The Libyan International Medical University

Faculty of Basic Medical Science

Primary Sjögren syndrome clinical and prognostic correlation

Narden Mahgoub

2146

Supervised by: Dr. Osama Othman

Assisted by: Marwa Yousef

Report Submitted to fulfill the requirements for Scientific Research Activity

Date of Submission: 12/03/2020

Abstract

Sjogren's syndrome (SS) is a slowly progressing autoimmune disease, affecting predominantly middle-aged women, with a female to male ratio reaching 9:1. It is characterized by lymphocytic infiltration of the exocrine glands, mainly the lacrimal and salivary glands, resulting in reduced secretory functions and oral and ocular dryness. The syndrome can present alone as primary SS (pSS) or in the context of underlying connective tissue disease as secondary SS (sSS).While the pathogenesis of the disease remains elusive, environmental, genetic and hormonal contributors seem to be involved. Over the last years, compelling evidence has suggested a pivotal role of the epithelium in orchestrating the immune response in the histopathological lesion of Sjogren's syndrome and the term “autoimmune epithelitis” has been proposed as an etiological term. Although the clinical manifestations of pSS patients are mainly those of an autoimmune exocrinopathy, almost half of patients develop extraglandular disease, which may be manifested either by epithelial lymphocytic invasion of lung, liver, or kidney (resulting in interstitial nephritis) or by skin vasculitis, peripheral neuropathy, glomerulonephritis, and low C4 levels. The latter reflect immune-complex mediated disease and confer increased risk for lymphoma development.[1]

Introduction

Sjögren's syndrome (SS) is a chronic autoimmune disorder characterized by decrease salivary and lacrimal gland function, due to lymphocytic infiltration of these glands, leading to progressive destruction, Sjogren's syndrome is a multifactorial condition with genetic, environmental and hormonal factors playing a role in establishing the condition. B-cell activating factor (BAFF) is an important mediator in the induction and perpetuation of this condition patients are classified as having primary SS (pSS) and secondary SS. In pSS, decreased exocrine gland function leads to the 'sicca complex', a combination of dry eyes (Xerophthalmia) and dry mouth (xerostomia). Secondary SS (sSS) is associated with other rheumatic conditions, of which the most common is rheumatoid arthritis .

The clinical manifestations of SS include both exocrine gland involvement and extra-glandular disorder features. SS occurs in a primary form not associated with other diseases and cause this manifestation xerostomia, keratoconjunctivitis sicca, and parotid gland enlargement. Primary SS, symptoms do not always present concurrently , Prognosis is predicting the likely or expected development of a disease, including whether the signs and symptoms will improve or worsen (and how quickly) or remain stable over time.

The aim of this report is to study the clinical manifestation and prognostic correlation of PSS .[2]

Material And Methods

In this report, several studies were reviewed about pSS, by searching in google scholar and Pupmed using key-words such as (Primary Sjögren syndrome , clinical and prognostic correlation) .

The results revealed the disease and other correlated subjects such as :

Primary Sjögren SyndromeClinical and Immunologic Disease Patterns in a Cohort of 400 Patients .

Geoepidemiology of Sjögren's syndrome .
Serologic features of primary Sjögren’s syndrome: clinical and prognostic correlation .Which will be discussed in this report . The clinical spectrum of primary Sjögren’s syndrome: beyond exocrine glands .

Results

Primary Sjögren’s syndrome (pSS) is a mild indolent chronic disease mainly characterized by mucosal dryness in the majority of cases, a consistent subgroup of patients display extra-glandular manifestations. Virtually any organs and systems can be affected Up to 40% of pSS patients develop extraglandular disease, but only 5–10% of pSS patients suffer from severe extraglandular manifestations. In particular, palpable purpura, hypocomplementemia, cryoglobulinemia and non-Hodgkin’s lymphoma are associated with increased mortality After a mean follow-up of 91 months, 127 (9.8%) patients developed 133 cancers. The most frequent type of cancer was B-cell lymphoma (including 27 MALT and 19 non-MALT B-cell lymphomas). Systemic activity at diagnosis of primary SjS correlated with the risk of hematological neoplasia and cryoglobulins with a high risk of either B-cell or non-B-cell lymphoma subtypes Neurological complications occur in about 20 p. 100 of primary Sjögren's syndrome patients. It most frequently involves the peripheral nervous system, predominantly sensorimotor and sensory polyneuropathy. Sensory neuronopathy and trigeminal nerve involvement are less frequent but quite suggestive of primary Sjögren's syndrome. Among central nervous system involvements, focal or multifocal lesions of the brain or the spinal cord are the most frequent .

Discussion

Sjögren syndrome primarily affects women, with a female-male ratio of 9:1, and may occur in patients of all ages but typically has its onset in the fourth to sixth decades of life.[3]

Central to the pathophysiology of SS is chronic immune system stimulation. The processes that underlie the humoral and cellular autoimmune reactions observed in patients with SS are not known, but both B and T lymphocytes are involved.

B-cell hyperreactivitiy is expressed to hypergammaglobulinemia and circulating autoantibodies. Organ-specific autoantibodies include antibodies to cellular antigens of salivary ducts, the thyroid gland, the gastric mucosa, erythrocytes, the pancreas, the prostate, and nerve cells. Non–organ-specific autoantibodies are found in approximately 60% of patients with SS. These autoantibodies include rheumatoid factors, antinuclear antibodies, and antibodies to the small RNA-protein complexes Ro/SS-A and La/SS-B. These autoantibodies may contribute to tissue dysfunction before inflammation is evident .

Clinical symptoms include fatigue debilitating fatigue occurs in approximately 50% of patients with primary SS Joint disease in primary SS is typically an intermittent polyarticular arthropathy primarily affecting small joints Dry skin, another exocrine manifestation of SS, was found to affect 55% of SS patients in a recent study. More than 10% of SS patients reported a skin rash.

Pulmonary involvement is seldom clinically significant in patients with SS. Cough is often the main respiratory symptom and is usually a symptom of xerotrachea. Other potential pulmonary complications include lymphocytic alveolitis, lymphocytic interstitial pneumonitis and fibrosis may have involvement of their entire gastrointestinal tract. Malabsorption due to lymphocytic infiltrates of the intestine rarely occurs in patients with SS, and esophageal dysmotility has been reported in 36% to 90% of patients may have tubulointerstitial involvement of the kidneys affecting the tubules (eg, distal renal tubular acidosis, impaired concentrating ability, hypercalcinuria, or proximal tubule defects).

One of the most common significant systemic manifestations of SS is neurologic disease, which can involve the cranial and peripheral nerves and, infrequently, the central nervous system. Peripheral neuropathy, primarily sensory, was found in 22%

Treatment of SS is mainly symptomatic and is directed toward recognizing and treating complications of the disease early.

About Prognostic correlation with manifestation most patient of SS remain healthy except in some rare complication like cancer (lymphoma) or renal failuar and respiratory fibrosis.[4]

Conclusion

SS carries a generally good prognosis; however, the prognosis and mortality are more closely related to the presence of other autoimmune diseases and the potential complications.

In systemic involvement, Attention should be paid to identification of the pathogeny of the primary disease to achieve early identification, diagnosis and treatment. In pSS patients, the risk of developing lymphoma is 15–20-times higher than in people without pSS .

References

1. Mavragani, C., & Moutsopoulos, H. (2020). The geoepidemiology of Sjögren's syndrome. Retrieved 11 March 2020, from **.**

2. Primary Sjögren Syndrome: Clinical and Immunologic Disease... : Medicine. h 2020, from https://journals.lww.com/mdjournal/Fulltext/2002/07000/Primary_Sj_gren_Syndrome__Clinical_and_Immunologic.3.aspx?fbclid=IwAR2fPslvBmtBOjYJb7SEfrdB8OnOnoGYtxZFlOaPi7Cj8W76nE80NWrHlJA (2020). Retrieved 11 Marc .

3. Zeron, P. B., Retamozo, S., Bové, A., Kostov, B. A., Sisó, A., & Ramos-Casals, M. (2013). Diagnosis of Liver Involvement in Primary Sjögren Syndrome. Journal of clinical and translational hepatology, 1(2), 94–102. https://doi.org/10.14218/JCTH.2013.00011 .

4. Kassan, S., & Moutsopoulos, H. (2020). Clinical Manifestations and Early Diagnosis of Sjögren Syndrome. Retrieved 11 March 2020, from .