Case Report

Diffuse large B-cell lymphoma presenting as bilateral parotid enlargement, mimicking Sjögren's syndrome

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ABSTRACT

Parotid gland lymphomas are rare with an incidence of 2% of all salivary gland tumors and are therefore commonly overlooked. They can occur *de novo*, arise in the background of a preexistent Sjögren's syndrome, or can be involved in disseminated non-Hodgkin's lymphoma (NHL). We report here a case of a 45-year-old female patient with disseminated NHL, presenting as bilateral parotid enlargement, clinically thought to be Sjögren's syndrome, diagnosed on fine needle aspiration cytology. A high index of suspicion and distinction from its more common mimics such as Sjögren's syndrome is needed to provide a quick and correct diagnosis as well as appropriate management.

Key words: Fine needle aspiration cytology, non-Hodgkin lymphoma, parotid gland, Sjögren's syndrome

INTRODUCTION

Non-Hodgkin's lymphomas (NHLs) constitute only 2% of salivary gland malignancies.[1] They can occur *de novo*, arise in the background of preexistent lymphoepithelial sialadenitis (LESA), or can be involved in disseminated NHL.[2] Involvement of bilateral parotid glands can be clinically indistinguishable from other nonmalignant lesions as well as other more common epithelial tumors causing bilateral affection.[3] Fine needle aspiration cytology (FNAC) is a rapid and effective diagnostic tool which can solve this diagnostic dilemma. This report describes a case of disseminated NHL diagnosed on FNAC, in a clinical setting of bilateral parotid swelling mimicking Sjögren's syndrome.

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Access this article online

Quick Response Code:

Website: www.ccij-online.org

DOI: 10.4103/2278-0513.197885

CASE REPORT

A 45-year-old female presented with bilateral, rapidly progressive, painless parotid swelling for 5 months. She also complained of joint pains and mouth dryness. Clinical impression thus was Sjögren's syndrome. However, SSA Ro and SSB La were found to be negative. She had no history of malignancy or other autoimmune disease. Examination revealed bilateral parotid masses measuring 7 cm × 5 cm and 4 cm × 4 cm respectively. Both were firm, fixed, tense, and nontender. There was no enlargement of the cervical lymph nodes. No hepatosplenomegaly was present. Erythrocyte sedimentation rate was 185 mm at the end of 1 hour, and serum lactate dehydrogenase level was 621 IU/L. Serological tests for human immunodeficiency virus, hepatitis B, and hepatitis C were found to be nonreactive. Serum uric acid, serum globulin, and alkaline phosphatase were mildly raised. Hemogram and other blood investigations were within normal limits, ultrasonography showed multiple

Cite this article as: Agnihotri MA, Sathe PA, Kothari KS, Naik LP. Diffuse large B-cell lymphoma presenting as bilateral parotid enlargement, mimicking Sjögren's syndrome. Clin Cancer Investig J 2016;5:473-5.

hypoechoic lesions in the parotid, suggestive of a neoplastic etiology, and the patient was advised FNAC.

Cytology smears were highly cellular and showed a monomorphic population of large, dispersed, malignant lymphoid cells with high nucleo-cytoplasmic ratio, prominent nucleoli, and brisk mitosis. Background showed mature lymphocytes and many lymphoglandular bodies [Figure 1a-c]. No lymphoepithelial lesions were identified. A diagnosis of high-grade NHL was suggested. Incisional biopsy of left-sided parotid swellings revealed atypical lymphoid cell proliferation with infiltration of the fibrocollagenous tissue and adipose tissue with entrapment of nerve bundles. Immunohistochemistry was performed using standard techniques. The tumor cells expressed CD20 and CD10 and were immunonegative for CD3, CD 138, Bcl2, and AE1/AE3. The Mib-1 labeling index was approximately 90%. Diagnosis of NHL-diffuse large B-cell (DLBCL) type was confirmed. Bone marrow examination showed involvement by CD5 and CD10 negative clonal B-cells, favoring NHL. Positron emission tomographic scan of the whole body revealed active disease in supra and infra-diaphragmatic lymph nodes and extranodal sites such as nasopharynx, hard palate, pleura, lung, stomach, bone, and subcutaneous tissue. The final diagnosis was disseminated DLBCL. The patient was started on chemotherapy following which swelling regressed and the patient is doing well 2 years postchemotherapy.

DISCUSSION

NHL arises from the cellular components of nodal or extranodal tissues.[4] Although head and neck region is the most common site of presentation, involvement of the salivary gland is rare, accounting for 4.7% of lymphomas at all sites.[4-6] The parotid gland is most commonly affected, followed by the submandibular gland.[1,5] Multiple glands, especially bilateral, are involved in about 10% of the cases.[7]

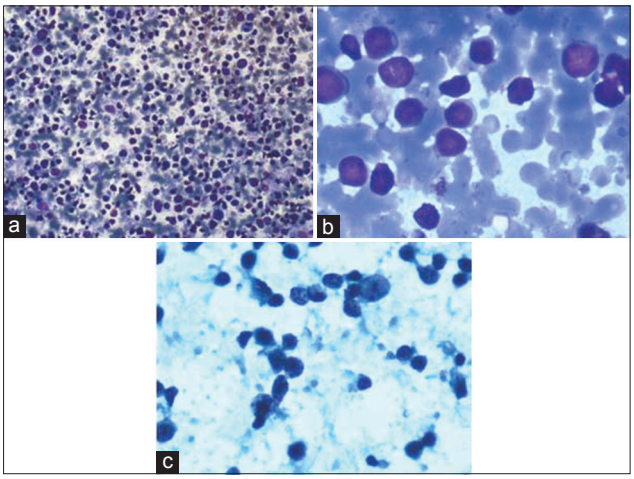

Figure 1: (a) Cellular smears showing monomorphic appearance of cells (Giemsa, ×100). (b and c) Large dispersed malignant lymphoid cells with high nucleocytoplasmic ratio and prominent nucleoli. Background showing lymphoglandular bodies (Giemsa and Papanicolaou, ×400)

Salivary gland lymphoma mostly presents as a painless progressively enlarging mass indistinguishable from other nonneoplastic or neoplastic conditions.[3,6] A predominant involvement of bilateral parotid glands is common in LESA, amyloidosis, cystic lymphoepithelial lesion, Warthin tumor, and acinic cell carcinoma.[8] Since salivary gland lymphoma is uncommon, it is often overlooked and is rarely suspected at initial presentation.[3] In the present case, with bilateral parotid gland enlargement and dryness of mouth with joint pains in a middle-aged female, Sjögren's syndrome was the initial clinical diagnosis.

Lymphomas of the parotid gland can be primary or secondary to disseminated NHL, involving the gland parenchyma or intraglandular lymph nodes.[8] The most common type of primary parotid lymphoma is extranodal marginal zone B-cell lymphoma (EMZBCL) of mucosa-associated lymphoid tissue type. It typically develops in the background of LESA or Sjögren's syndrome.[6,8] DLBCL accounts for about 15% of all NHL of the salivary glands and is speculated that it might represent transformation from the underlying EMZBCL.[7]

Rarely, disseminated NHL can present as bilateral, enlarged parotid glands leading to a diagnostic dilemma. In such situations, FNA is a rapid and reliable tool for making the diagnosis. The primary differential diagnosis on cytology includes reactive intraparotid lymph nodes, LESA, cystic lymphoid hyperplasia, chronic sialadenitis, and epithelial neoplasms with a prominent lymphoid component.[8] However, all these listed lesions show a polymorphous population of lymphoid cells as compared to the monomorphic appearance of intermediate- to large-sized atypical lymphoid cells of lymphoma. The low-grade NHL can sometimes cause difficulty in diagnosis, but a high index of suspicion with appropriate immunophenotyping will confirm the diagnosis.

A thorough clinical examination for nodal and other extranodal sites should follow for staging of the disease as was done in our case.

CONCLUSION

Bilateral parotid gland enlargement is usually seen in Sjögren's syndrome. NHL is rarely suspected at initial presentation. This case emphasizes the importance of FNAC as a primary diagnostic tool which can help the clinician plan appropriate management with minimum inconvenience to the patient.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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