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SJOGREN'S SYNDROME- A CASE REPORT

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ABSTRACT

Sjogren's Syndrome (SS) is a systemic autoimmune chronic disorder affecting the exocrine glands that is characterized by infiltration by lymphocytes leading to cumulative loss of glandular function. This case report depicts a detailed clinical and labarotory investigations required for diagnosis of Sjogren's syndrome.

Keywords- Sjogren 's syndrome, Rheumatoid Arthritis, Immunoassay.

INTRODUCTION

Sjogren's Syndrome (SS) is a systemic autoimmune chronic disorder affecting the exocrine glands that is characterized by infiltration by lymphocytes leading to cumulative loss of glandular function.[1]It was first described by Swedish ophthalmologist Henrik Sjögren hence the name Sjogren's syndrome.

SS occurs with an incidence of 50%in patients with rheumatoid arthritis (RA) affecting 0.5% to 1.0% of the population. The symptoms can occur in people of any age but most commonly occurs in patients aged 45 to 55 years with marked predilection in females with a ratio of 9:1 to males.[2]

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The clinical manifestations progress as the severity increases. SS typically exist in two forms which are-primary form which is characterized by dryness of the mouth and eyes, also known as sicca syndrome, and the secondary form where characteristic connective tissue disorders such as rheumatoid arthritis, systematic lupus erythematosus occur along with ocular dryness and xerostomia.[3]

To prevent advancement and complications, early diagnosis is important. Here, we present a case of SS, with aim to lay further emphasis on prompt diagnosis using advanced diagnostics and management protocol.

CASE REPORT

A 50 year old female patient reported to the Department of Oral Medicine with a chief complaint of dryness and burning sensation in mouth since 1 year. On elicitation of medical history the patient reported that she is under medication for diabetes since five years and reported of joint pains in both hands and feet since 2 years. On clinical examination, extraoral swelling was seen on the right and left parotid region , measuring approximately 3 cm × 2 cm, extending from the tragus of the ear to the angle of mandible.Ear lobe was lifted on both sides (Fig 1). On palpation the swellings were firm, diffuse, non-movable; which was warm and tender with normal overlying surface texture. She also presented with dryness of eyes and non tender, movable, palpable bilateral sub mandibular lymph nodes.

Intra-oral inspectory findings revealed the presence of melanotic diffuse pigmentation on left and right buccal and palatal mucosa. Depapillation was seen in anterior 2/3rd and lateral border of tongue(Fig 2). Reduced salivary pooling was present along with severe xerostomia.Tongue blade sign was positive. On milking of the parotid glands, it revealed a muco-purulent discharge (Fig 3). The exudate was collected in a sterile syringe and was sent for histopathological evaluation. A provisional diagnosis of generalised stomatitis with subacute sialadenitis and papillary atrophy was formulated. Differentials included Sjogren's syndrome, IgG4-related Disease- Küttnertumor, Mikulicz disease, Benign lymphoepithelial sialadenitis, Dacryoadenitis.The patient was then referred for an ophthalmologist's opinion, blood investigations and USG of bilateral salivary glands. Ultrasonography showed bilateral parotid and submandibular gland enlargement and multiple hypoechoic lesions showing high vascularity. Hemogram revealed high erythrocyte sedimentation rate & iron deficiency anaemia. Enzyme linked immunoassay revealed high positivity for ANTI RO and ANTI-LA antibodies, as well as ANA was positive on immunofluorescence. These antibodies reveal the

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presence of connective tissue disorders like rheumatoid arthritis or systemic lupus erythematosus along with SS.

Ophthalmic opinion confirmed the presence of ocular dryness with positive Schirmer's test and rose bengal dye test. The patient however was unwilling for lip biopsy.

Based on the clinical presentation and investigations a final diagnosis of SS was formulated (revised American-European diagnostic criteria) [4]

The patient was prescribed tab Augmentin 625mg twice daily for 5 days paracetamol 500mg . Xerostomia was managed by Wet mouth solution and xerophthalmia by Maxmoist topical application four times daily.

DISCUSSION

SSincludes a triad of xerostomia, xerophthalmia and connective tissue disorder ,with rheumatoid arthritis being the most common. Some of the general symptoms include fatigue, arthralgia/arthritis, myalgia/myositis, and associated lymphadenopathy. Neuropathy, thyroiditis, interstitial lung disease, chronic atrophic gastritis, celiac disease, primary biliary cirrhosis and other liver symptoms, vasculitis, glomerulonephritis, hearing problems, vaginal dryness, dyspareunia, and interstitial cystitis are some of the internal organ alterations seen in such patients.[5][6][7]

Although a number of criteria are suggested for the diagnosis of SS, the foremost widely accepted is the American-European Diagnostic criteria. According to the criteria, the above case showed the presence of rheumatoid arthritis along with ocular, oral signs , salivary gland involvement and presence of serum antibodies indicative of secondary SS.

In addition to ultrasonography, sialography of the glands show the presence of diffuse sialectasia(branch less fruit laden appearance) and salivary scintigraphy may show reduced uptake, and reduction in concentration of tracer and delay in excretion.[4]

The treatment protocol is predicated on the management of sicca symptoms and extraglandular manifestations of the syndrome by a multidisciplinary approach. The symptomatic treatment involves use of salivary substitutes and artificial tears to stop episodes of conjunctivitis. Extraglandular features are often managed by corticosteroids but however should be limited in patients with severe organ damage and leukopenia.
[8]

A systematic review on treatment protocols by Ramos-Casals M et. al concluded that there are presence of enough evidence in literature that pilocarpine and cevimeline are often used for sicca features and topical cyclosporine for moderate to severe dryness of the eyes.[9]

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SS features a complex and unknown pathogenesis, but it's likely to be related to benign lymphoephelial lesions. due to the invasion of lymphocytic cells, 6–10% of patients develop into lymphoma.[10]

CONCLUSION

Prompt diagnosis using advanced investigative modalities will help in the diagnosis of SS. Regular follow up necessitates the importance of the progress of the disease. Patient education by the clinician will also help to efficiently manage the symptoms thus improving the quality of life.

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FIGURES & LEGENDS

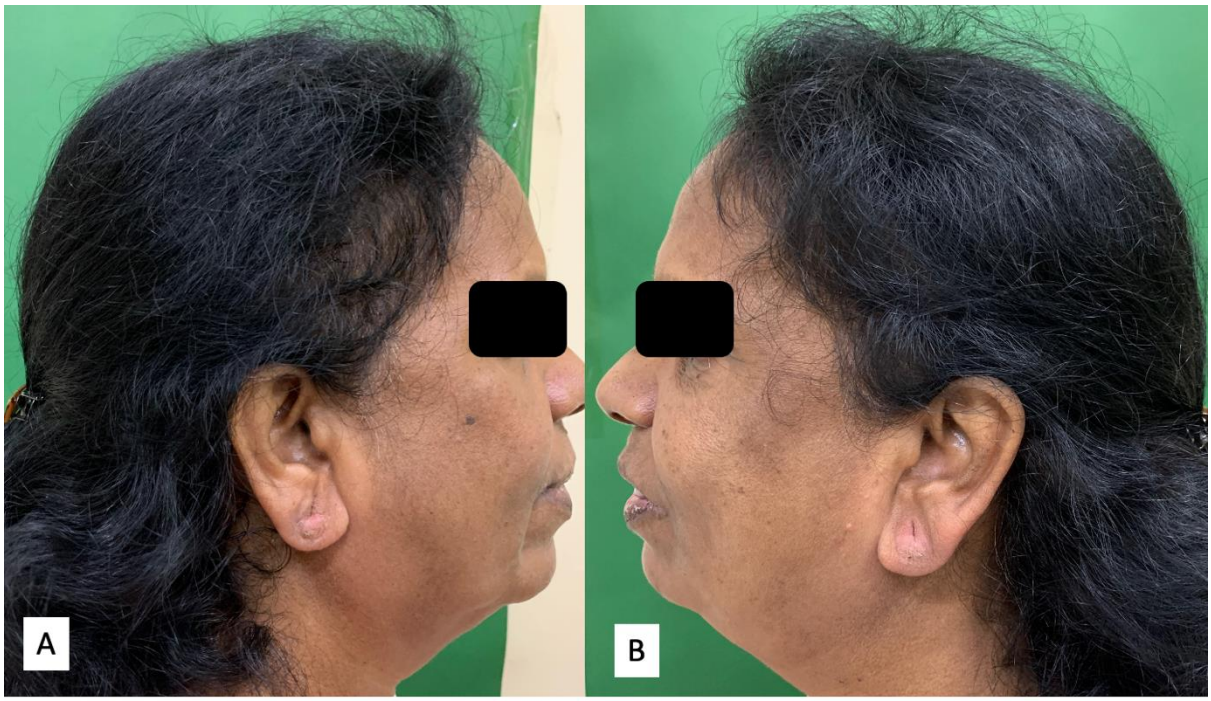

FIG. 1A & 1B

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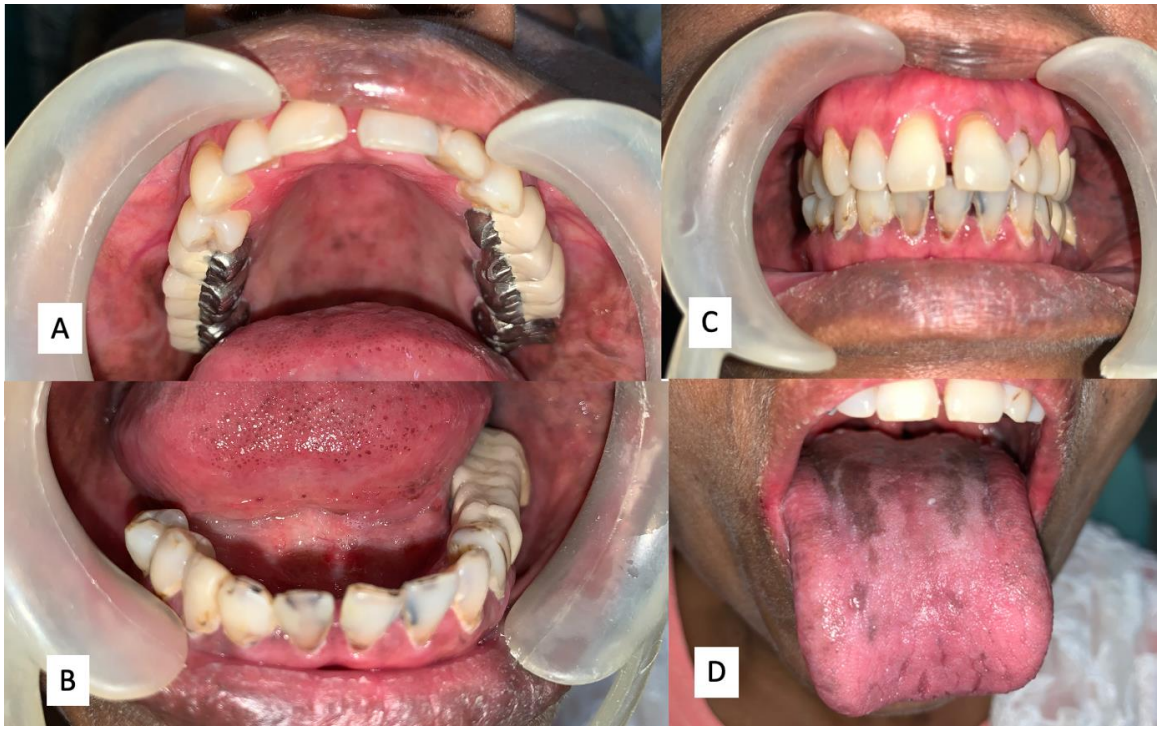

FIG- 2A, 2B, 2C, 2D

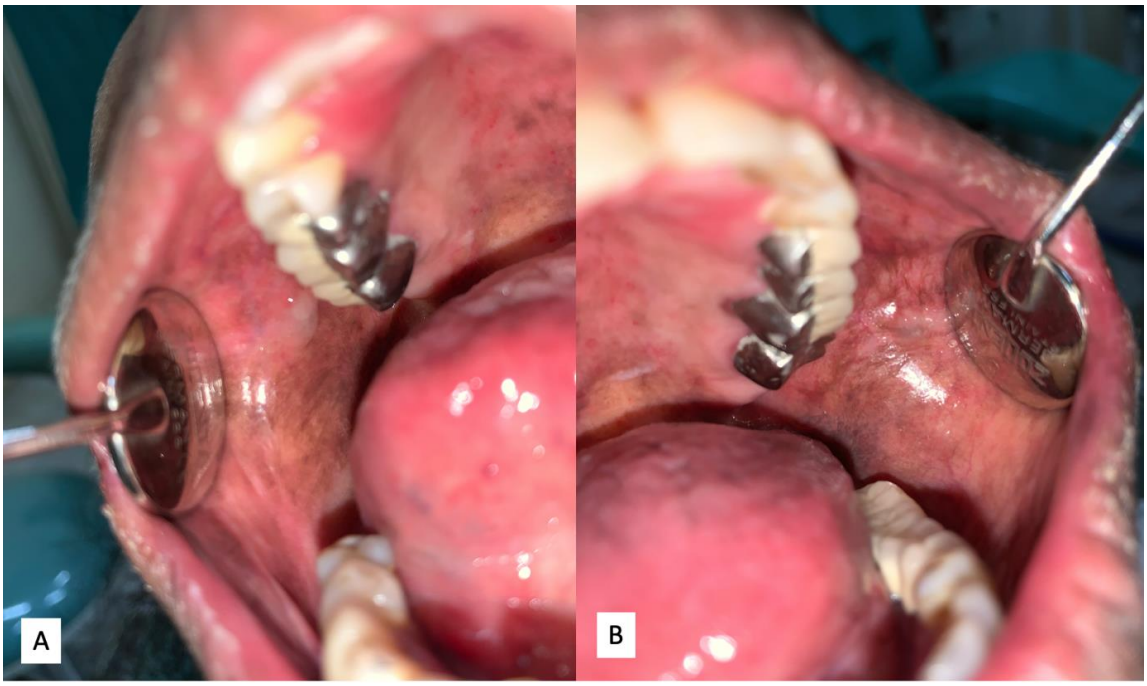

FIG- 3A, 3B.

LEGENDS-

FIG 1A & 1B- Extraoral bilateral swelling of parotid glands.

FIG 2A, 2B, 2C,2D- Intraoral examination revealing multiple caries, prostheses along with generalised stomatitis and papillary atrophy of the tongue.
FIG 3A & 3B- Mucopurulent discharge bilaterally from Stenson's duct.