

A Rare Case of a Diffuse Large B-Cell Lymphoma (DLBCL) arising From The Parotid Gland In a Patient With Sjogren's Syndrome

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ABSTRACT

Primary lymphoma of the salivary gland is rare, with NHL being 5% of all extra nodal lymphomas and 2% of all salivary gland tumors. Most are low grade lymphomas with MALT lymphomas being the most common. DLBCL lymphomas are uncommon with most thought to be due to transformation from underlying low-grade lymphomas, postulated to arise in those with sialadenitis and Sjogren's syndrome. We report a rare case of DLBCL in a patient on treatment for Sjogren's Syndrome.

Key words: Sjogren's syndrome, Parotid primary lymphoma, Diffuse large B-cell lymphoma.

Key Messages:

Non-Hodgkin's lymphoma is the most serious complication in Sjogren's syndrome. The high grades are thought to be a transformation from the low grade. This is a rare case of a probable de novo diffuse large B-cell lymphoma.

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INTRODUCTION

Primary lymphoma of the salivary gland is rare, with NHL being 5% of all extra nodal lymphomas and 2% of all salivary gland tumors.^[1] Most are low grade lymphomas with MALT lymphomas being the most common. DLBCL lymphomas are uncommon with most thought to be due to transformation from underlying low-grade lymphomas, postulated to arise in those with sialadenitis and Sjogren's syndrome. We report a rare case of DLBCL in a patient on treatment for Sjogren's Syndrome.

CASE HISTORY

A 63 Year old female, a diagnosed case of Sjogren's Syndrome since 2012 and on oral medications—namely tab. Prednisolone 5 mg od, tab. Azathioprine 50 mg od and tab. HCQ 200 mg od, presented with complaints of fever and painful swelling over her left parotid gland for 2 months duration. Initially received treatment for suspected bacterial parotitis with tab. Levofloxacin and linezolid. Since there was no response, a fine needle aspiration was done which revealed a low grade lymphoma at a general hospital and was referred to our hospital. She has no other comorbidities. On examination- her ECOG PS was 1, afebrile with no pallor or lymph nodes/ tensile enlargement. She had dry mouth and dry eyes. There was a 10*8 cm swelling of her left parotid gland, smooth surfaced however firm in consistency. The overlying

skin was erythematous and non-tender, with dilated veins over the swelling. Systemic examination was unremarkable. Investigations at our hospital—showed a normal CBC, Biochemistry with negative viral markers. The LDH- 152 U/L. there was no evidence of tumour lysis syndrome. The FNAC from the left parotid swelling was suspicious of a lymphoma, hence a trucut biopsy was done. The microscopy of which was reported as a malignant lymphoma and the IHC—showed neoplastic cells positive for CD20 with a high Ki-67. the cells were negative for CD3, CD5, CD23, lambda, Cyclin D1 and LMP-1, suggestive of a Diffuse large B-cell lymphoma. The CT imaging done—revealed a multiloculated heterogeneously enhancing lesion in both the superficial and deep lobes of the left parotid, predominantly in the superficial lobe. The lesion infiltrated the skin posterolaterally, the left para vertebral muscle and anterior wall of the external auditory canal and abutted the mastoid bone. Anteriorly, it abutted the masseter muscle and the left ramus of the mandible with loss of fat planes and compression of the retro mandibular vein. Medially, it extended to the left parapharyngeal space displacing the branches of the ECA and abutting the styloid bone. Few discrete enlarged enhancing lymph nodes were visualized in the left level II, III and V. There were atrophic submandibular and lacrimal glands. The thoracic and abdominal & pelvic imaging was unremarkable. The bone marrow examination showed no involvement of disease.

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Figure 1: Image 1- left parotid swelling; Image 2 – photograph documenting complete regression of the parotid swelling after 5# of chemotherapy.

She was diagnosed as a case of stage II EAX DLBCL and she received 3 cycles of chemotherapy- with standard doses of the CHOP regimen. Reassessment CT scan of the neck revealed a residual lesion and few left cervical lymph nodes. She was continued on CHOP chemotherapy and is doing well with complete regression of the parotid swelling at the end of 5 cycles of chemotherapy. She refused for further chemotherapy and involved field radiation.

DISCUSSION

Primary malignant lymphomas of the salivary gland are uncommon, accounting for 1.7–3.1% of all salivary gland neoplasms and 0.6–5% of all tumor and tumorlike lesions of the parotid gland.² Most cases of lymphoma involve the major salivary glands, frequently the parotid (50–93%) and submandibular glands. These neoplasms may arise from an intraparotid lymph node or in the gland itself.³ Primary malignant lymphomas of the salivary gland are predominantly B-cell type lymphomas and they include MALT-lymphoma, follicular and diffuse large B-cell lymphoma. Rarely T-cell and Hodgkin's do occur.³ Sjogren's syndrome is a chronic inflammatory autoimmune disorder characterized by lymphocytic infiltration of lachrymal and salivary glands, which results in exophthalmia, keratoconjunctivitis sicca and xerostomia; rheumatoid arthritis and hypergammaglobulinemia are also found. Sjogren's syndrome appears in association with reactive and neoplastic lymphoproliferative diseases, frequently with B-cells. This association of benign lymphoepithelial lesions and Sjogren's syndrome is believed to be a precursor lesion for extranodalmarginal B-cell lymphoma of MALT-type.³ The other lymphomas mentioned to arise from the parotid in the general population have been in those with SS. The relative risk of lymphomas in primary Sjogren's syndrome was found to be 16–44 times higher than in the general population in 2 large case series studies.⁴ The prevalence of NHL in SS is 4.3%, the median time to development is around 7.5 years.⁵ Our patient had a diagnosed disease and was on treatment for a 3 year duration. In Sjogren's syndrome, lymphoma genesis

is attributed to local T-cell-dependent antigen-driven proliferation of B cells that eventually become monoclonal and develop into primarily low-grade MALT lymphoma. Sustained antigenic drive during lymphoma transformation is implied by the presence of selected immunoglobulin variable gene mutations and clonal heterogeneity in the hyper variable regions within the tumor cells.⁶ It is known that MALT-lymphoma can progress to a diffuse large B-cell lymphoma but the criterion (on how) to define large-cell transformation is not universally accepted.³ There are reports that diffuse large B-cell lymphoma with a nodal origin have a worse prognosis.³ Our patient was diagnosed as a case of DLBCL arising from the parotid gland and not lymph node. The pathological examination showed no underlying low grade tumor which has commonly been seen in case studies.⁷ this could probably explain her good response to chemotherapy. Patients with primary Sjogren's syndrome have risk factors for progression to lymphoma, such as persistent enlarged parotid glands, splenomegaly, lymphadenopathy, palpable purpura, leg ulcers, peripheral nerve involvement, anemia, neutropenia, low-grade fever, low levels of C3 and C4, and mixed cryoglobulinemia.⁴ No viruses or characteristic chromosomal abnormalities have been associated with these lymphomas in SS.⁸ Epstein-Barr virus (EBV)-positive diffuse large B-cell lymphoma (DLBCL) is rarer and is associated with poorer overall survival than EBV-negative DLBCL.⁴ The LMP-1 which is a surrogate marker for EBV was negative in our patient. The outcomes of these patients with aggressive lymphomas have been reported to be dismal, with a median survival of only 1.8 years.⁵ the survival for all lymphomas associated with SS varying from 0.5 to 1 years in a case series.⁷ Whether the extra nodal origin, de novo origin of the tumor, lack of persistent enlargement of the parotid glands prior to development of the lymphoma and LMP-1 negativity portend a better outcome in our patient will be known in due course.

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