Case Report

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Primary Sjögren's syndrome without ocular manifestation: a case report

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Introduction and importance: Sjögren's Syndrome is a systemic immune disorder, manifested in dry eyes and mouth. Primary Sjögren's syndrome without ocular manifestation is seldom mentioned in the literature.

Case presentation: The authors report a case of a 48-year-old female who complained of dryness of mouth and dysphagia for 6 months. Physical examinations showed dry lips with angular cheilitis, an erythematous tongue, and dry buccal mucosa, with multiple carious teeth. The salivary flow was scanty from the Stenson's and Wharton's ducts on both sides. Her ophthalmological examination was normal. Laboratory tests revealed leukopenia, anemia, thrombocytopenia, elevated levels of C-reactive protein and erythrocyte sedimentation rate, a strongly positive antinuclear antibody, anti-SS-A, anti-SS-, and rheumatic factor. Hyperechoic nodules in both parotids were shown by Ultrasonography. Salivary gland biopsy showed lymphocytic infiltration. Diagnosis of primary Sjögren's syndrome was made. She was treated with Pilocarpine 5 mg for 3 months, Vitamin C, and artificial saliva for oral dryness. She is under continuous follow-up with 50–60% relief, without any systemic complications

Discussion: Sjögren's Syndrome affects the exocrine glands causing dry mouth and eyes, and can cause systemic symptoms, including fatigue and joint pain. The incidence of ocular involvement among the reported cases is 86.1%, whereas our patient did not have any ocular involvement, and this represents a rare condition. The differential diagnosis included diabetes mellitus, hypothyroidism, chronic virology infection, and some medications that cause dryness, which were very much ruled out. Treatment of sicca symptoms involves artificial tears and medications that stimulate saliva flow while treatment of systemic disease includes corticosteroids, and various DMARDs, Rituximab. this disease has an increased relative risk for the development of B-cell non-Hodgkin's lymphoma. Therefore, patients need to be monitored, especially in the presence of risk factors.

Conclusion: It is very important to diagnose this disorder early, using the various diagnostic criteria.

Keywords: case report, dry eye, dry mouth, primary Sjogren's syndrome

Introduction

Sjögren's Syndrome (SS) is a systemic auto-immune syndrome, manifested by dry eyes and mouth, resulting from the infiltration of lymphocyte cells to the exocrine glands, including the lacrimal and the salivary glands[1]. The presence of ocular manifestation among the reported cases is 86.1%[2]. When the ocular symptoms are non-specific, it may lead to a delay in diagnosis. Lack of knowledge about its potential presentations can lead to overlooking the diagnosis and has therapeutic implications[3].

It frequently occurs in the fifth decade of life, with a female predominance and increased risk of developing life-threatening complications, such as lymphoma[4,5]. It is

HIGHLIGHTS

- Sjögren's syndrome (SS) is a systemic auto-immune syndrome, manifested by dry eyes and mouth. It frequently occurs in the fifth decade of life, with a female predominance and increased risk of developing life-threatening complications.
- The presence of ocular manifestation among the reported cases is 86.1%. When the ocular symptoms are non-specific, it may lead to delay in diagnosis, and can lead to therapeutic implications.
- Primary Sjögren′s syndrome (pSS) without ocular symptoms is rarely reported in the literature. Therefore, we intend to emphasize the importance of an early diagnosis of pSS in patients presenting with sicca symptoms as a delay in diagnosis may lead to complications.

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classified either as primary (pSS) when occurring alone or secondary (sSS) when associated with other rheumatic immune diseases[1,4].

The final classification criteria of the American College of Rheumatology with the European League Against Rheumatism are based on the weighted sum of 5 items: anti-SS-A/Ro antibody positivity and focal lymphocytic sialadenitis with a focus score of greater than or equal to 1 foci/4 mm^2, each scoring 3; an abnormal ocular staining score of greater than or equal to 5 (or van Bijsterveld score of ≥ 4), a Schirmer's test result of less than or equal to 5 mm/5 min, and an unstimulated salivary flow rate of less than or equal to 0.1 ml/min, each scoring 1. Individuals with signs and/or symptoms suggestive of SS who have a total score of greater than or equal to 4 for the above items meet the criteria for primary SS[6].

pSS without ocular symptoms is rarely reported in the literature. Therefore, we intend to emphasize the importance of an early diagnosis of pSS in patients presenting with sicca symptoms as a delay in diagnosis may lead to complications.

Case report

A 48-year-old Syrian female came to the Modern Medical Hospital in November 2022, complaining of dryness of mouth along with dysphagia for 6 months; with no other complaints. No past medical, family or personal histories were found.

Her general physical examinations showed dry lips with angular cheilitis, an erythematous tongue (Fig. 1), and dry buccal mucosa, with multiple carious teeth.

The salivary flow was scanty from the Stenson's ducts and Wharton's ducts on both sides. Her ophthalmological examination was normal including clinical examination along with Schirmer-I Test and Rose Bengal Test.

Laboratory tests showed Laboratory examination revealed leukopenia: 3300 mm^3 ($n = 4000–1100$), anemia: hemoglobin 9.8 g/l (n:12–14),and thrombocytopenia:132 000) ($n = 150\,000–400\,000$) C-reactive protein 8.2 mg/dl ($n < 6$), and erythrocyte sedimentation rate 38 mm/h1) (n = 0-20).The rest of the chemical tests were normal. Further workup revealed a strongly positive antinuclear antibody (ANA) screen (1:320 titers), anti-SS-A (Ro) 24, anti-SS-B (La) 18, and rheumatic factor 22 IU/ml. Virology including EBV, parvovirus, herpes simplex 1/2, HIV, and hepatitis panels was all negative.

Chest X-ray chest was normal. Ultrasonography of the parotid gland showed hyperechoic nodules in both parotids. A salivary gland biopsy was taken from the lower lip, which showed lymphocytic infiltration around the ducts and acinus in a small area, proliferation of the lining cells, and formation of epimyoepithelial cell islands, without atypical cells (Figs. 2, 3).

A diagnosis of pSS was made, based on the clinical examination, laboratory evaluation, and the ACR/EULAR criteria[4]. She was treated with Pilocarpine 5 mg QID for three months, chewable Vitamin C, and artificial saliva for oral dryness. She is under continuous follow-up with 50–60% relief, without any systemic complications.

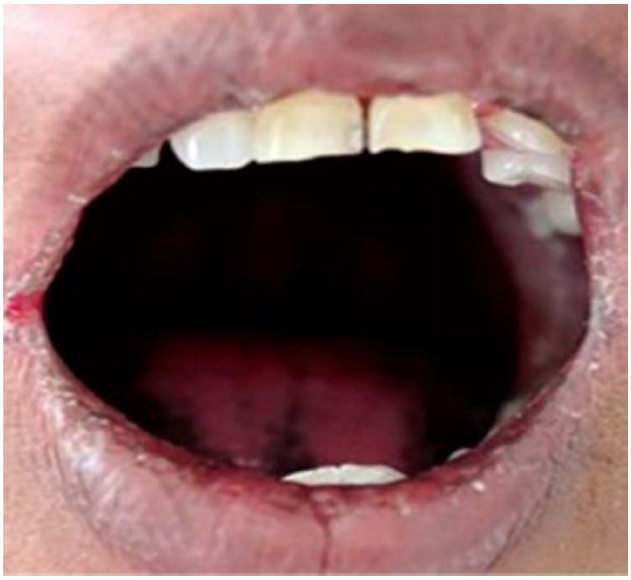

Figure 1. Dry lips with angular cheilitis, an erythematous tongue.

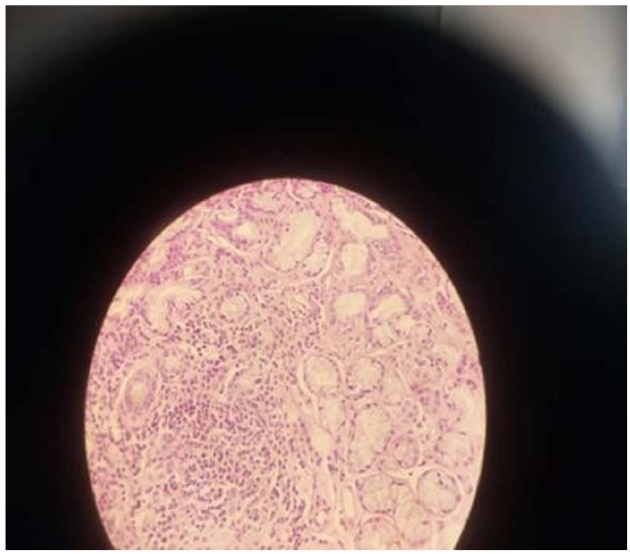

Figure 2. Lymphocytic infiltration in a biopsy of the lower lip.

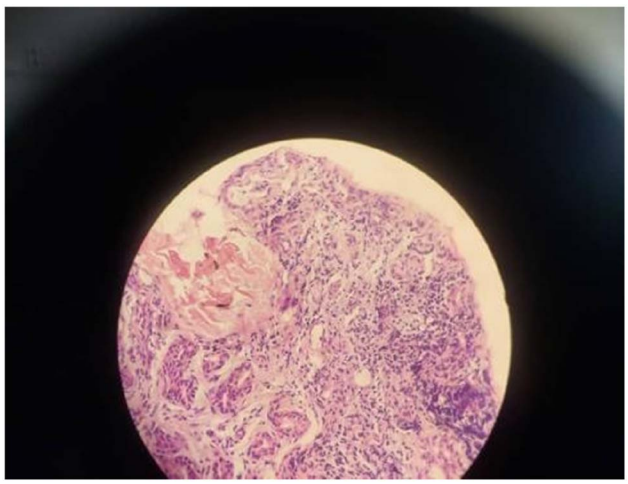

Figure 3. Lymphocytic infiltration in a biopsy of the lower lip.

Discussion

pSS, an auto-immune disorder, targets moisture-producing glands and can cause systemic symptoms, including fatigue and joint pain[1–3]. Typically, it affects women around the age of menopause[2], as in our case.

The incidence of ocular involvement among the reported cases is 86.1%[2,3] whereas, our patient did not have any ocular involvement. To the best of our knowledge, there has been no case report published from Syria on pSS without ocular involvement, and very scarce data is present in the literature. It is reported that the enlargement of the major salivary glands occurs in 25–66% of pSS[3,5,6], which was not manifested in our patient.

Various immunological markers found in different studies showed that RF, ANA, and anti-SS-A, SSB antibodies were positive in different percentages. The sensitivity for Rheumatoid Factor and ANA for pSS is high[7] All these parameters were positive in our patient.

The differential diagnosis included diabetes mellitus, hypothyroidism, chronic virology infection such as Mumps, hepatitis C infection, and HIV infection, sarcoidosis, Parkinson's disease, rheumatoid arthritis, dehydration, treatment causing dryness such as anxiety, antipsychotic, and anticholinergic drugs, and head and neck radiation[3], which were very much ruled out.

To assess disease activity and treatment efficacy, ACR/EULAR criteria were used (Table 1)[4], as we did.

Treatment of SS depends on the extent and severity of the clinical manifestations[3]. Treatment of sicca symptoms involves artificial tears, punctual occlusion, cyclosporine drops, medications that stimulate saliva flow, such as pilocarpine or cevimeline, drinking water, chewing gum, or using saliva substitutes[3,8]. If patients develop yeast infections, anti-fungal therapies may be used[3]. The first approach to systemic major organ-system disease is oral/parenteral corticosteroids, and various DMARDs (methotrexate, azathioprine, and cyclophosphamide) have been successfully employed as steroid-sparing agents[3,8]. Hydroxychloroquine is recommended to treat inflammatory polyarthritis. B-cell depletion may have a role in keratoconjunctivitis sicca, vasculitis, xerostomia, severe parotid gland swelling, and vasculitis). TNF-alpha inhibitors have not proved effective for Sjogren[8].

p SS patients have an increased relative risk for the development of B-cell non-Hodgkins lymphoma[3,8]. Risk factors include persistent salivary gland swelling, enlarging lymph nodes, leukopenia, palpable purpura, and low complement C4 at presentation[3]. Our patient had none of these factors. These patients need monitoring and referring for oncologic consultation when needed.

Mild Sjogren disease has a good prognosis, but those with moderate to severe disease have a very poor quality of life[3]. The dry mouth and eyes often cause irritable symptoms which are not well tolerated[3,8]. With advancing age, the symptoms also tend to worsen[3]. In the long run, patients with Sjogren syndrome will develop a lymphoproliferative disorder, which can lower life expectancy[1–3].

A case report was revealed in literature by Phulambrikar *et al.*[2], that described a case of a 45-year-old female, who presented with dry mouth and dysphagia, without any ocular and systemic manifestations, treated with Pilocarpine 5 mg QID for three months, chewable vitamin C lozenges, and artificial saliva 'Bioxtra' for oral dryness.

The strength of our case is that this is a case report of pSS with xerostomia, without keratoconjunctivitis, which is a rarity and hardly reported in the literature.

The limitation of a case report refers to the limited possibility of generalizing its validity and the impossibility of establishing a cause-effect relationship.

Table 1

ACR/EULAR criteria classification of pSS.

Inclusion criteria	Score[a]
Labial salivary gland with focal lymphocytic sialadenitis and a focus score of ≥ 1[b]	3
Anti-SS-A antibodies (anti-Ro)	3
Ocular staining score, in at least 1 eye[c] ≥ 5 (or van Bijsterveld score ≥ 4)	1
Schirmer test in at least 1 eye ≤ 5 mm/5 min	1
Unstimulated whole saliva flow rate ≤ 0.1 ml/min	1
Patients with exclusion criteria do not have this syndrome:	
Head and neck radiation treatment	
Active hepatitis C infection	
AIDS	
Sarcoidosis	
Amyloidosis	
Graft-vs-host disease	
IgG4-related disease	

pSS, primary Sjögren's syndrome.

[a]Inclusion criteria included patients with a score of ≥ 4, at least 1 symptom of eye or oral dryness.

[b]A focus score is the infiltration of at least 50 inflammatory cells present in 4 mm^2 of gland surface unit.

[c]Ocular staining score (OSS) :is performed using a sequential examination with fluorescein dye for the cornea, followed by lissamine green dye to assess the conjunctiva, the total score of OSS is (0-12).

Conclusion

This is a rare hardly reported literature case of pSS without keratoconjunctivitis. Therefore, we intend to emphasize the importance of an early diagnosis of pSS in patients presenting with sicca symptoms as a delay in diagnosis may lead to serious complications such as lymphoma. Despite the limited clinical presentation of our patient, the early diagnosis of this rare disorder was possible by using the latest ACR/EULAR diagnostic criteria.

Ethical approval

Ethical approval was obtained.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Author contribution

A.M.: writing the paper and data collection. S.B.: writing the paper. Z.M.: writing the paper. A.A.: data collection. H.A.: data collection. M.K.: writing the paper and data analysis.

Conflicts of interest disclosure

There are no conflicts of interest.

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Data availability statement

Data are available.

Provenance and peer review

Not commissioned, externally peer-reviewed.

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