Case Report:

Tearless lady with yellow eyes

Sharma S K*, Sangram Mangudkar*** , Ajinkya Dhakne**, Ramdas Barure**, Swaraj Shelke*, Sukritsingh Sethi*, Suryaprakash R Kothe***

***Professor in Medicine, ** Assistant professor in Medicine, *Junior Resident-3,
Department of Medicine, Padmashree Dr. D. Y. Patil Medical College,Pune – 411018, India
Corresponding author: Dr. Ajinkya B Dhakane,

Abstract:

Sjogren's syndrome is a chronic autoimmune disorder with unknown cause and usually secondary to other autoimmune connective tissue disorders. Herein, we report a patient with autoimmune hepatitis and thyroiditis associated with Sjogren's syndrome presenting with pancytopenia.

Keywords: Sjogren's syndrome

Introduction

Autoimmune disorders usually affect women and tend to be together with two or more disorders in one patient. Moreover; patients with any autoimmune disease are prone to develop other diseases.[1] Increased titers of serum autoantibodies which target specific tissues and hypergammaglobulinaemia determine disease characteristics. Therefore; symptoms and range between slight fatigue to severe tissue damage. In this paper we present a patient who has Sjogren's syndrome, autoimmune hepatitis, and thyroiditis with pancytopenia.

Case report

40 year old female patient presented with yellowish discoloration of eyes & urine, swelling & pain in abdomen & Right parotid region, loss of appetite, low grade fever, dryness of mouth & eyes, bleeding through nose since 2-3 months. Patient also had H/O jaundice 2years back similar symptoms reoccurring since then for same symptomatic treatment taken outside. No The laboratory examination showed Hemoglobin 5.0gm%, TLC 3800, Platelet count 1.8lakh/cumm, Retic count 4.6%, negative serology for hepatitis A, B, C & D. We found a considerably reduced liver function with low albumin and prothrombin time(1.25), as well as a moderate elevation of liver enzymes(ALT 100,AST 161,ALP 183) and a high bilirubin(3.1mg/dl), hypergammaglobulinaemia(1.82gm%) were found. Schirmer's test showed abnormality. Liver biopsy consistent with autoimmune hepatitis & parotid biopsy suggestive of lymphocytic infiltration with chronic sildenitis. Autoimmune antibodies were performed showing high titers of antinuclear antibodies (ANA), nRNP/Sm, SS-A, Ro-52, Antimicrosomal antibody (AMA-M2), Anti dsDNA. Ultrasound examination revealed hepatic parenchymal changes, splenomegaly. We diagnosed an autoimmune hepatitis with Sjögren's syndrome by means of laboratory values, histological findings and detection of typical autoantibodies. The patients score for autoimmune hepatitis according to international autoimmune hepatitis group scoring system was 22.[2,3]

Patient was discharge on Tb.Prednisolone 10mg, Tb. Azathioprine 50mg with significant clinical improvement.

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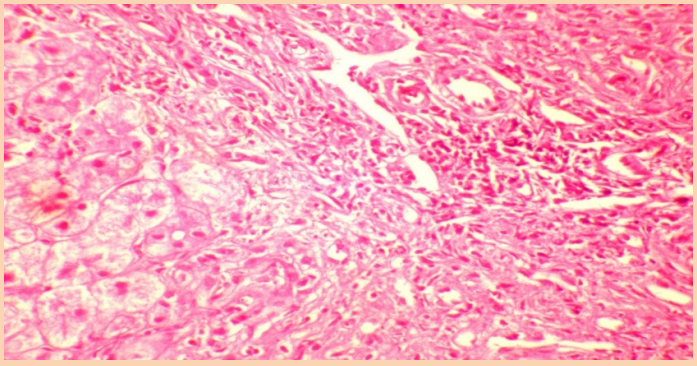
1 The initial and final laboratory values of the patients

	Normal	Presentation	Discharge	Three months after
White blood (cells/cumm)	4000-11000	3800	4600	7200
Hemoglobin (gm %)	11.5-14.5	5.0	7.6	9.8
Platelet count (cells/cumm)	150000-450000	180000	200000	230000
Reticulocyte (%)	1-2	4.6		
ESR (mm/hr)	10	48	32	`18
ALT	0-40	100		
AST	0-40	161		
ALP	0-110	183		
Total Bilirubin (mg/dl)	0-1.2	3.1	2.4	1.9
Thyroid stimulating hormone	0.3-5	8.78		
Free T_3	2-4.4	2.1		
Free T_4	0.9- 1.8	0.87		
Prothrombin time (INR)	1.25	1.13	1.12	1.08
Total proteins (gm/dl)	6.5-8	4.8	5.5	6.3
Albumin (gm/dl)	3.5-5	2.8	3	3.7



Liver Biopsy picture

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Discussion

Sjogren's syndrome (SS) is a chronic, slowly progressive autoimmune disease characterized by lymphocytic infiltration of exocrine glands resulting in xerostomia and dry eyes. The disease presents alone (primary Sjogren's) or in association with other autoimmune rheumatic diseases (secondary Sjogren's). Although SS is an autoimmune exocrinopathy, involvement of non exocrine organs such as thyroid, kidneys and skin have been reported. At the same time chronic autoimmune hepatitis, digestive disorders, diabetes mellitus, pulmonary disorders, Raynaud's phenomenon, joint and muscle disorders, peripheral neuropathies and depressive syndromes are reported. [4] Furthermore, it can be associated with number of other autoimmune disease especially autoimmune hepatitis and thyroiditis.[5-7] We report this case as a rare coexistence of SS, AIH and autoimmune thyroiditis with pancytopenia. Our patient presented with completely non-related symptoms of this overlapping autoimmune disorders. She was admitted with anemia and advanced investigation in the hospital showed that she had Sjögren's syndrome, autoimmune liver disease and thyroiditis.

The main symptoms of SS are xerostomia and keratoconjunctivitis Sicca caused by B lymphocytic infiltration of exocrine gland.[8] In our patient initial main symptom was easy fatigability, yellowish discoloration of eyes and dryness of mouth and eyes. Our patient showed deranged liver function test and underwent liver biopsy which was suggestive of interface hepatitis and lymphocytic infiltration. Ultrasound shows liver enlarged and decreased echogenicity. Parotid biopsy suggestive of lymphocytic infiltration and sialadenitis with enlarged on ultrasound with multi hypoechoeic lesions. We used scoring system and full response to treatment supporting our diagnosis. The patients score was 22 for autoimmune hepatitis according to international autoimmune hepatitis group scoring system (in his system an aggregate score greater than 15 prior to therapy constitute a definite diagnosis of AIH. A score of 10-15 is interpreted as probable AIH).[2, 3]The presence of autoantibodies of thyroid and thyroid dysfunctions has previously been reported in SS. Thyroid autoantibodies can be 28-38% positive in SS.[9] In our case thyroid hormones were TSH was raised with fT_3 and fT_4 were normal and autoantibodies were positive suggestive of subclinical hypothyroidism (Hashimotos thyroiditis).

Conclusion

Elevated liver enzymes should always be further investigated. Autoimmune hepatitis is a rare disorder and its association with Sjögren's syndrome is further rarer. Rapid response to immunosuppressive therapy, such as Prednisolone, Azathioprine is characteristic. Early diagnosis and therapy are essential for the patient's prognosis.

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