## Sjögren's Syndrome Is Overlooked, Undertreated

BY DENISE NAPOLI

Assistant Editor

ith many female baby boomers reaching the age when Sjögren's syndrome presents, rheumatologists must sharpen their skills to diagnose and manage this second most common autoimmune disease in the United States, said several experts.

"It is not a benign disease, which is a perception that many physicians have," said Dr. Frederick B. Vivino, director of the Sjögren's syndrome center at Penn Presbyterian Medical Center, Philadelphia. "It has significant morbidity, and even mortality, when people develop internal organ manifestations or complications like lymphomas."

Dr. Arthur Weinstein, director of the section of rheumatology at the Washington



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DR. VIVINO

Hospital Center, said that among internists and family physicians, Sjögren's syndrome (SS) is "grossly underrecognized."

Whether or not physicians follow the diagnostic criteria for the disease is a matter of debate. "My colleagues in rheumatology don't do as good a job diagnosing and treating Sjögren's as they do rheumatoid arthritis," said Dr. Vivino. "Most physicians, including rheumatologists, aren't really aware of the current diagnostic criteria for the disease, though they were published in 2002. Or if they are aware, they don't follow them."

Dr. Weinstein is aware of the criteria (Ann. Rheum. Dis. 2002;61:554-8), but said he finds them better suited to classification than to diagnosis. Some criteria are major, he said, and indicative of disease, whereas others are minor. For instance, a biopsy plus ocular and oral symptoms, or autoantibodies (anti-SSA and/or anti-SSB) with the dryness symptoms, are often sufficient to make a diagnosis, he said.

Ultrasound of the salivary glands has recently been looked at as a diagnosis tool (Rheumatology News, February 2008, p. 18), but Dr. Vivino, chairman of the medical and scientific advisory board for the Sjögren's Syndrome Foundation, said, "We haven't used it, because it's not included in the current criteria."

For now, misdiagnosis of SS is common. Dr. Weinstein said, "Many of the SS patients are misdiagnosed as having RA because they have rheumatoid factor, or lupus, or because of a positive [antinuclear antibody test]. . . . Early on, it could look like those, but ultimately [these patients] have different problems. So it takes an awareness to ask the right questions."

Dr. Vivino said many women regard dryness, the primary symptom in SS, as a part of menopause, which is the approximate age of disease onset. On average, the mean time between symptom onset and diagnosis is 7 years, he added.

Mouth burning or oral ulcers from the dryness can make eating difficult, leading to weight loss. Many patients develop dental caries, yeast infections of the mouth, or bacterial infections of the salivary glands.

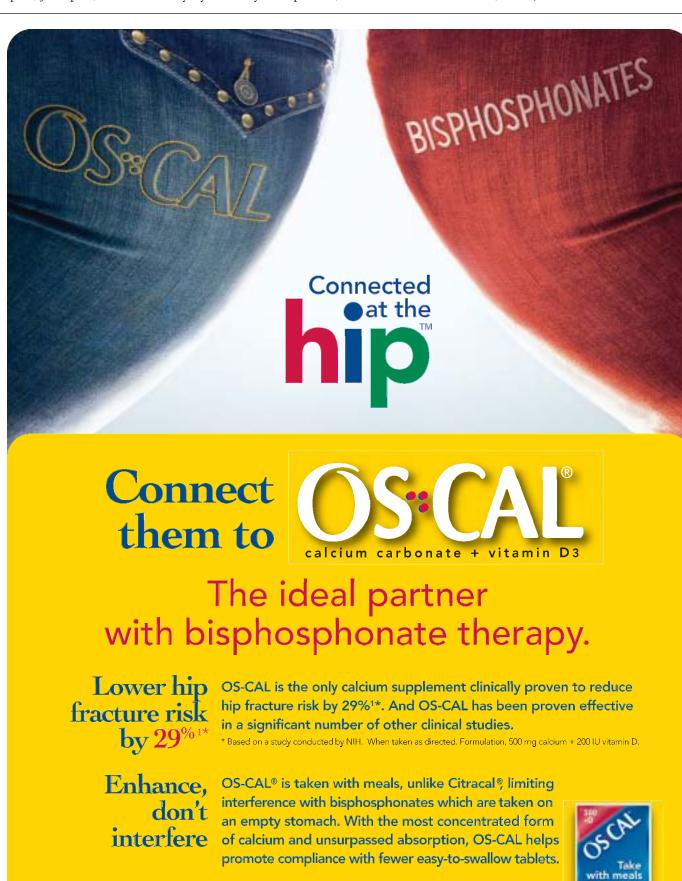
Ocular dryness can cause infections. "In some cases, the cornea can perforate," said Dr. Vivino. Vaginal dryness also occurs.

More seriously, SS is tied to fatigue, muscle pain, joint pain, liver and kidney dysfunction, and non-Hodgkin's lymphoma.

The drug of choice for SS is hydroxy-chloroquine (Plaquenil), which came on the market in 1955. Dr. Weinstein said one new therapy that shows promise is B-cell-targeted rituximab. "There's this [fascinating] link between Sjögren's and the development of B-cell lymphomas," he said; one study put the incidence of lymphoma in this cohort at more than 40 times higher than in the general population. In a study of 16 patients, rituximab was tied to

remission in SS-associated lymphomas, but did little to lessen symptoms of dryness (Ann. Rheum. Dis. 2007;66:351-7).

Another investigational B-cell-targeted drug, epratuzumab, showed efficacy in a 16 patient-, open-label, phase I/II study. More than half of patients had a greater than 20% improvement in the Schirmer I test of tear production, unstimulated whole salivary flow, fatigue, erythrocyte sedimentation rate, and IgG levels (Arthritis Res. Ther. 2006;8:R129).



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