggressive DLBCL of the parotid with metastasis to the vertebrae (stage IV). Treatment depends on clinical staging and irradiation is considered as treatment of choice in localized lesions in early stage. However, it is both or either radiotherapy or chemotherapy which is considered along with surgery in disseminated forms [9, 10]. Our patient underwent RCHOP chemotherapy following total parotidectomy and is on follow-up.

CONCLUSION

Parotid gland swelling is a common presentation in clinical practice and lymphomas affecting parotid gland is clinically indistinguishable from other benign or malignant lesions. Imaging modalities and fine-needle aspiration cytology is not always helpful and hence majority of patients require a parotidectomy for definitive diagnosis.

Author Contributions

Devika Gupta – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

GPS Gahlot – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Vandana Rana – Acquisition of data, Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Rajat Jagani – Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Davendra Swarup – Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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REFERENCES


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