Case Report

Sjögren’s syndrome—literature review and clinical case presentation

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Abstract:
Sjögren’s syndrome (SS) is a systemic disorder with clinical presentation of dry eyes, dry mouth and autoimmune disorder. Recently the number of patients with SS is increasing. In middle age female patients, having symptoms of dry mouth and salivary glands enlargement, SS should always be suspected. We present a clinical case report of a patient with SS. Based on the therapy and long term follow-up, we may conclude that via appropriate treatment significant improvement could be achieved.

Keywords: Sjögren’s syndrome, sicca syndrome, dry mouth, dry eyes.

Introduction:
Sjögren’s syndrome (SS) is a fairly common chronic, autoimmune, multisystemic disorder mainly affecting the lacrimal and salivary exocrine glands [1,2]. A lymphocytic infiltrate is accumulated in the gland, causing destruction of the tissue and significant reduction or even termination of the secretory activity [3]. Based on histopathological analysis, lymphoid infiltrate in SS could be described as reactive, malignant or atypical hyperplasia [4]. The impaired function of the exocrine glands is followed by persistent dry mouth (xerostomia) and dry eyes (keratoconjunctivitis sicca). The prevalence of SS ranges from 0.2 to 1.4% of the population and is more commonly observed in women [5]. The highest incidence is during the middle age. The etiology of SS remains uncertain. The etiopathogenesis is probably associated with intrinsic factors (genetic predisposition) and extrinsic factors (wide range of exogenous agents), contributing to the disease development.

SS can be either primary when the clinical symptoms are limited to the exocrine glands, or secondary to another connective tissue disease including systemic lupus erythematosus, rheumatoid arthritis, systemic sclerosis, etc. [6]. In 1993 the Preliminary European Classification criteria for SS have been proposed and broadly used in the clinical practice for classification and diagnosis of the disease [7]. These principles were thoroughly revised in 2002 and the current version – American-European Consensus Group (AECG) criteria is accepted as a standard reference for SS [8]. According to the criteria there are six main symptoms associated with SS and the presence of any four of them is considered enough to establish the diagnosis. The six items are as follows: 1. Ocular symptoms; 2. Oral symptoms; 3. Ocular sings (Schirmer-I test, Rose Bengal score); 4. Evaluation of salivary gland function by sialography, scintigraphy and unstimulated salivary flow; 5. Histopathological examination of the minor salivary glands confirming lymphocytic infiltration; 6. Autoantibodies such as antinuclear antibodies (ANA), anti-Sjögren A syndrome (anti-SS-A / Ro) antibodies, Sjögren B syndrome (anti-SS-B / La), rheumatoid factor (RF). There is also exclusion criteria added, including hepatitis C infection, acquired immunodeficiency syndrome (AIDS), radiotherapy in the head and neck region, pre-existing lymphoma, graft-versus-host disease, sarcoidosis, anticholinergic drugs.

The treatment of SS is mainly empirical and symptomatic. For the ocular symptoms, artificial tears and parasympathomimetics are usually prescribed. Treatment of oral symptoms is provided by the application of artificial saliva, sugarless chewing gums and mucolytic drugs. Non-steroidal anti-inflammatory drugs (NSAID) are prescribed to relief salivary glands edema. Oral corticosteroids and immunosuppressants are given for the treatment of the systemic symptoms.

The purpose of this article is to report a case of a patient suffering from SS; to discuss the clinical presentation, paraclinical tests, differential diagnosis and treatment in the context of the current literature.

Case Report:
A 62-year-old female patient is presented with symmetric enlargement of both parotid glands. Periods of moderate pain and tension in the parotid region were reported, mainly prior and during food intake. The chief complaints are associated with severe dry mouth (xerostomia) and dry eyes (xerophthalmia), itching, photophobia and significantly reduced to absent tearing when crying. Multiple dental caries have occurred within the past 10-12 months as well as occasional oro-mucosal infections, without any particular treatment performed. Permanent moderate edema of the limbs, pain in the lumbar region and hip joint pain, history of angular
Chelitis, infections of the urogenital tract and family history of rheumatoid arthritis were reported. The presence of dry mouth and dry eyes necessitated frequent ingestion of fluids, especially during mastication, and application of tear substitutes. During the physical examination swelling of both parotid glands together with bilateral congestion, dry mouth and viscous saliva was detected. Unstimulated whole salivary flow was decreased with less than 1.5 ml in 15 min. Histopathological examination of a minor salivary gland obtained through normal-appearing mucosa showed signs of sialoadenitis with lymphocytic infiltrate (more than one focus per 4 mm2).

Consultation with ophthalmologist has been performed to confirm the diagnosis. Schirmer test on both eyes was conducted with 3 mm for the left eye wetting of the test strip after 5 min and 4 mm for the right eye. In patients older than 40 years, 10-15 mm is considered normal. Paraclinical blood test demonstrated elevated anti-Ro antibodies (anti-SSA Ro >200) and anti-La antibodies (anti-SSb La >200). Erythrocyte sedimentation rate was slightly increased (27 mm/h).

Treatment with Resochin, Imuran and Medrol has been conducted for nearly 3 months. Resochin was withdrawn because of hypersensitivity and Imuran didn’t show any therapeutic effect. After comprehensive clinical examination and paraclinical tests the therapy was replaced. Methotrexate 15 mg once a week, Medrol 1 tablet every day and Folic acid were prescribed. Folic acid was avoided at the day of Methotrexate intake. Seven weeks later the ocular and mouth symptoms were significantly improved. Regular paraclinical tests and consultations with rheumatologist, endocrinologist and neurologist are strongly recommended.

Discussion:

SS is a pathologic condition usually presented with so called sicca syndrome: dry eyes and dry mouth, as extraglandular manifestation is also quite frequently observed. Impaired function and enlargement of the glands usually occur in a combination with vasculitis, pulmonal or renal disorders, as well as fatigue and arthralgia in nearly 20 to 30% of the cases [1,9].

Wide range of causes leading to the development of sicca syndrome is well known, as they can be divided to glandular and nonglandular (or extraglandular). In the nonglandular causes, neurologic and psychological alterations, dehydration, oral sensory dysfunction, mouth breathing, and idiopathic predisposing factors are included. Of the glandular causes, immunodeficiency syndrome, diabetes, sarcoidosis, end stage renal disease, radiotherapy, trauma and tumor of the glands are found to be the most significant [10]. Xerostomia and keratoconjunctivitis sicca are symptoms that might be observed in patients taking tricyclic antidepressants, beta blockers, antihistamines or diuretics. These facts should be considered in the anamnesis, and getting as much information about the general condition of the patient as possible is mandatory. Medical history should be thoroughly revised in order to establish the correct diagnose and to choose the most appropriate treatment.

During the clinical examination of the patients, especially women in their middle age with symptoms of dry mouth and salivary glands enlargement, SS should always be suspected. Further interrogation will confirm the presence of additional symptoms, particularly keratoconjunctivitis sicca and arthritic complaints. In the present case the diagnosis of primary SS is established according to the revised international classification criteria described by Vitali et al. [8]. Recurrent chronic parotitis with at least two inflammatory episodes and pain with varying intensity, separated by remissions is indicative for SS. The patient in the present case reported use of pain-killers and NSAIDs with transient effect. Treatment of SS remains controversial. Probably conservative treatment together with regular precise clinical examinations and paraclinical tests are of primary importance for the proper control over the symptoms.

Conclusion:

SS is an autoimmune systemic disorder, clinically presented with xerostomia, xerophthalmia and autoimmune disorder (rheumatic arthritis). Although, it is typical progressive pathology, an appropriate conservative treatment together with long-term follow up, clinical examinations and laboratory tests, proper control over the symptoms could be achieved.

References:
