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ERUPTIVE SYRINGOMAS OF THE NECK

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Abstract

Syringomas are benign adnexal tumors of eccrine origin. Eruptive syringomas clinically present as multiple, skin colored tiny papules that usually develop during a short period of time and are frequently misdiagnosed with other entities. Histopathological examination is essential in the diagnosis and usually reveals numerous, small ducts in the dermis lined with a double row of epithelial cells. Herein, we present an additional case of multiple eruptive syringomas on the neck in a 25 year old female.

Key words: syringoma; papules; neck

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Introduction

Syringomas are benign adnexal tumors of eccrine origin. The name "syringoma" is derived from the Greek word syrinx, which means pipe or tube, and describes the acrosyringium, the intraepidermal portion of eccrine sweat ducts. Syringomas commonly appear as soft, flesh colored single or multiple dermal papules localized on the eyelids in women during puberty. According to Friedman and Butler's classification scheme, 4 principal clinical variants of syringoma are recognized: a localized form, a form associated with trisomy 21, a familial form and a generalized form that includes multiple and eruptive syringoma [1]. In 1987 Jacquet and Darier first described eruptive syringoma, a rare variant of syringoma [2]. Clinically, eruptive syringoma may be confused with acne vulgaris, milia, sebaceous hyperplasia, lichen planus, eruptive xanthoma, hidrocystoma, urticarial pigmentosa, trichoepithelioma and xanthelasma on the face, and granuloma annulare on the trunk, therefore, histopathological examination is essential [3,4]. Herein we present a case of multiple syrinogmas of the neck in a 25 years old female patient.

Case Report

A 25 year old female patient was admitted to the dermatologist's office with the complaint of a rash and itch on the skin on the anterior neck (Fig. 1, left-sided arrows). Patient was otherwise healthy and had no other complaints.

Biopsy was taken from one of the lesions. It showed a well-

circumscribed neoplasm in the upper and middle parts of the dermis surrounded by a fibrous stroma (Fig. 2). The neoplasm was composed of small ducts composed of two-layers of cuboidal epithelium. Some ducts were dilated and some had a short "comma-like" tail (Fig. 2, 3). The diagnosis of "eruptive syringoma" was established. The patient refused to undergo treatment

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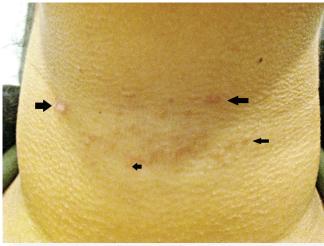


Figure 1. Multiple pink and skin colored papules on the anterior neck. Scar after skin biopsy (right sided arrow).

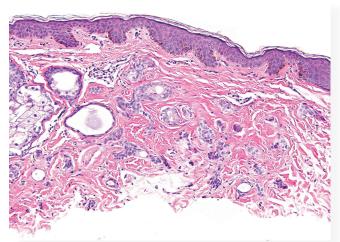


Figure 2. Well-circumscribed eccrine neoplasm in the upper and middle parts of the dermis surrounded by fibrous stroma. Hematoxyllin & Eosin stained sections, ×100.

Discussion

Eruptive syringomas clinically present as multiple, skin colored tiny papules that usually develop during a short period of time. Neck, axillae, pubis and anterior trunk are the most frequent localization. Rare cases with widespread involvement of the body have also been reported [5,6].

The diagnosis is rarely established based on the clinical picture alone, hence, histopathological examination is mandatory. Syringomas are associated with multiple conditions, most frequently with Down syndrome and Nicolau-Balus syndrome (eruptive syringomas, milium cysts and atrophoderma vermiculata) [7].

Histopathologically, eruptive syringoma is characterized by numerous, small ducts in the dermis lined with a double row of epithelial cells. Ductal lumina are filled with an amorphous, periodic acid-Schiff-positive material. Some of the ducts possess small, comma-like tails of epithelial cells, giving them the appearance of tadpoles. Clear cell variant of syringoma and squamous metaplasia were also described [8]. Rarely, especially if located in genital area, syringomas may be located in the deeper areas of the dermis. In such cases differential diagnosis with microcystic adnexal carcinoma is especially difficult.

Absence of follicle differentiation helps to distinguish syringomas from trichoepitheliomas and infundibular-cystic basal cell carcinomas. The differential diagnosis between syringomas and microcystic adnexal carcinoma and reactive eccrine gland metaplasia is more complex and often requires clinico-pathological correlation.

The pathogenesis of eruptive syringoma is unclear. Association with drug intake and variety of inflammatory conditions was described. Guitart et al. propose the term syringomatous dermatitis assuming a hyperplastic response of the eccrine duct to an inflammatory reaction rather than a true adnexal neoplasm [9]. Yoshii et al. reported a case of Syringoma-like eccrine sweat duct proliferation in association with radiation dermatitis [10]. Furthermore, eruptive syringoma may be linked to alopecia areata, prurigo nodularis or lymphocytic inflammatory reactions. Öztürk et al. described a case of post-pubertal eruptive syringoma triggered by anti-epileptic drugs, namely, valproic acid and carbamazepine [11]. In our patient, careful assessment did not reveal any associations.

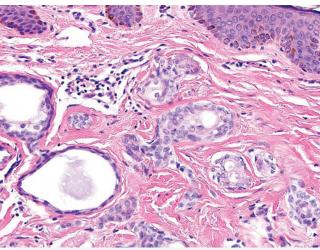


Figure 4. Dilated eccrine ducts lined with two-lavers of cuboidal epithelium and short "comma-like" tails, surrounded by fibrotic stroma. Hematoxyllin & Eosin stained sections, ×400.

Treatment of eruptive syringoma includes cryosurgery, dermabrasion, chemical peeling, electrodessication, curettage, CO2 laser and demonstrate variable cosmetic results [12]. Treatment with oral isotretinoin, topical tretinoin, adapalene was also reported. Sánchez et al. suggest the use of topical atropine to alleviate the pruritus in symptomatic eruptive syringoma [13].

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