

MALT Lymphoma of Minor Salivary Glands in a Sjögren's Syndrome Patient: a Case Report and Review of Literature

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ABSTRACT

Background: Sjögren's syndrome is a chronic systemic disease, characterized by lymphocytic infiltration and destruction mainly of the salivary and lacrimal glands, resulting in xerostomia and xerophthalmia. Sjögren's syndrome patients have a 44-fold excess risk for the development of non-Hodgkin's lymphoma particularly mucosa-associated lymphoid tissue (MALT) lymphoma, prevalently affecting the major salivary glands. In this report, a rare case of MALT lymphoma of minor salivary glands in a patient with Sjögren's syndrome is described. A review of the published cases of MALT lymphoma located in the minor salivary glands of patients with Sjögren's syndrome is provided.

Methods: In a 64-year-old female patient previously diagnosed with Sjögren's syndrome, an asymptomatic soft tissue mass at the palate was noticed, exhibiting rapid enlargement within one month. With a main differential diagnosis of salivary gland neoplasm or lymphoproliferative lesion, a partial biopsy was performed accompanied by proper immunohistochemical analysis.

Results: A final diagnosis of MALT lymphoma was rendered and the patient was referred for further multidisciplinary evaluation. Gastric endoscopy and biopsy revealed a *Helicobacter pylori*-negative gastric MALT lymphoma, while spleen involvement and bone marrow infiltration were also identified. Patient was classified as having stage IV disseminated disease and a standard chemotherapy protocol was administered; the treatment was well tolerated and resulted in complete remission.

Conclusions: This case emphasizes the need for close monitoring of patients with Sjögren's syndrome by oral medicine specialists, which, besides ensuring proper management of xerostomia and its sequelae, may also lead to early recognition of lymphoma development.

Keywords: MALT lymphoma; minor salivary glands; Sjögren's syndrome.

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INTRODUCTION

Sjögren's syndrome (SS) is a chronic systemic autoimmune disorder, characterized by lymphocytic infiltration of the exocrine glands, mainly the salivary and lacrimal glands, as well as by systemic symptoms and extraglandular manifestations. It may present as a solitary entity (primary SS) or it may occur in conjunction with another autoimmune condition, such as rheumatoid arthritis, systemic lupus erythematosus or scleroderma, in which case it is termed secondary SS [1]. SS has a strong female predisposition (female to male ratio of 9:1), usually beginning during the 4th or 5th decade of life. Patients with SS suffer from classic sicca symptoms of dry eyes and dry mouth, associated with objective evidence of keratoconjunctivitis sicca and xerostomia, respectively. The most significant complication of SS is the development of lymphoproliferative malignancy, which occurs in about 5% of SS patients [2].

Malignant lymphoma, particularly mucosa-associated lymphoid tissue (MALT) lymphoma, is a relatively frequent complication of SS with an incidence ranging between 5 - 10% and a median time from SS to lymphoma diagnosis of 7.5 years [3-6]. Potential explanations for this incidence variation include differences in the criteria used for the diagnosis of SS and the variable duration of follow-up. The risk of lymphoma development in SS patients has been assessed to be 44 times higher compared to the normal population [4]. According to a more recent study, this high risk is probably valid only for selected patient populations with severe disease, while a lower danger appraisal of 16-fold is probably more representative of a low-risk SS subpopulation [7]. According to a meta-analysis of 20 studies, SS exhibits a higher standardized incidence rate (SIR 18.8%) of non-Hodgkin's lymphoma development compared to other immune systemic diseases, e.g. systemic lupus erythematosus (SIR 7.5%) and rheumatoid arthritis (SIR 3.3%) [8].

The purpose of the present report is to describe a rare case of MALT lymphoma of the palatal minor salivary glands, which was detected during routine oral examination of a patient previously diagnosed with secondary Sjögren's syndrome. Additionally, a summary of the published cases of MALT lymphoma affecting the minor salivary glands of Sjögren's syndrome patients as well as the risk factors for lymphoma development in Sjögren's syndrome patients, are also discussed.

CASE DESCRIPTION AND RESULTS

A 64-year-old female patient was referred by her prosthodontist for evaluation of dry mouth and oral burning sensation to the Department of Oral Medicine and Pathology, School of Dentistry, National and Kapodistrian University of Athens, Greece.

The patient reported a 15-year history of secondary SS associated with rheumatoid arthritis, managed by her rheumatologist with hydroxychloroquine and pilocarpine intermittently for several years. Medical history was also significant for haemorrhagic spastic colitis and iron deficiency anaemia, which were managed with appropriate medication.

Clinical examination revealed signs of xerostomia (including lack of saliva secretion during palpation of major salivary glands), erythematous candidiasis (confirmed by a positive cytologic smear) and angular cheilitis. Appropriate systemic (per os fluconazole 100 mg once daily for 15 days) and topical antifungal (miconazole 2% oral gel 4 times daily diffusely on the oral mucosa, and miconazole 2%/hydrocortisone 1% cream 4 times daily on the commissures for 15 days) medication, along with antixerostomic products (topical gel and mouthwash), were prescribed and the patient was placed on regular follow-up. During follow-up, an asymptomatic submucosal mass of soft tissue consistency was noticed at the junction of the hard and soft palate crossing the midline (Figure 1). The lesion, which was not present at the initial visit, exhibited rapid enlargement (reaching approximately 1 cm in diameter) within one month of observation. With a main differential diagnosis of a salivary gland neoplasm or a lymphoproliferative lesion, a partial biopsy was performed.

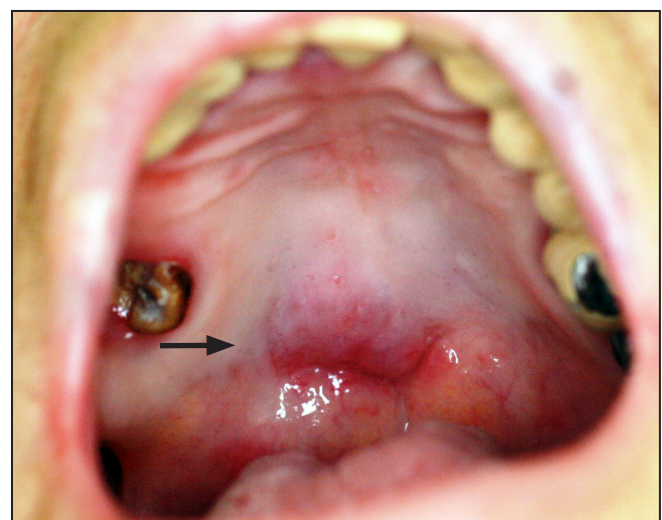


Figure 1. Asymptomatic mass noticed at the junction of hard and soft palate (arrow).

Histopathologically, a dense and diffuse infiltration of minor salivary glands by small sized centrocyte-like neoplastic lymphoid cells with formation of abundant lymphoepithelial lesions was observed. A few scattered blast cells were also noted (Figures 2 and 3). Immunohistochemistry revealed positivity of the neoplastic cells for CD20, CD43 and BCL-2; individual cells were also positive for DBA44 and MUM1. In contrast, stains for BCL-6, CD10, cyclin D1, CD5 and CD3 were negative, while Ki-67 positivity was observed in about 8 - 10% of the neoplastic cells (Figures 4 and 5).

A final diagnosis of an extranodal NHL, MALT-type was rendered and the patient was referred for further multidisciplinary evaluation, disease staging and management to a specialized Hematology-

Oncology Center. Gastric endoscopy and biopsy revealed a *Helicobacter pylori*-negative gastric MALT lymphoma, while further imaging showed spleen involvement. Bone marrow infiltration by neoplastic lymphocytes (at approximately 10%) was identified. Neutropenia, lymphocytopenia, anaemia, low IgM and IgA, as well as elevated β 2-microglobulin, beta 1 globulin and LDH levels were also observed.

The patient was classified as having a stage IV disseminated disease and a standard treatment chemotherapy protocol, including rituximab and fludarabine, was recommended. Although the patient initially refused treatment (for approximately one year), no significant disease progression was noticed during this time after a complete re-evaluation of lymphoma with computed tomography scan

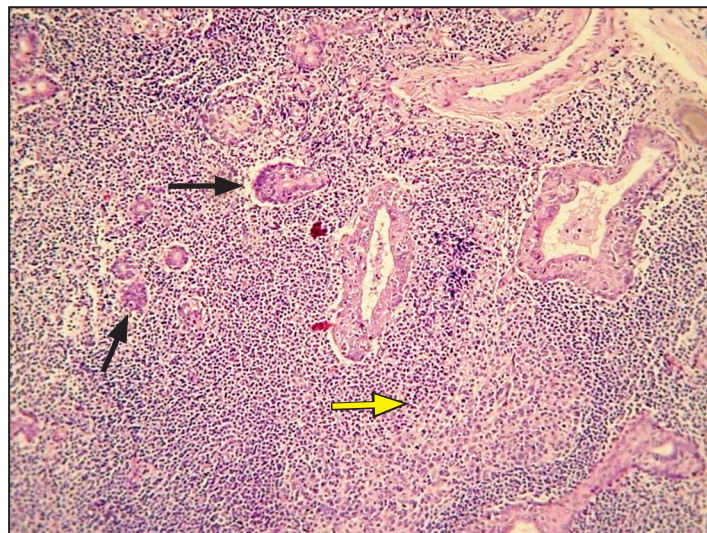


Figure 2. Dense, diffuse infiltration of a minor salivary gland by a monotonous population of lymphoid cells, destroying the normal architecture. An organized lymphoid follicle (arrows) and lymphoepithelial islands (yellow arrow) are also observed (haematoxylin and eosin stain, original magnification x100).

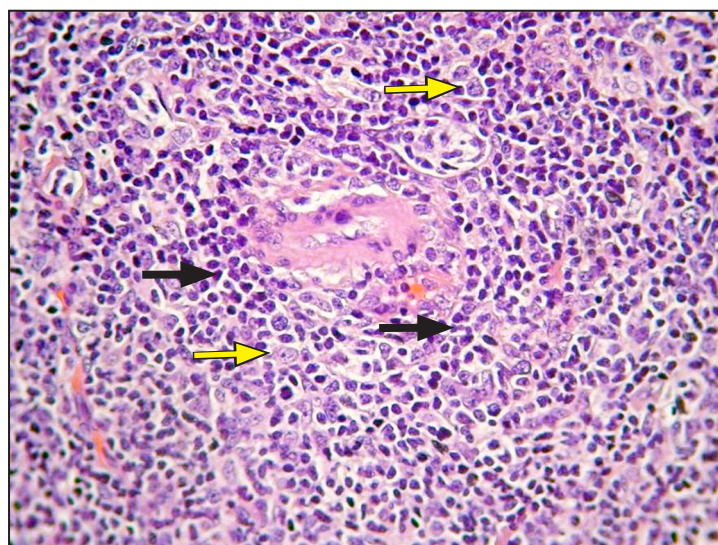


Figure 3. Infiltration by numerous centrocyte-like cells (yellow arrows) is observed; scattered blast cells are also noted (arrows) (haematoxylin and eosin stain, original magnification x400).

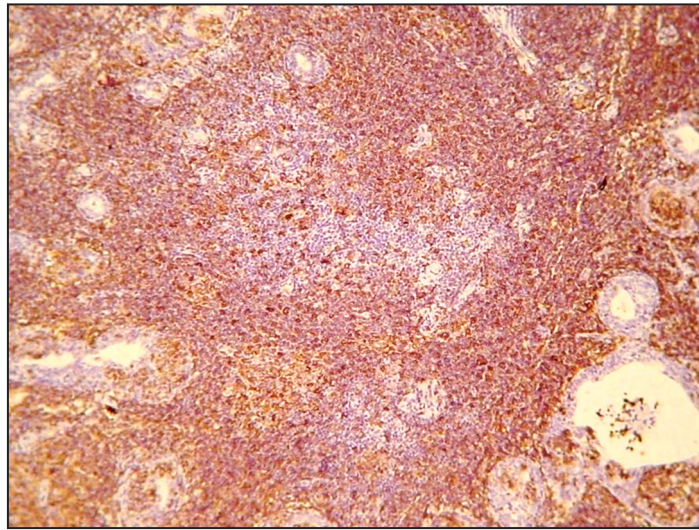


Figure 4. Neoplastic cells exhibiting positivity for anti-CD20 (original magnification x100).

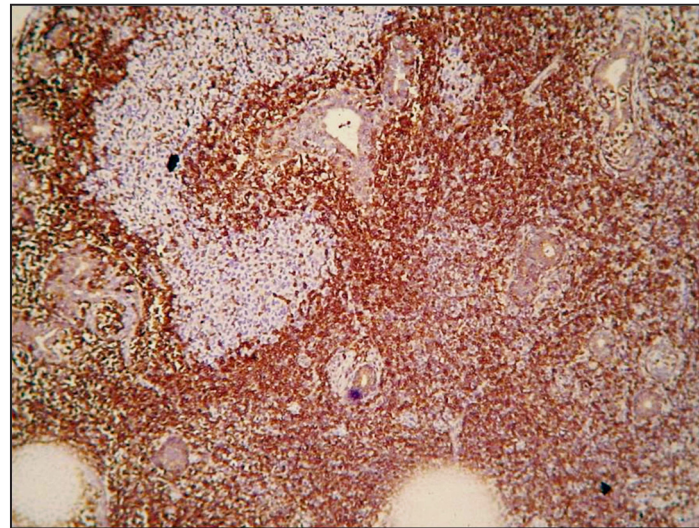


Figure 5. Strong positivity for anti-BCL2 (original magnification x100).

and bone marrow biopsy. Eventually, the patient complied with the suggested treatment; no major complications developed and sustained complete remission of the lymphoma, including resolution of the palatal mass, was achieved; no signs of recurrence were noticed during the subsequent 2 years follow-up period.

DISCUSSION

Malignant lymphoma development is a major and potentially lethal complication in patients with SS. The standardized mortality ratio in SS patients with lymphoma is 3.25 compared to 1.08 in SS patients without lymphoma [9]. Many clinical and biological parameters have been related to lymphoma development in SS (Table 1) [7,10-19]. According to Anderson et al. [11], decrease in serum

immunoglobulin (Ig) levels and the disappearance of a previously positive rheumatoid factor (RF) may herald progression to lymphoma. Kassan et al. [4] showed that SS patients with lymphadenopathy, splenomegaly and persistent parotid gland enlargement share an increased probability of lymphoma development and can be classified as a high risk group.

A variety of histologic subtypes of lymphoma can occur within the salivary glands of SS patients, MALT lymphoma accounting for 46 - 56% of all cases [10,20]. Firstly described by Isaacson and Wright in 1983 [21], MALT lymphoma has been reported in various organs, but the gastrointestinal tract is by far the most common site of occurrence. In SS patients a predilection for extranodal sites is observed with a strong predisposition for major salivary glands, especially the parotid and submandibular glands [20,22]. In contrast, MALT lymphomas affecting minor salivary glands in SS patients appear to be rare,

Table 1. Predictors of lymphoma development in Sjögren’s syndrome

Clinical	Serological
Persistent parotid gland enlargement	Mixed monoclonal cryoglobulinemia
Splenomegaly	Low serum C4 levels
Lymphadenopathy	Serum or urine monoclonal bands
Palpable purpura	CD4 + T lymphocytopenia ^a
Leg ulcers	Neutropenia ^a
Peripheral neuropathy	High serum β2 microglobulin ^a
Previous low-dose irradiation or chemotherapy	Low serum IgM levels ^a
	Disappearance of previously positive rheumatoid factor

^aIndicates detected findings in the present case.

Table 2. Reported cases of MALT lymphoma of the minor salivary glands in Sjögren’s syndrome patients

Author	Year	No. of cases	Age	Sex	Oral location	Other locations	Risk factors	Treatment	Follow-up
Speight et al. [23]	1994	4	NR	NR	Lip ^a	Stomach	NR	NR	NR
			NR	NR	Lip ^a	Palate	NR	NR	NR
			NR	NR	Lip ^a	Cervical lymph nodes	NR	NR	NR
			NR	NR	Lip ^a	Cervical lymph nodes	NR	NR	NR
Van Mello et al. [24]	2005	3	53	F	Lip ^b	Possibly chest area	Non-blanching purpuric lesions on the lower limbs, C4 low	NR	NR
			51	F	Lip ^b	NR	Intermittent swollen parotids, raised levels of total protein, IgG, IgA, IgM, while C4 levels and white blood cell count were decreased	NR	NR
			55		Lip ^a	NR	Intermittent swelling of the parotid gland	Leukeran (4 mg daily) and prednisone 20 mg/day	NR
Pijpe et al. [25]	2005	1	42	F	Hard palate ^b	Right Parotid 10 years earlier neck lymph nodes	Increased IgG	Weekly, course of 4 infusions of rituximab	NS, no recurrence
Sakuma et al. [26]	2006	1	70	F	Hard palate ^a	-	-	Self regression	38 months, no recurrence
Keszler et al. [27]	2012	1	60	F	Lip ^a	-	-	Watch and wait	Evaluation performed every 6 months during the first year and then, once a year, no recurrence
Shwetha et al. [28]	2014	1	62	F	Upper and lower lip ^b	-	-	Watch and wait	NS, no recurrence
Present case	2017	1	64	F	Hard palate ^a	Stomach; spleen; bone marrow	CD4 + T lymphocytopenia, neutropenia, high serum β2 microglobulin, low serum IgM levels	Refused treatment for 1 year (Rituximab + Fludarabine)	2 years, no recurrence

^aMALT lymphoma diagnosed after Sjögren’s syndrome initial diagnosis.

^bMALT lymphoma diagnosed simultaneously with Sjögren’s syndrome.

NR = not reported; F = female; NS = not specified.

with only 11 well documented cases hitherto reported in the English language literature (Table 2) [23-28]. MALT lymphoma is generally an indolent neoplasm and lesions tend to remain localized to the original site for prolonged periods of time, leading sometimes to a mistaken diagnosis of an inflammatory process.

However, in extragastrintestinal areas, the clinical advancement and biologic behaviour is heterogeneous [29]. In the case presented herein, the indolent nature of the neoplasm was supported by the lack of significant disease progression despite the delay in treatment for almost one year.

Selection of treatment regimen for MALT lymphoma depends on the presence of either localized or disseminated disease and the International Prognostic Index (IPI) score based on five parameters (age, stage, involvement of many extranodal sites, performance status and lactate dehydrogenase - LDH). In asymptomatic, localized MALT lymphomas without bone marrow or lymph node involvement and an IPI score between 0 and 1, the patient may receive radiotherapy or remain under close follow-up with no therapeutic intervention. Typically, over an average time of 4 years, these patients may either show no disease progression or exhibit transformation into a high-grade malignant lymphoma [29]. In disseminated MALT lymphomas with a high IPI score, bone marrow and lymph node involvement, cyclophosphamide, hydroxydaunorubicin, oncovin and prednisone (CHOP) chemotherapy regimen is administered with or without rituximab (anti-CD20 monoclonal antibody) [25]. In general, prognosis of MALT lymphoma is satisfactory, with a 5-year survival rate of 90%. The IPI score is of crucial prognostic significance,

with high values indicating patients with a poorer prognosis [30,31].

CONCLUSIONS

Close monitoring of patients with Sjögren's syndrome by oral medicine specialists, ensures not only proper management of xerostomia and its sequelae but may also lead to early recognition of lymphoma development. Especially, high risk Sjögren's syndrome patients should be scrutinized and repeated biopsies may be needed for prompt diagnosis of lymphoma development.

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The authors report no conflicts of interest related to this study.

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