**Case Report**

Otitis externa sicca/fibrotising external otitis (FEO) as a complication of Sjögren’s syndrome

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**ABSTRACT**

Sjögren’s syndrome (SS) is a condition characterized by sicca symptoms and by autoimmune features. We describe two SS patients with otitis externa fibroticans/sicca. One of these 2 patients developed a lesion of the tympanic membrane making it necessary to perform a tympanotomy and meatoplasty. Our findings suggest firstly that the epithelial cell-mediated secretion of lamellar bodies and the production of the permeability barrier are defective in SS. Secondly, local moisturing and/or topical corticosteroid treatment in SS patients with sicca symptoms in the auditory canal could help to avoid reconstructive surgical treatment.

**Introduction**

Sjögren’s syndrome (SS) is an autoimmune exocrinopathology occurring mainly in elderly women. According to the classification criteria (1, 2) it is characterized by: (i) dry eyes; (ii) dry mouth; (iii) objective evidence of dry eyes; (iv) objective evidence of dry mouth; (v) autoantibodies (SS-A/Ro, SS-B/La, rheumatoid factor and/or anti-nuclear antibodies); and (vi) a focal sialadenitis of the labial salivary glands (3). If at least 4 of these criteria are fulfilled, the diagnosis of primary Sjögren’s syndrome can be made. SS is thus characterized by sicca symptoms/findings and autoimmune features. In addition, at least lymphoma, sarcoidosis, HIV (and HCV) infection and graft-versus-host reaction should be excluded.

The most common extra-glandular symptoms are fatigue, arthralgias/myalgias and Raynaud’s phenomenon. Visceral manifestations include chronic atrophic gastritis, renal tubular acidosis of the distal type (type 1) and autoimmune thyroiditis.

Although the sicca symptoms are usually relatively mild, they can also be severe. For historical and practical reasons dry eyes (keratoconjunctivitis sicca) and dry mouth (sialopenia/xerostomia) have been emphasized, but any mucosal surface may be affected. In addition, due to involvement of the sweat and/or sebaceous glands, the skin is often dry, a condition referred to as xerosis. This makes the skin sensitive so that it may become itchy and sunburn develops easily. In addition to these sicca symptoms, the patient may suffer from skin changes caused by vasculitis, typical features being a vasculitic purpura in the legs and urticaria-like, but more long-lasting skin lesions (4).

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Patient no. 1

The diagnosis of this patient was based on the typical symptoms of dry eyes and dry mouth. The patient felt that her eyes had been dry and gritty for more than 3 months in spite of the frequent application of artificial tears (Tears Naturales; Alcon, Couvreur, Belgium); she did not have enlarged parotid glands, but she felt that her mouth was dry so that she had to consume liquids when eating dry food. The Schirmer I test demonstrated diminished tear fluid flow (1 mm/5 min on the right side and 0 mm/5 min on the left side) and lesions in the anterior conjunctival and corneal surfaces of the eye. Rose Bengal score values were 8 and 9, calculated according to van Bijsterveld (5). Finally, she had a slightly increased titer of rheumatoid factor (18 IU/ml, reference values 0-14 IU/ml), but at the time of the study, her SS-A/Ro and SS-B/La antibodies and anti-nuclear antibodies were negative. The focus score of her labial salivary glands was 2.9 calculated according to Greenspan et al. (3).

She did not have lymphoma, sarcoidosis, HIV or HCV infection or graft-versus-host reaction. Because she did not have any underlying autoimmune disorder, she fulfilled the EU criteria for primary SS (1). All of the above mentioned tests were performed with careful attention to the recommendations published in the Manual of Methods and Procedures (6).

Her first disease symptoms appeared at the age of 45 years in 1982, 14 years before the present examination and report. The first symptoms she noticed were gritty and irritated eyes. Eight years later dry mouth symptoms developed. These symptoms had been very prominent so that during the year preceding the present examination (1995) she visited the Institute of Dentistry, University of Helsinki, for 52 times for treatment of com-

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...lications of sialopenia/xerostomia and e.g. a dental bridge was constructed and dentures with moisture reservoirs were made for her. Her dry mouth was frequently complicated by secondary Candida superinfections, which necessitated repeated treatments with fluconazole capsules, amfotericine B resoriblets and miconazole gel. She also often developed an associated angular cheilitis. She suffered from vaginitis sicca, which was complicated by labial synchiae. This was noticed when samples were going to be taken for a routine cervical cytological Papanicolaou smear. These synchiae were initially treated with a dilatation in local anesthesia. She had been on hormone replacement therapy since her menopause and local estrogen treatment was initiated. She had dry skin and was frequently using moisturizing skin emulsions and occasionally local corticosteroid ointments. She was also relatively tired, but did not display any visceral organ manifestations.

In addition to the symptoms described above, both ears were repeatedly deafened and she heard a continuous hum. She did not have aches or pains in her ears. An otitis externa sicca was diagnosed and in spite of moisturising ear drops and corticosteroids recommended for temporary use, her condition progressed during many years leading to fibrotising external otitis and hearing loss. At that time she was referred to a specialist, who as a result of these bothersome and progressive symptoms recommended a tympanotomy with metaplasty, which operations were performed with good short-term results.

Patient no. 2
Case no. 2 was 55-year-old male patient. He had had an auditory injury in his right ear when he was serving in the army. He engaged in sports hobbies such as hunting and shooting. The hearing of the injured right ear had been worse than in the healthy left ear, which was consistent with his age. However, from the spring of 1999 the hearing in his left ear became worse and the patient was sent to a specialist for further examinations. The otorhinolaryngological status was otherwise normal, but the ears were dry on both sides. The left ear canal ended in a blind-sac, making the tympanic membrane thick and blunted. This finding was regarded as the cause of his conductive hearing loss. On the right side the ear canal was narrow, but the tympanic membrane appeared normal. In addition, Rinne’s test was negative on the left side. Furthermore, the patient had dry eyes and dry mouth. His parotid and submandibular glands had been enlarged several times. The patient was sent to have a labial salivary gland biopsy which showed focal sialadenitis with a focus score of approximately 2.2. On this basis primary SS was diagnosed. As a complication, based on the symptoms and clinical findings, bilateral otitis externa fibroticans was diagnosed, being more severe on the left side. The patient is now undergoing medical examination and the plan is to treat him surgically after local and systemic therapy for SS have been initiated.

Discussion
Xerosis in Sjogren’s syndrome has been attributed to involvement of the sweat and/or sebaceous glands, which have been considered to be the major sources of lubrication of the skin (4). More recently, it has become clear that a mixture of ceramides, cholesterol and free fatty acids produced by the epithelial cells forms the permeability barrier that permits terrestrial life (7, 8). The lateral cells form the permeability barrier rather than by involvement of the sweat and/or sebaceous glands. Sensory nerve hearing impairment has been described in SS (13). We have described here a case of otitis externa sicca progressing to fibrotising external otitis and hearing loss, to add a new otorhinolaryngological dimension to this syndrome.

In summary, Sjogren’s syndrome is an autoimmune exocrinopathy, where most of the symptoms cluster either as autoimmune manifestations or as a dysfunction of the exocrine glands/secretion. Our patient was characterized by only mild autoimmune manifestations, but by severe sicca symptoms. In her case the specialized skin surface of the external ear and ear channel were involved in such a severe way, that finally an otitis externa sicca and a complicating fibrotising external otitis developed with subjective symptoms and hearing loss as a result. The short-term results of meatoplasty and tympanotomy seem promising, at least in our patient, and this mode of treatment can be recommended in advanced cases. Perhaps more importantly, when SS patients with severe sicca symptoms suffer from affection of the external ears, long-term prophylactic treatment with moisturizing drops and/or topical corticosteroids might be worth a trial. At least other local treatments, such as that of the dry eyes and dry mouth, have gained wide acceptance in SS centers as part of the standard treatment regimen, when local symptoms and signs are evident.

References