Darier's Disease with Involvement of Both Submandibular Glands

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Salivary gland obstruction in association with Darier's disease is described. Histological examination of both submandibular glands revealed squamous metaplasia of the ducts with suprabasal cleft formation and occlusion of the lumen.

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To our knowledge, only one report has previously been published, clearly documenting epithelial Darier-like changes in the salivary gland ducts and causing obstruction and sialoadenitis (1). The present case is reported because both submandibular glands were involved in a patient with Darier's disease.

CASE REPORT
The patient is a 41-year-old man whose mother has skin changes, probably Darier's disease. He has one brother and two sisters, but none of them has any skin changes, nor has any of his three children. The skin changes appeared when he was a teenager. He was seen at our Department for the first time when he was 18 years old. There were greasy brown papules typical of Darier's disease on his chest, abdomen and back. He also had longitudinal streaks in the nails and abnormal deratoglyphic patterns. Histology confirmed the diagnosis of Mb. Darier. Over the years the skin changes have spread, now also involving his face, neck, ears, upper arms and upper legs. No macroscopic mucosal lesions have been noticed.

For several years he has suffered repeated attacks of severe abdominal pain. The diagnosis of acute intermittent porphyria has been suspected, but not confirmed.

In 1970 the left submandibular gland was extirpated because of recurrent episodes of swelling and sialolithiasis. Since 1974 the patient has had recurrent episodes of swelling of the right submandibular gland. Sialography has revealed chronic inflammation with dilated ducts within the gland, as well as multiple strictures. No stones have been found. During the last 2 years the painful swellings have recurred more frequently. The right submandibular gland was therefore also extirpated, in October 1989.

Histopathological examination of the right gland revealed a marked dilation of both the greater interlobular ducts and the intralobular ducts. Around the latter a slight fibrosis and varying degree of lymphocytic infiltration were observed. The reaction around the greater interlobular ducts was more pronounced, with considerable fibrosis and prominent lymphoid tissue with follicles. The ductal epithelium in both types of duct showed a pronounced squamous metaplasia, often with a papillomatous architecture, as well as intra-epithelial cleft formations (Fig. 1) with evidence of acantholysis and dyskeratosis (Fig. 2). The dilated lumina of the principal duct and interlobular ducts were sometimes filled with desquamated epithelium, lymphocytes and inspissated secretions (Fig. 3). The smaller intralobular ducts were, however, lined by normal columnar epithelium. The histologic picture was consistent with changes in the salivary ducts described in Mb. Darier (1). Moreover there was a mild sialoadenitis, probably secondary to the changes in the ducts.

Microscopic re-examination of the previously extirpated left submandibular gland revealed similar changes in the ducts, and in this gland sialolithiasis and more pronounced sialoadenitis were also seen.

COMMENTS
Salivary gland obstruction in association with Darier's disease was first observed in 1966 (2). In a paper on oral involvement in Darier's disease the authors concluded their discussion by stating: «The significance of a fairly definite history of chronic recurrent obstruction of the parotid duct occurring in two of our four cases (and in one of the patients who did not have oral lesions) must provide material

Fig. 1. Interlobular duct showing squamous metaplasia of epithelium with suprabasal cleft formation and detached epithelium desquamating into the lumen. Htx-eosin, × 200.

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Notalgia Paresthetica – Puzzling Posterior Pigmented Pruritic Patch
Report on two cases

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In the following study we discuss two extensively investigated patients with a pruritic pigmented patch on the back.

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In the neurological and dermatological literature two syndromes are related with regard to the body area affected. These are “notalgia paresthetica” (1, 2, 3) and “puzzling posterior pigmented pruritic patch” (4). The former is a sensory neuritis affecting the upper part of the back and corresponding to the 2nd and 6th thoracic dermatomes. It may appear on either side of the spine. Subjectively, the patient usually feels a tickling, creeping or pruritic sensation. A careful examination usually discloses zones of hyperesthesia to pinprick close to either scapula, and encompassing the above-mentioned dermatomes. A hereditary variant has also been described (5). The disease may occur either as a single localized symptom or as part of a migratory sensory neuritis (6). Intense localized paroxysmal itching may also occur in multiple sclerosis (7).