ORIGINAL RESEARCH PAPER
Volume-9 | Issue-3 | March-2020 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

HISTOPATHOLOGICAL FINDING IN LIP BIOPSY OF PATIENT SUSPECTED WITH AUTO IMMUNE DISEASE PRESENTING WITH FETAL LOSS: A RARE Case study at RIMS

Pathology

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ABSTRACT

Sjögren syndrome is chronic, systemic autoimmune disease characterized by lymphocytic infiltration of the exocrine glands. It is an elaborate involvement of the lacrimal and salivary glands, which eventually lead to keratoconjunctivitis sicca and xerostomia. It may occur in two forms - Primary and secondary, which is associated with another autoimmune disease, most commonly rheumatoid arthritis. Numerous criteria were proposed for diagnosis of Sjögren syndrome. Most widely accepted are American and European group developed international classification criteria for Sjögrens syndrome. These criteria include ocular symptoms, oral symptoms, ocular signs, histopathology, salivary gland involvement and sialography. The classification requires four of the six items, one of which must be positive minor salivary gland biopsy or a positive antibody test. Early diagnosis is important to prevent further complications. The aim of this paper is to emphasis on oral changes, advanced diagnosis, and management of Sjögren's syndrome.

KEYWORDS

Sjogren Syndrome, ANA

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 manifestions are concomitant with such xerophthalmia and xerostomia.This includes neural, renal, rheumatological, vascular, gastric and pulmonary manifestations

Case 1

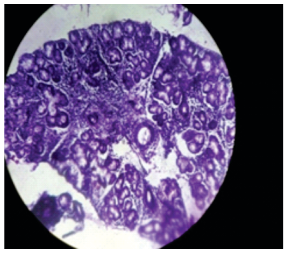
A 20 years young female married in Feb 2018 who conceived and underwent missed abortion in 18th April and again conceived 2nd time with positive UPT on 27 June ! Considering high risk pregnancy kept in regular ANC by obstetrician without any obvious medical problems and USG done on 01 /09/18 for foetal 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 ! So again advised USG for foetal well being showed gestational age of 20 weeks and 7 days with normal flow of uterine artery and absence of diastolic notch . Also treating obstetrician advised for foetal echo showed fetal bradycardia (HR - 82 beats /mins) with calcified aortic valve leaflets! Paediatrician reference sought and Autoantibodies work up done !

ANA -5.9(neg <0.8),anti-dsDNA- 0.4 (neg <20),antiSSA(Ro60)-6.47(neg <1) ,AntiSSB(Ro52) - 5.91 (neg <1) antiSSB(La) - 6.31 (neg <1) ,No APLA workup done !

Patient was referred to physician who advised T.Dexona (4mg) /day from 06-04-18 till 22 -02-2019 ! inspite steroid foetal bradycardia (FHR ~80 -100 beats/mins) persisted ,at that time patient has severe dry mouth ,dry Eye and hairfall but no joint symptoms! USG for foetal well being showed live foetus with gestational age 31 weeks and 1 day ! patient noticed less foetal movement 2 days later with Absence of fetal HS examined by her obstetrician USG done showed IUD ,so admitted and continuous induction done for spontaneous expulsion which occurred on 28 February 2019 .

The patient visited to Rheumatology clinic on and retrospectively history reviewed and it revealed that she was symptomatic since July 2018 around second conception with H/o Dry mouth ,hairfall ,teeth carries but no rashes and joint pain ,they neglected which aggravated approximately 20 weeks of the pregnancy but still joint pain were absent ! When evaluated in Rheumatology OPD pateint had mild synovitis in MTPs 2nd -4th bilaterally which confirmed by HRUSG 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 adviced 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) Stroma shows diffuse lymphocytic infiltration with area of fibrosis consistent with the diagnostic criteria of " Primary Sjogren's syndrome (pSS)



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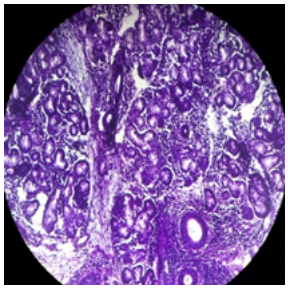

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Case 2

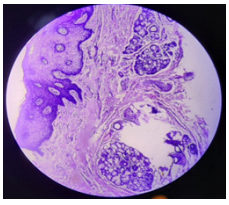
Savitri Kunkal age 25/F with Anti Ro positivity diagnosed retrospectively after early pregnancy loss !

This young female who was under Ante natal checkup,underwent missed abortion in 10 weeks of pregnancy inspite of utmost precaution ,So her obstetrician order immmunological workup and found to have

ANA ,Anti Ro (Anti -SSA) positivity,so refered to Rheumatologist!

She was evaluated by Rheumatologist and found to have dry mouth and caries teeth and minimal synovitis of small hand joints and malar rash . patientwas adviced 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 ,Hydroychloroquine (HCQ) ,calcium and Vit D3 supplementation !Her Dry mouth improved and advised for dental consultation and preconceptional counselling done for next pregnancy for better pregnancy outcome!



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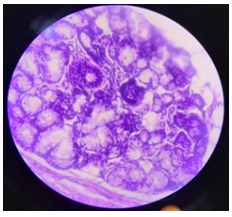

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DISCUSSION -

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 hypergamaglobulinemia,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 Gyanecologist. 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 !

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