Net Letter

Concomitant Darier's disease and Sjögren's syndrome

Sir,

Darier's disease, also known as keratosis follicularis is an autosomal dominant disease characterized by dirty-looking, warty, papular lesions located in the seborrheic areas.[1] Sjögren's syndrome is a systemic autoimmune disease in which the exocrine glands are damaged by immune cells.[2] Herein, we describe a patient with the rare coexistence of Darier's disease and Sjögren's syndrome.

A 48-year-old woman came to our outpatient clinic with gray, itchy papules on her body. The patient said that the papules had appeared 20 years ago. She had been admitted to the rheumatology department 10 months earlier for dry mouth, foreign body sensation in the eyes and arthralgia involving knees and ankles. A diagnosis of Sjögren's syndrome was established and subsequently hydroxychloroquine and systemic steroid therapy were started. There was no history of similar complaints in any family member.

Physical examination revealed widespread, gray, erythematous papular lesions over her body which mainly involved the neck, axillae, inframammary regions and abdomen [Figures 1a and b]. In addition, longitudinal streaks and distal notches were observed on the nails [Figure 1c]. Her oral and genital mucosa were normal. An ophthalmological examination showed mild bilateral blepharitis and corneal punctate epitheliopathy. Laboratory tests showed a rheumatoid factor of 45.3 IU/mL (reference range: 0-15 IU/mL), positive anti-nuclear antibodies (ANA) at 1:320 dilution with a speckled pattern, an anti-Ro antibody level of >240.00 IU/mL (reference range: <5 IU/mL), an anti-La antibody level of 45.10 IU/mL (reference range: <5 IU/mL), and an erythrocyte sedimentation rate (ESR) of 94 mm/h (reference range: 0–30 mm/h). Other tests (complete blood count, urinalysis, serum biochemistry) were within normal limits. Labial salivary gland biopsy revealed mild glandular atrophy and a dilated small duct with a lymphocyte focus score of greater than one (>50/4 mm^2) [Figure 2a]. A punch biopsy obtained from one of the papular lesions on the neck revealed marked hyperkeratosis, parakeratosis, papillomatosis, suprabasal clefts and acantholysis-associated dyskeratosis with corps ronds and grains. A mild superficial perivascular infiltrate of lymphocytes was also noted [Figure 2b]. Based on the clinical and laboratory findings, the patient was diagnosed with sporadic Darier's disease in conjunction with Sjögren's syndrome.

Systemic methotrexate (20 mg/week) was started after rheumatology consultation. At follow-up after 3 months, the symptoms and the skin lesions had significantly improved.

Darier's disease is a rare, autosomal dominantly inherited genodermatosis.[1] The characteristic feature

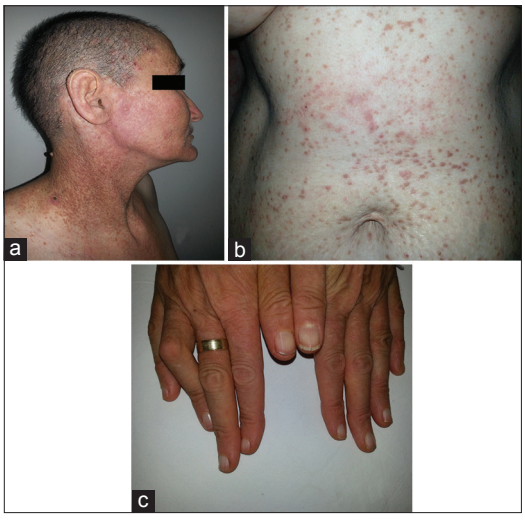

Figure 1: (a) Confluent gray, erythematous papular lesions on the neck, (b) Diffuse, discrete papular lesions on the trunk, (c) Longitudinal streaks and distal notches on the nails

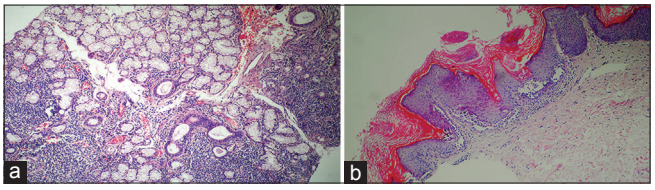

Figure 2: (a) Lymphocytic sialadenitis. (H and E, ×100), (b) Biopsy of a papule showing marked hyperkeratosis, parakeratosis, papillomatosis, suprabasal clefts, and acantholysis-associated dyskeratosis with corps ronds and grains. (H and E, ×40)

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is the presence of firm, hyperkeratotic papules located predominantly in the seborrheic areas but mucous membranes may also be involved. Oral lesions can be seen on the palate, gingiva, buccal mucosa, and tongue. The disease may cause obstruction of the salivary gland channels, leading to xerostomia. Several studies have also reported the obstruction of salivary gland ducts even in patients who did not have any symptoms.[3]

Sjögren's syndrome is an autoimmune connective disorder characterized by xerophthalmia and xerostomia due to lymphocytic infiltration of the lacrimal and salivary glands.[2] Earlier studies detected mitochondrial alterations in cells from these patients, which were related to the action of antimitochondrial autoantibodies. Recently, Perez *et al.*, found that gene loci such as 1p34.2 (D1S3721), 3q13.31-q13.32 (D3S2460) were differentially expressed in labial salivary glands and epithelial cells in Sjögren's syndrome.[4] Various neurological and psychiatric diseases, skin cancers, esophageal carcinoma, polycystic kidney disease, renal and testicular agenesis, bone cysts, and horseshoe kidney have been reported to be associated with Darier's disease;[1] however, Sjögren's syndrome is a rare association. We were able to find only one previously reported case of Darier's disease concomitant with Sjögren's syndrome. Oxholm *et al.*, reported a patient with Darier's disease who presented with dry mouth and foreign body sensation in the eyes. They thought that these symptoms were caused by the primary disease but after further investigation due to persistence of these symptoms, the patient was diagnosed with Sjögren's syndrome.[5]

Darier's disease and Sjögren's syndrome do not usually appear concurrently. As xerophthalmia and xerostomia can be encountered in both conditions, when these symptoms occur in patients with Darier's disease, the possibility of a concomitant Sjögren's syndrome should be kept in mind.

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