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Sjogren's Syndrome Presenting as Sialadenitis – A Rare Case

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Abstract:
Sjogren's syndrome, an autoimmune disease with lymphocytic glandular infiltration, can be primary or secondary. It is more commonly seen in middle aged females above 50 years of age. Sjogrens syndrome mainly affects the ocular and oral system of the body, eventually causing keratoconjunctivitis sicca (dry eye) and xerostomia (dry mouth). It has been commonly seen in association with other immune disorders like lupus and rheumatoid arthritis. The index case involves a 50-year-old female who was initially diagnosed with sialadenitis; however, later turned out to be a case of Sjogrens syndrome on a detailed clinical and histopathological evaluation.

Keywords: Keratoconjunctivitis sicca, xerostomia, rheumatoid arthritis, focus score, histopathology.

INTRODUCTION

Sjogren's syndrome is a multisystem autoimmune disease characterized by hypo-functioning of lacrimal and salivary glands along with multiorgan manifestations.[1-2] The median age at the time of diagnosis is 40 years with female preponderance. It is a disorder of immune system presenting with two common symptoms, i.e., keratoconjunctivitis sicca (dry eyes) and xerostomia (dry mouth). The moisture secreting glands of eyes, mouth and mucus membranes are firstly affected thereby causing dry eyes and dry mouth.[2-4] These patients are immunocompromised and hence present with dental cavities, oral thrush, blurred vision & corneal damage. The patients can also present with swollen salivary glands, prolonged fatigue, skin rashes, dry skin, joint pain, swelling and stiffness. Rare complications like bronchitis, pneumonia, hepatitis, cirrhosis of liver, peripheral neuropathy can also be noted in few patients.

Keeping in mind the varied presentation of this syndrome, a proper clinicopathological approach is needed to arrive at an early diagnosis and intervention. Here, we present a case of Sjogrens syndrome in a 50-year-old female patient who was initially diagnosed as Sialadenitis.

CASE REPORT

A-50-years-old female patient presented to the hospital with swelling on both sides of preauricular region for last one year [Figure 1a]. The swellings measured approximately 2x2 cms in size, and were diffuse, immobile, firm in consistency and tender on palpation with no local rise of temperature. Often pain was intensified after ingestion of food.

Fine needle aspiration cytology showed few clusters of ductal epithelial cells and lympho-histiocytic aggregates along with some polymorphs in a background of mucoid material. Based on cytological evaluation, a diagnosis of Acute Sialadenitis was made. Ultrasonographic findings revealed bulky and diffusely hypoechoic parotid glands bilaterally with small hypoechoic nodular lesions. NCCT neck revealed enlargement of superficial lobe of bilateral parotid glands. Also noted were multiple enlarged non-significant bilateral cervical lymph nodes.

Hence patient was treated as a case of acute sialadenitis. Adequate hydration, oral antibiotics, and anti-inflammatory drugs were prescribed for 7 days. They were extended for another 7 days on next follow up. Even after 15 days of conservative treatment there was no improvement. But this time the patient gave history of foreign body sensation in throat, dry cough and history of tooth extraction few months back. On further questioning she complained of dryness of mouth and dryness of eyes, skin rashes and coating of tongue [Figure 1b&c].

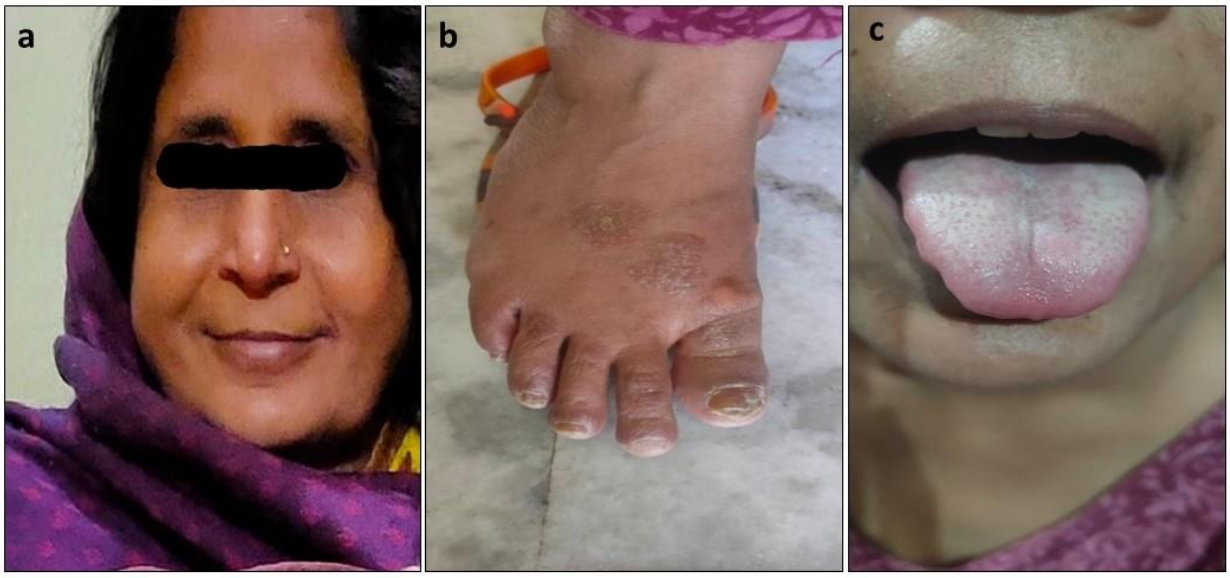

Figure 1a-c: a- Clinical image showing bilateral parotid enlargement; b- Skin rash over right foot; c- Oral candidiasis

Routine investigations including hemogram, ECG, liver n kidney function tests, chest x ray were done. Mediastinal widening and prominent bilateral hilum appeared on chest x ray. CECT thorax was done which showed bilateral apical fibro-parenchymatous changes. Positive Schirmer test and Rose Bengal test showed evidence of dry eyes. RA factor (256) was positive for Rheumatoid arthritis (Normal range: 0-20). Anti-RO/SSA (12.30, normal range being 0.1-3.5) and anti- LA/SSB (20.7, normal range being 0.1-3.5) were found to be positive.

Further, on the basis of above tests labial salivary biopsy was done for this patient. Sections examined showed lymphocytic sialdenitis with a positive focus score of 2; consistent with Sjogrens syndrome [Figure 2a-b]. Hence the diagnosis of primary sjogrens syndrome was made and treatment was started accordingly. The patient was followed up for 6 months and was doing well till the last follow up period.

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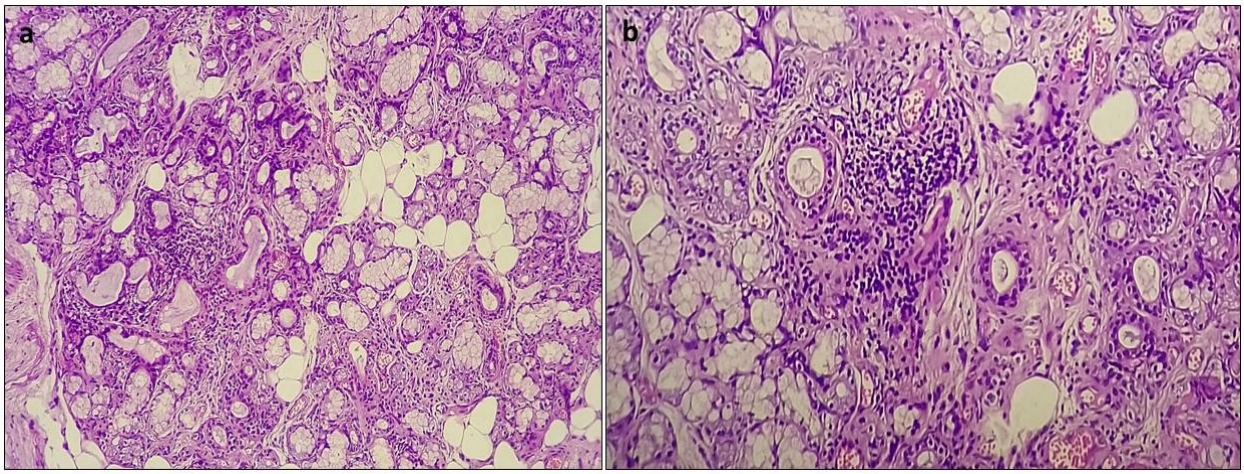

Figure 2a-b: Sections examined showed sheets of ductal epithelial cells with dense aggregates of lymphocytes surrounding the ducts along with interspersed adipocytes

[Figure 2a- H&E, 20X and Figure 2b- H&E, 40X]

DISCUSSION

Sjogrens syndrome (SS) is a slowly progressive autoimmune disease that exhibits a wide range of organ specific and systemic manifestations. Although exact etiology is not known, immunological background with genetic and environmental predisposing factors are blamed for this disease. Both B and T lymphocytes are involved in SS. Non organ specific autoantibodies are found in approximately 60% of patients with SS.[2-5] They are rheumatoid factors, antinuclear antibodies and antibodies to small RNA protein complexes RO/SSA and LA/SSB. These autoantibodies are responsible for tissue dysfunction.

SS can either occur alone in absence of an underlying disease (Primary SS) or in association with systemic autoimmune rheumatic disease (Secondary SS). Secondary Sjogren's syndrome is associated with another underlying rheumatic disease such as SLE (systemic lupus erythematosis), RA (rheumatoid arthritis) or scleroderma. Prevalence of secondary SS has been reported to be 4 – 31 % depending on the criteria applied, methodological design and associated connective tissue disorder.[3-6] Among patients with RA prevalence ranges between 4 – 31 %, in SLE between 8 – 19% and in scleroderma between 14 - 29 %.[3-6] Secondary SS has not been studied extensively and there is doubt whether secondary SS is merely a manifestation of underlying disease or a true overlap of primary SS with another connective tissue disease.

SS poses a diagnostic challenge with prevalence of approximately 2% of adult population. It has been seen that more than half of the cases remain undiagnosed. Since the wide range of symptoms of SS do not appear together ophthalmologist, dentist and ENT specialist treat each symptom individually unaware that a systemic disease is present.[4-7] The index case also presented with bilateral parotid swellings and was treated in the line of management of sialadentis as was demonstrated in FNAC. Only later when a battery of investigations was ordered, did the diagnosis of SS got confirmed in labial salivary biopsy and with positive anti-RO & anti-LA antibodies. The patients of SS are frequently misdiagnosed as their symptoms mimic those of other diseases. Also, symptoms are deceptively non-specific, and spectrum of clinical manifestations is wide.

SS is more common in perimenopausal women. The symptoms of cutaneous, oral and vaginal dryness may be attributed to menopause. Xerostomia, xerophthalmia and xerotrachea can be contributed to a topic disease and anxiety. However, xerophthalmia is most prominent ocular feature. Itching, grittiness, soreness, decreased visual acuity, corneal erosions can all result because of xerophthalmia.[1, 5-8] Xerostomia is another feature of the triad of SS. The patients have difficulty in eating dry fruits, soreness and foreign body sensation in throat. Usual pooling of saliva in floor of mouth is absent and there are increased chances of dental caries and dental cavities. Angular cheilitis associated with candidiasis may coexist. Absence of glandular secretions of respiratory tract can lead to dryness of nose, throat and trachea resulting in persistent hoarseness and a chronic non-

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productive cough. Involvement of exocrine glands of skin leads to skin dryness, itching, rashes. Leukocytoclastic vasculitis resulting in palpable purpura with slightly raised haemorrhagic skin lesions are quite common findings, as noted in our patient also.

The differential diagnosis of SS includes conditions that can cause parotid swellings, xerostomia and xerophthalmia.[5-8] Bilateral parotid swellings can be seen in acromegaly. metabolic diseases like DM, chronic pancreatitis, cirrhosis or infections like mumps, HIV, sialadenitis. Dry eyes can be caused by amyloidosis, blepharitis, pemphigoid, sarcoidosis, steven Johnson's syndrome. Dry mouth can be seen in conditions like DM, amyloidosis, sarcoidosis, viral infections and trauma.

There is no single diagnostic test for SS. Clinical diagnosis of SS is made in presence of compatible clinical and laboratory features and after exclusion of other causes of ocular and oral dryness. Objective findings of oral or ocular dryness are because of glandular parenchymal damage. The patient has an objective marker of dry eye (Schirmer test < 5mm/ 5 minute or abnormal ocular surface staining) or salivary hypofunction as seen in Sialometry. Alternatively, MRI or ultrasound evidence of significant glandular parenchymal abnormality is noted in such patients. Serological evidence of autoimmunity shows the presence of anti-RO/SSA antibodies with or without anti-LA/SSB antibodies.[3-8] A positive labial salivary gland biopsy (focus score >=1) aids in the confirmation of the disease. Often, a well-established systematic rheumatic disease, i.e., RA, SLE or systemic sclerosis is also noted in the patients of SS.

A complete workup of SS involves a battery of investigations and coordination of multiple specialists like rheumatologists, ENT specialists, ophthalmologists and dentists. For accurate diagnosis of SS, proper assessment of ocular and oral involvement is essential. Schirmer and Rose Bengal tests are effective in demonstrating dry eye. Sialometry, parotid sialography and salivary gland scintigraphy demonstrates decreased salivary flow. Minor salivary gland biopsy remains highly specific test for salivary component of SS. Focal lymphocytic sialadenitis defined as multiple dense aggregates of 50 or more lymphocytes (1 focus) in perivascular or periductal areas in the majority of sampled glands is characteristic histopathologic feature of SS.[6-8] Serological and laboratory findings, several autoantibodies that includes rheumatoid factors, antinuclear antibodies and antibodies to RO/LA are found positive in 80% of patients.

Treatment is mainly symptomatic depending on extent and severity of clinical manifestations and requires a multidisciplinary approach.[7-8] Treatment is intended to limit the damage resulting from xerostomia and keratoconjunctivitis. Artificial tears, muscarinic agonists such as pilocarpine and cevimaline hydrochloride have therapeutic effects on xerophthalmia. Salivary substitutes are recommended for patients with severe dryness and no residual salivary function. Dental care and frequent dental examinations are required. Antifungal drugs, like clotrimazole lozenges can be given for oral candidiasis. NSAIDS provide relief from minor musculoskeletal problems. Corticosteroid use is limited to treatment of extraglandular manifestations of SS. They can be given intermittently in pruritus, vasculitis and renal tubular acidosis. Humidification and secretagogues can help manage xerotrachea.

CONCLUSION

Sjogren's syndrome is an autoimmune disease which can be primary or secondary (in association with underlying disease). The disease presents with wide spectrum of clinical manifestations and all of them don't present simultaneously leading to delay in diagnosis and treatment. Increased awareness of SS and a multidisciplinary teamwork of specialists are required for its early diagnosis. Diagnosis should be suspected in individuals with persistent dry eyes/ mouth, parotid gland enlargement and dental caries. SS is non-life threatening and treatment is conservative. The patients should be prescribed appropriate treatment as early as possible to prevent complications and improve lifestyle of the patients of SS.

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