



PIC QUESTION OF THE WEEK: 7/26/10

Q: Can you describe some of the general characteristics of Sjogren's syndrome?

A: Sjogren's syndrome (also known as keratoconjunctivitis sicca, xerophthalmia-xerostomia, and sicca syndrome) is an autoimmune disease whose key manifestations are *dry mouth* and *dry eyes*. Symptoms result from infiltration of lymphocytes into lacrimal and salivary tissue and the subsequent destruction of these exocrine glands. *Primary* Sjogren's syndrome generally involves only dryness of the eyes and mouth, but may be associated with systemic complications such as Raynaud's phenomenon, low-grade fever, parotid gland enlargement, arthralgia, and easy fatigability. Interestingly, lymphoma has been documented in a significant number of patients with primary Sjogren's syndrome. *Secondary* Sjogren's has been reported in approximately 30% of patients suffering from autoimmune *rheumatic diseases*, most commonly rheumatoid arthritis and systemic lupus erythematosus. It has also been observed in patients with scleroderma, Hashimoto thyroiditis, primary biliary cirrhosis, and chronic active hepatitis. The syndrome primarily affects women (90% of patients) between the ages of 40 and 60 years; however, it can occur at any age. Systemic symptoms are rare in patients whose condition is secondary to rheumatoid arthritis. Diagnostic criteria include evaluation of oral and ocular signs and symptoms, performance of a Shirmer test (extent of tear flow), presence of auto-antibodies to Ro/SS-A or La/SS-B, and the results of a minor salivary gland biopsy. The treatment of Sjogren's syndrome is centered on alleviating ocular symptoms and dry mouth. As the disease progresses, response may be less evident. Frequent application of artificial tear solutions (Comfort Tears, Tearisol, etc.) and ointments (Lacri-Lube NP, Refresh PM, etc.) relieve symptoms and reduce further ocular damage. Patients should be encouraged to sip water frequently. The use of lozenges, sugar-free chewing gums, and saliva substitutes (Moi-Stir, etc.) are useful in relieving the symptoms of dry mouth. Oral administration of pilocarpine (Salagen; 5 mg four times daily) and the acetylcholine derivative cevimeline (Evoxac; 30 mg three times daily) may also be helpful in the management of xerostomia. Rituximab and other immunosuppressive agents may be beneficial for some of the systemic manifestations of primary Sjogren's syndrome. Anticholinergic compounds (especially atropine derivatives) and decongestants can decrease salivary secretions and should not be administered. Dental hygiene programs and frequent evaluation by an ophthalmologist are essential components in the management of patients with Sjogren's syndrome. Treatment of the oral and ophthalmic symptoms of this disorder has no direct effect on progression of the rheumatic diseases or other systemic complications that frequently accompany the syndrome.

References

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