INTRODUCTION

Sjögren's syndrome (SS), either primary or secondary, has been defined as an autoimmune epithelitis characterized by lymphocytic glandular infiltration and various extraglandular manifestations [1], [2]. Transcending Copenhagen diagnostic praxis, SS is nowadays diagnosed according to the European-American inclusion and exclusion criteria and classification. Clinical manifestations of SS develop gradually along its pathological course. The first clues in primary SS are, most often, lacrimal hypofunction (xerophthalmia), and dry mouth (xerostomia) secondary to hyposalivation which result from self-perpetuating immune-mediated loss of acinar and ductal cells of lacrimal and salivary glands. In secondary SS, rheumatoid factors and several extraglandular manifestations are concomitant with such xerophthalmia and xerostomia. This includes neural, renal, rheumatological, vascular, gastric and pulmonary manifestations.

Case 1

A 20 years young female married in Feb 2018 who conceived and underwent missed abortion in 18th April and again conceived 2nd time with positive UPT on 27 June! Considering high risk pregnancy kept in regular ANC by obstetrician without any obvious medical problems and USG done on 01/09/18 for foetal well being showed single live foetus of 7 weeks 4 days age and both mother and foetus were doing well till 27/11/2018 when found foetal bradycardia on routine ANC! Considering high risk pregnancy kept under missed abortion in 18th April and again conceived 2nd time.

Patient was referred to physician who advised T.Dexona (4mg)/day and also she have mild low backache and heel pain and tested high titres of RA factor and ACPA fulfilling classification criteria to Diagnose RA! Her APLA work up were negative! and patient advised for histopathological examination of lip biopsy(minor salivary gland biopsy). Under local anesthesia, four minor salivary glands from a normally appearing labial mucosa were harvested. The extract was immersed immediately in 10% formalin to be submitted for microscopic examination. The histologic picture viewed a confluence of lymphocytic infiltrate which replaced most of the glandular parenchyma (greater than fifty lymphocytes in several foci).

Histopathological examination of lower lip biopsy specimen done .DERMIS SHOWS: a) increase in the number of glands and few duct, heavy periglandular lymphocytic infiltration (>100 lymphocyte/hpf)
b) Perivascular lymphocytic infiltration also seen c) Stromas shows diffuse lymphocytic infiltration with area of fibrosis consistent with the diagnostic criteria of “Primary Sjogren's syndrome (pSS)”.

Case 2

Savitri Kunkal, age 25/F with Anti Ro positivity diagnosed of Sjogren Syndrome, ANA.

ABSTRACT

Sjögren syndrome is chronic, systemic autoimmune disease characterized by lymphocytic infiltration of the exocrine glands. It is an elaborate exclusion criteria and classification. Clinical manifestations of SS develop gradually along its pathological course. The first clues in primary SS are, most often, lacrimal hypofunction (xerophthalmia), and dry mouth (xerostomia) secondary to hyposalivation which result from self-perpetuating immune-mediated loss of acinar and ductal cells of lacrimal and salivary glands. In secondary SS, rheumatoid factors and several extraglandular manifestations are concomitant with such xerophthalmia and xerostomia. This includes neural, renal, rheumatological, vascular, gastric and pulmonary manifestations.

KEYWORDS

Sjogren Syndrome, ANA
ANA .Anti Ro (Anti-SSA) positivity, so referred to Rheumatologist!

She was evaluated by Rheumatologist and found to have dry mouth and carries teeth and minimal synovitis of small hand joints and malar rash. Patient was advised for lip biopsy (minor salivary gland biopsy).

Histopathological examination of lower lip biopsy specimen shows typical of Sjogren’s syndrome (SS). DERMIS SHOWS: a) periglandular lymphocytic infiltration (>50 lymphocyte/hpf) b) perivascular lymphocytic infiltration c) area of fibrosis consistent with the diagnostic criteria of “Primary Sjogren’s syndrome (pSS).”

She was on steroid, Hydroxychloroquine (HCQ), calcium and Vit D3 supplementation! Her dry mouth improved and advised for dental consultation and preconceptional counseling done for next pregnancy for better pregnancy outcome!

DISCUSSION -
Sjögren's syndrome is an Autoimmune Exocrinopathy involving lacrimal and salivary glands leading to progressive destruction of glands resulting in dry eyes and mouths (Cardinal symptoms of the disease), it may exist alone (primary SS) or in association with other Connective tissue disease (secondary Sjogren's syndrome). Primary SS constitutes only 0.5% in India and is an uncommon and under diagnosed entity due to lack of awareness amongst treating physicians, ophthalmologist and dentist and even gynaecologist as in this case! The absence of joint deformities, joint erosions on hands/feet radiograph, normal CRP, marked hypergammaglobulinemia, Presence of Anti-La antibodies help in differentiating primary SS from that associated with RA! Almost half of asymptomatic mother giving birth to children with CHB ultimately develop SS! Anti-Ro/SSA and Anti-La/SSB antibodies are detected in 50 - 70% of Primary SS patients! Anti-Ro/SSA and Anti-La/SSB antibodies positivity are of younger age at diagnosis as this case! Autoimmune CHB occurs in 1-2% of Anti-Ro/SSA antibodies pregnancies, developed in absence of cardiac structural abnormalities and has recurrence rate of 12 - 20% in subsequent pregnancy as in this index case and CHB usually diagnosed between 18 and 24 weeks gestation by foetal echo! Autoantibodies against Anti-CCP in SS have been estimated 3-10%. RA factor commonly found in sera of SS patient and is associated with Anti-Ro/SSA and Anti-La/SSB positivity as well as systemic disease as this case with non erosive and deforming Arthritis!

CONCLUSION -
Primary SS is uncommon and under diagnosed disease and incidence rate only 0.5% due to lack of awareness amongst physicians, ophthalmologist, Dentist and Gynaecologist. Autoimmune CHB occurs in 1-2% of Anti-SSA-Ro positive mother and half of the mother giving birth to CHB children developed SS latter, so there should be high index of suspicion and require multidisciplinary approach including Rheumatology service as main part of the treatment team!

REFERENCES