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 dermatoglyphic 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

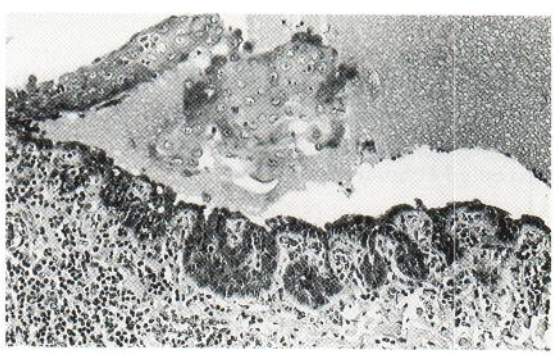

Fig. 1. Interlobular duct showing squamous metaplasia of epithelium with suprabasal cleft formation and detached epithelium desquamating into the lumen. Htx-eosin, × 200.

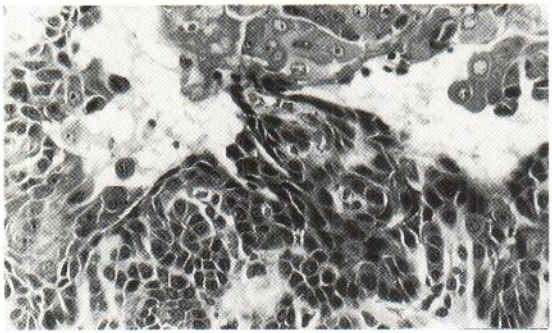

Fig. 2. Detail of duct epithelium with cleft formation and signs of acantholysis and dyskeratosis, Htx-eosin, × 800.

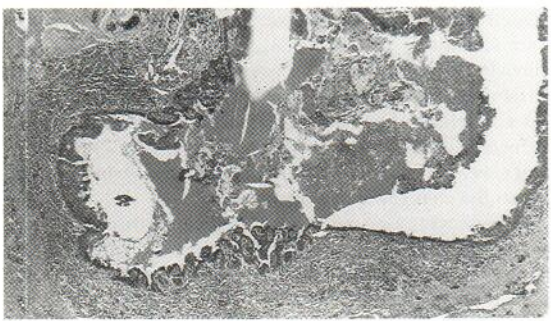

Fig. 3. Low-power view of dilated interlobular duct. Lumen is partly filled with detached epithelium and inspissated secretion products. Htx-eosin, × 40.

for some speculation, but biopsy or further investigation of this aspect was not possibble.«

In 1983, Graham-Brown et al. reported on 2 patients with Darier's disease, in whom clearly documented salivary gland obstruction occurred in association with epithelial changes in the salivary ducts near their oral orifice. These consisted of squamous metaplasia and Darier-like changes (1). Our patient with Mb. Darier had suffered from salivary gland obstruction of both his submandibular glands. Histologic examination of both glands revealed squamous metaplasia, suprabasal cleft formation and occlusion of duct lumina.

This case calls attention to the fact that the dyskeratotic process of keratosis follicularis, Darier's disease, may also involve the ducts of the salivary glands and lead to obstruction.

REFERENCES

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