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CASE REPORT

Carcinoma of the Tongue, a Rare Complication in Severe Sjogren's Syndrome: A Description of a Case and a Review of the Literatures

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Introduction

Sjögren's Syndrome (SS) is a chronic inflammatory disorder characterized by diminished lacrimal and salivary gland function. SS occurs in a primary form not associated with other disease and in a secondary form that complicates other rheumatic conditions. The most common disease associated with secondary SS is rheumatoid arthritis. In primary or secondary SS, decreased exocrine gland function leads to the sicca complex, a condition of dry eyes (keratoconjunctivitis sicca) and dry mouth (xerostomia) [1].

Although it has been shown that patients with primary SS have a lifespan not significantly different from that of normal population, primary SS patients have a greater risk of developing lymphoproliferative malignancy [2]. The high incidence of lymphoma in primary SS was first reported in 1963 by Bunium, et al. [3] and has been verified in several studies since [4].

There have been case reports of other cancers, including solid organ tumors, also occurring in patients with primary SS [5]. The data about non-lymphoid cancer is not conclusive, it was suggested that there might be certain subgroups that are at higher risk of developing non-lymphoid cancer [6].

This case report describes a patient with primary SS who developed squamous cell carcinoma of the tongue.

Case Report

A 48-year-old woman with a 2-year history of dry eye, dry mouth, and lower extremities skin rash was evaluated in the rheumatology clinic. Her physical examination revealed conjunctival congestion, mucosal atrophy of the tongue, dental cavities, enlarged parotid glands, and multiple purpuric lesions on both legs varying from 0.5 to 1 cm in size.

Laboratory investigations showed positive anti-nuclear antibody (1:2560, speckled), positive Rheumatoid factor (> 1770 U/ml), positive anti-Sjögren's Syndrome A antigen (SSA) (66 U/ml), anti-Sjögren's Syndrome B antigen (SSB) was negative, erythrocyte sedimentation rate (ESR) was highly elevated (122 mm/hr), C-reactive protein (CRP) (2 mg/dl), serum protein electrophoresis showed polyclonal increase in the gamma region without M-spike. Complete blood count (CBC) showed hemoglobin (12.0 g/dl), with normal red blood cells, white blood cells, and platelets count.

Anti-double stranded DNA (anti-dsDNA), anti-Smith antibody (anti-Sm), anti-neutrophil cytoplasmic antibody (ANCA), and test for cryoglobulins were negative. Complement 3 and complement 4 levels were normal.

Serology for hepatitis B, hepatitis C, and human immunodeficiency virus (HIV) were negative. She was referred to oral surgery for labial salivary gland biopsy, and the result showed chronic sialadenitis with lymphocytic infiltrate consistent with the diagnosis of Sjögren's Syndrome.

She was treated with hydroxychloroquine,

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cevimeline, and local measures for her dry eye and dry mouth. She noticed some improvement in her sicca symptoms and her skin rash resolved.

During follow-up visit in the rheumatology clinic, there was enlargement in her left parotid gland, computerized tomography (CT scan) of the neck was ordered which showed bilateral parotid gland enlargement and parenchymal destruction with fatty infiltration and fibrosis, and multiple cystic and solid intra-parotid lesions within the left parotid gland.

Her condition raises the concern about lymphoma in her parotid gland, she also developed an ulcer on her left lateral margin of the tongue. She was referred to oral surgery for biopsy of her tongue ulcer and was referred to otolaryngology for evaluation of her parotid lesion.

The biopsy of her tongue ulcer showed moderately differentiated squamous cell carcinoma of the tongue, otolaryngology performed left superficial parotidectomy. Histopathologic examination of the excised parotid tissue did not show evidence of lymphoma or squamous cell carcinoma metastases.

She was treated with radiation therapy and chemotherapy for her squamous cell carcinoma of the tongue, surgical resection of the tongue was not done initially to preserve her tongue, however her ulcer did not resolve completely, and repeat biopsy of the tongue ulcer showed invasive moderately differentiated squamous cell carcinoma. She was treated surgically with hemi glossectomy which cured her tongue cancer.

Discussion

A major long-term concern for patients with SS is the development of lymphoma. SS is characterized by polyclonal B cell activation as well as lymphocytic infiltration of the exocrine glands. In ways that remain unclear, the B cell activation in SS predispose some patients to the development of lymphoma. Most lymphoma arises from a reactive infiltrate called lymph epithelial sialadenitis or benign lymphoepithelial lesion [7].

The lifetime risk of non-Hodgkin lymphoma is approximately 5 percent [8], which is 16 to 44 times higher than that of the normal population [9].

Physicians taking care of SS patients face the challenge of early diagnosis and detections of lymphoma. Several predictors of lymphoma development have been identified. Clinical signs such as lymphadenopathy, swollen salivary glands, palpable purpura or skin vasculitis, peripheral nerve involvement, leg ulcers, use of cytotoxic drugs, younger onset primary SS, and laboratory predictors such as anemia, lymphopenia, low levels of C3 and C4, rheumatoid factor positivity, and cryoglobulinemia have been described [10]. The relation between primary SS and other malignant tumors is not well described.

Xerostomia, the primary symptom of SS, can affect the oral mucosa. Xerostomia occurs when the rate of salivary flow is reduced to less than 50%. Signs of xerostomia include dry and cracked lips, oral mucosal sores, and tongue depapillation [11]. There are few reports about squamous cell carcinoma of the tongue in primary SS, Endo, et al. [12] reported a patient with primary SS who developed squamous cell carcinoma of the tongue.

Clinical studies that evaluated the risk of malignancy in primary SS reported few cases of carcinoma of the tongue and oral cavity, the incidence of the reported cases was not significant to suggest a risk higher than the general population similar to what was detected in lymphoproliferative malignancy [10,13].

Squamous cell carcinoma of the oral cavity is rare in patients of age 50 and younger, the classic risk factors include alcohol and tobacco consumption [14].

The patient reported in this article is a 42-year-old woman with no history of alcohol and tobacco use. The younger age for this patient than the common age for most cases of Squamous cell carcinoma of the tongue and the lack of the classic risk factors for this cancer could suggest an etiopathological link between primary SS and Squamous cell carcinoma of the tongue in this patient. This could be related to local chronic inflammation or chronic damage from loss of saliva fluid [13]. The patient reported here had clinical and laboratory features that predict the development of lympoproliferative malignancy. Here parotid gland biopsy showed lymphoepithelial lesion that put her also at risk for developing lymphoma.

Lazarus, et al. [6] confirmed that patients with primary SS have an increased risk of developing lymphoma. The incidence was calculated as being 37.5 times that of the general population. The overall incidence of cancers was 2.6 times that in the general population, but this was almost entirely due to the excess occurrence of lymphoma. Of the 11 patients with lymphoma, five developed additional cancers: one developed a bilateral renal granular cell carcinoma and four developed skin cancers (three basal cell carcinoma and one melanoma). They suggested that there may be a potential link between the development of lymphoma and the subsequent development of additional cancer due to common etiology. One study has shown evidence of defective repair of a promutagenic DNA base lesion, O6-methylguanine, in the lymphocytes of patients with primary SS pre-dispose to lymphoma [15]. This raises the possibility that primary SS patients, pre-disposed to lymphoma, may have defective DNA repair mechanisms in other cell types. A number of oncogenes, cell surface molecules and viruses have also been implicated in the development of lymphomas [16] and these could also possibly have a role in causing additional cancers.

This case report shows that carcinoma of the tongue could be a potential complication to primary SS.

Conflict of Interest

The author declares that he has no conflict of interest.

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