Stay Alert for Atypical Sjögren’s Signs, Symptoms

ARTICLES BY HEIDI SPELE
Senior Writer

FORT LAUDERDALE, Fla. — Sjögren’s syndrome is the second most common autoimmune disorder that affects the musculoskeletal system, and yet the average time to diagnosis is 6 years, said Yvonne Sherrer, M.D., said at a meeting sponsored by the Sjögren’s Syndrome Foundation.

Until more reliable biomarkers for the syndrome are identified, diagnosing the disease may require physicians to have a higher level of suspicion for some of the less common manifestations of Sjögren’s, said Dr. Sherrer, medical director and director of clinical research at the Centre for Rheumatology, Immunology, and Arthritis in Fort Lauderdale.

Although causes of the Sjögren’s is still unknown, researchers suspect that a combination of genetic, environmental, and hormonal factors contribute to predisposition for the disease. Indeed, for every male with the syndrome, an estimated nine women are affected, underscoring the relevance of hormonal influences.

Inflammation of the exocrine glands, the common denominator of Sjögren’s syndrome, most obviously affects the eyes, mouth, and vagina, Dr. Sherrer said.

Typically, Sjögren’s occurs in the context of a primary autoimmune disorder, such as lupus, rheumatoid arthritis, or scleroderma.

The atypical patients with primary Sjögren’s “are our most challenging patients, because they don’t have an accompanying autoimmune disease.” Dr. Sherrer said.

These patients generally present with neuropathy, accelerated dental caries, salivary gland swelling, joint pain without overt arthritis or myositis, and connective tissues.

The current international criteria for diagnosing Sjögren’s require that patients demonstrate some objective evidence of autoimmunity, in addition to other symptoms. (See box.)

However, the following less typical symptoms may also warrant suspecting Sjögren’s syndrome:

- **Pulmonary.** Lung involvement and coronary involvement are rare but can develop due to dryness of bronchial tubes. Other potential manifestations include bronchitis, bronchiolitis obliterans-organized pneumonia, and interstitial fibrosis.
- **Neurologic.** Neuropathies tend to be less symmetrical in Sjögren’s patients, compared with other conditions. Central nervous system disorders might manifest as changes in cognitive function or as seizures.
- **Renal.** Renal involvement is rare, but when it occurs, it is usually interstitial tube involvement and is more likely to be chronic.

**Recommended Diagnostic Criteria**

The diagnosis of primary Sjögren’s syndrome requires that patients meet at least four of the following six criteria:

1. **The patient must have at least one of three ocular symptoms:**
   - Dry eyes for less than 3 months.
   - Need to use artificial tears more than three times daily.
   - Sensation of a foreign body in the eye.

2. **The patient must have at least three oral symptoms:**
   - Persistent dry mouth for more than 3 months.
   - Swollen salivary glands.
   - Need to add extra liquid to the mouth in order to swallow.

3. **The patient must have at least one of two ocular signs:**
   - Positive anti-SSA or anti-SSB tests.
   - Positive vital dye staining.

For patients who don’t respond to over-the-counter products, prescription humectants such as Carmol 20 and Carmol 40 may help.

He also advises patients to use humidifiers and fragrance-free laundry detergents to reduce irritation of dry skin.

In addition to dry skin, Sjögren’s patients may develop red or purple palpable spots related to small vessel disease. Urticarial vasculitis, which affects some patients, is characterized by hives that last hours to days. When these lesions are biopsied, they show inflammation in the blood vessels.

**Skin Care: Limit Washing, Study Product Labels**

FORT LAUDERDALE, Fla. — Skin manifestations of Sjögren’s syndrome may not be unique to the disease, but they require specific interventions and close monitoring, Darren L. Casey, M.D., said at a meeting sponsored by the Sjögren’s Syndrome Foundation.

Sjögren’s syndrome affects the body’s moisture-producing glands, resulting in chronically dry, itchy, and scaly skin.

Remind patients that they should try to maintain a protective barrier on the skin. “When we wash too aggressively, we get rid of that barrier,” said Dr. Casey, a dermatologist in private practice in Atlanta.

Dr. Casey suggests that patients limit washing to 20 minutes and recommend using Cetaphil, Dove, or Oil of Olay gentle skin cleansers.

Use of emollients such as Aquaphor immediately after a bath or shower can help retain moisture if lotions and creams do not provide relief.

**Manage Dry Eyes Multiple Ways**

FORT LAUDERDALE, Fla. — Optimal management of dry eyes associated with Sjögren’s syndrome is likely to involve just one solution.

Artificial tears come in assorted formulas, and patients who complain of burning and irritation for patients with occluded tear ducts, noted Ashley Behrens, M.D., of Johns Hopkins University in Baltimore.

Atypical patients with primary Sjögren’s may be more convenient to help keep liquid in the eye, Dr. Trattler said. Refrigeration is important in the treatment of external eye disease.

Patients can best manage their eye conditions by recognizing that a single product isn’t likely to meet their various needs, William B. Trattler, M.D., said at a meeting sponsored by the Sjögren’s Syndrome Foundation.

Patients who complain of burning and irritation from artificial tears should try a preservative-free formula, said Dr. Trattler, an ophthalmologist in private practice in Miami, with a special interest in external eye disease.

Preservative-free drops are typically sold as single-use vials, but a tear can be used in the morning and evening on two consecutive days if kept upright in the refrigerator, Dr. Trattler said. Refrigeration is important to inhibit the growth of bacteria, and cold drops can be more comfortable to use. However, drops with preservatives may be more convenient to keep in a pocket or purse or to carry when traveling.

Natural tears and some artificial products have an oily component to help keep liquid in the eye, Dr. Trattler said. Patients should be encouraged to select drops that meet their needs in various settings. Drops needed when working outside will be different than those needed when viewing a computer screen indoors.

If inflammation is present, patients can try cyclosporine ophthalmic emulsion (Restasis). Approved by the Food and Drug Administration in 2003, Restasis increases tear production and has anti-inflammatory properties, making it a good choice for patients with occluded tear ducts, noted Ashley Behrens, M.D., of Johns Hopkins University in Baltimore.

However, patients should be cautioned when using Restasis, since the effects of overdose of the drug are unknown. In the study that served as the basis for the FDA approval, the drops were used twice daily, but “using them more frequently won’t necessarily add benefits,” Dr. Behrens said.

Both ophthalmologists, neither of whom report an economic interest in any eye care product, recommended that Sjögren’s patients use over-the-counter lubricant eye gels for periods of sleep. Warm compresses can provide relief, and specialized moisturizing goggles are available. In addition, patients should be advised to turn off ceiling fans at night.

In treatment-resistant cases, Sjögren’s patients can have their tear ducts blocked with punctal plugs. The ducts drain the tears into the nose, and plugging them helps retain fluid in the eye. Cauterization of the tear ducts has the same result, but while plugs can be removed, cauterization permanently closes the ducts.

For patients who develop blepharitis, inflammation of the eyelid margins, Dr. Trattler recommends using a mild shampoo and warm compresses. The inflammation occurs when the oil that is a component of normal tears crystallizes on the lids, he explained.

For patients who don’t respond to over-the-counter products, prescription humectants such as Carmol 20 and Carmol 40 may help.

He also advises patients to use humidifiers and fragrance-free laundry detergents to reduce irritation of dry skin.

In addition to dry skin, Sjögren’s patients may develop red or purple palpable spots related to small vessel disease. Urticarial vasculitis, which affects some patients, is characterized by hives that last hours to days. When these lesions are biopsied, they show inflammation in the blood vessels.

Medium-sized vessel disease typically manifests as nodules on the hands and feet or as live-dot retinopathy, characterized by a whorl pattern of pigmentation brought on by constricted blood flow. Medium-sized vessel disease should be treated with a red flag to investigate central nervous system or severe organ involvement.