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General Medicine

A RARE CASE OF AUTOIMMUNE HEMOLYTIC ANAEMIA IN SJOGREN'S SYNDROME.

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ABSTRACT Sjogren syndrome (SS) is an autoimmune disease characterized by lymphocytic infiltration of the salivary glands and lacrimal glands. This syndrome also has various manifestations associated with other exocrine glands and non glandular tissues. 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. However, Autoimmune hemolytic anaemia (AIHA) in Sjogren's syndrome is a rare occurrence. We report a case of an elderly women with AIHA secondary to Sjogren's syndrome.

KEYWORDS : Sjogren's syndrome, AIHA

Introduction:
Sjögren's Syndrome (SS) is a chronic systemic autoimmune disorder, characterized by the lymphocytic infiltration of lacrimal and salivary glands respectively. The disease is usually diagnosed at around the fifth decade of life, with a female to male ratio of 9:1. Transcending Copenhagen diagnostic praxis, SS is nowadays diagnosed according to the European-American inclusion and exclusion criteria and classification . Although the exact etiology of SS is not totally fathomed, immunological background, with genetic and environmental predisposing factors, are blamed for priming this disease. There, apoptosis, IFN signaling, cytokine levels, expression of autoantigens, and T-cell and B-cell dysfunction are all likely to be of (a) salient role(s) in understanding the etiopathogenesis of SS . 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 selfperpetuating 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 . However, Autoimmune hemolytic anaemia (AIHA) in Sjogren's syndrome is a rare occurrence. The present study reports an atypical case of AIHA secondary to Sjogren's syndrome in a 47-year-old female.

Case report
A 47 year old woman came to the OPD with complaints of
1)fever
2) burning micturition and
3)dry mouth since 2 days.
On General examination : She was found to be pale and cachexic (Fig 1).

Figure 1: General Examination
On Systemic Examination Gross splenomegaly was found which was then confirmed on ultrasound (Fig 2) .

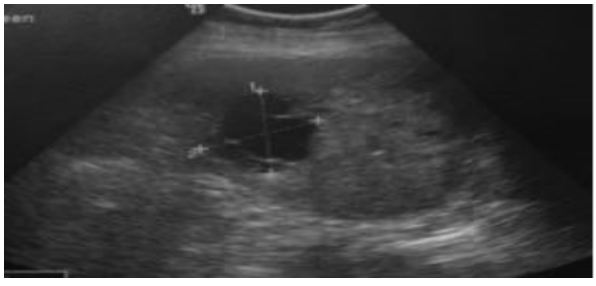

Fig 2 Enlarged spleen with well defined hypoechoic and hyperechoic areas.
On Blood examination her Hb was found to be 3.2g/dl and PCV 8.1 %, Peripheral smear showed microcytic hypochromic RBCs , red cell agglutinates, spherocytes, target cells, polychromatophils and nucleated RBCs (Fig 3), Reticulocytes count of 30 % (Fig 4) with no hemoparasites and was reported as Autoimmune Hemolytic Anaemia (AIHA).

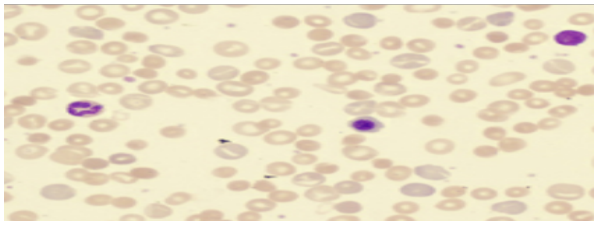

Fig 3: Peripheral smear

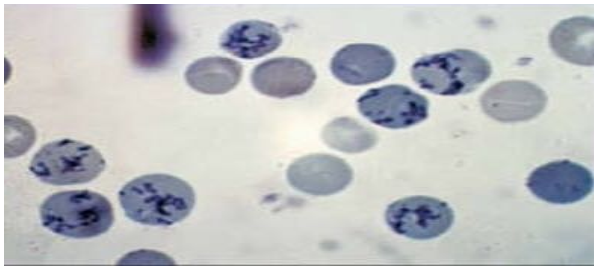

Fig 4 : Supravital stain

showing reticulocytes
Further investigations included Coomb's tests , both Direct Coob's Test (DCT) & Indirect Coombs Test (ICT) were positive.

High Performance Liquid Chromatography (HPLC) showed no abnormal haemoglobinopathy but elevated HbF of 2.5 % was noted.

Further, ANA profile was done which showed very strong positivity (++++) for SS-A and Ro-52 autoantibodies indicating Sjogren's syndrome according to latest The American college of rheumatology classification (ACR) diagnostic criteria.

Schirmer's Test was also positive.
Based on the clinical findings & laboratory investigations, it was concluded that the patient had AIHA due to underlying Sjogren's syndrome. Treatment with immunosuppressants was initiated which showed an improvement.

Discussion

Sjögrens syndrome is a defined as a clinical symptom complex. It is an autoimmune destruction of exocrine glands (primary salivary and lacrimal)that produces the clinical manifestations of dry mouth, dry eyes (keratoconjuctivities sicca), and in more than 50% of cases, parotid gland enlargement. Primary Sjögren syndrome is diagnosed when the syndrome is limited to this pattern of involvement. However, this pattern of involvement may be a manifestation of another -well-defined autoimmune disease such as rheumatoid arthritis, systemic lupus erythematosus, or primary biliary cirrhosis. In this context, it is referred to as secondary Sjögren syndrome. To the best of our knowledge there has been no case report published from India on AIHA secondary to Sjogren's syndrome , and very scarce data is present in the literature.

Treatment of Sjögren's syndrome depends on the extent and severity of the clinical manifestations and is better instituted through a multidisciplinary approach. Symptomatic treatment includes artificial tears, salivary substitutes to relieve the symptoms and prevent local infectious complications like conjunctivitis and corneal inflammation, development of caries and periodontal disease, a thorough dental preventive program should be implemented in all cases. Corticosteroid treatment should be reserved for all the cases showing evidence of organ damage, significant leukopenia or severe clinical symptoms.

This is a case report of Autoimmune hemolytic anaemia (AIHA) in Sjogren's syndrome , which is a rarity, and hardly reported in literature. In spite of the limited clinical presentation of our patient the early diagnosis of this rare disorder was possible by using the latest ACR diagnostic criteria. However, Primary Sjogren's syndrome should be included in the differential diagnosis of acquired AIHA.

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