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Das, Anupam
Kumar, Piyush

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Photo Vignette

Erythematous papules over the neck: a diagnostic conundrum

Anupam Das¹, Piyush Kumar²

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¹Department of Dermatology, KPC Medical College, Kolkata, India

² Department of Dermatology, Katihar Medical College, Katihar, India

Correspondence:

Piyush Kumar
Email: docpiyush@gmail.com

Abstract

Syringomas are common appendageal neoplasms, manifested as skin-colored papules, most commonly distributed over the periorbital region. We describe a patient with syringoma localized over the neck and upper chest.

Case synopsis

A 25 year-old otherwise healthy gentleman presented with multiple elevated lesions over his neck, present over the preceding 7 years. There were initially a few lesions over the front of the neck, but gradually, the lesions increased in number to attain the present status. Family history was non-contributory. Cutaneous examination revealed numerous 1-2 mm skin-colored flat-topped and oblong papules distributed over the anterior and lateral aspects of the neck. A few lesions were present over the upper chest as well (Figure 1).



Figure 1. Multiple, discrete, skin colored to slightly erythematous, oblong papules on neck.

Histopathological examination showed well-delineated lesions in the upper dermis in a fibrotic stroma. The lesion consists of single-to-double layered epithelial cells with pale eosinophilic cytoplasm, forming nests, cords, or tubules, with a typical comma-like tail (Figure 2). Based on the clinicopathological correlation, a diagnosis of syringomas at an unusual location was established.

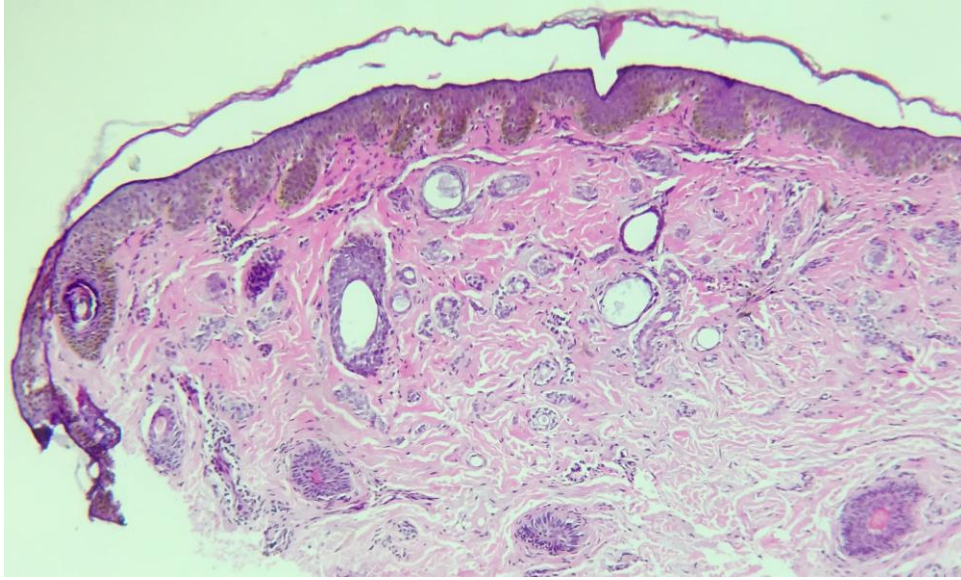


Figure 2. Single-to-double-layered epithelial cells with pale eosinophilic cytoplasm, forming nests, cords, or tubules. The stroma is fibrotic. (H&E x 100).

Discussion

Syringomas are benign adnexal neoplasms with an eccrine sweat duct differentiation. The clinical variants, described by Friedman and Butler in 1987, include local or disseminated syringoma associated with Down syndrome and inherited types [1].

Clinically, it manifests as an insidious onset of bilaterally symmetrical asymptomatic papules, most commonly located in the periorbital region [2]. It is more frequent in females, and is rarely familial [3]. Atypical sites include scalp, penis, buttocks, axillae, groin, and moustache area [4, 5, 6,]. In our case, the lesions were confined to the neck and upper chest. The entities considered in the differential diagnosis were verruca plana, sebaceous hyperplasia, cutaneous sarcoidosis, and appendageal neoplasms. But histology clinched the diagnosis of syringoma.

Histology is characterized by single to double-layered epithelial cells with pale eosinophilic cytoplasm. The hallmark finding is the formation of nests, cords, and tubules, with a typical comma-like tail, referred to as a “tadpole-like” appearance. The lumina are filled with PAS-positive eosinophilic material [7]. On immunohistochemistry, syringomas stain positive for S-100, carcinoembryonic antigen, epithelial membrane antigen, lysozymes, antibodies to the breast cystic fluid protein GCDFP-15 and GCDFP-24, succinic dehydrogenase, phosphorylase, and leucine aminopeptidase [8].

Eruptive syringomas are characterized by the sudden-onset of numerous papules on the neck, trunk, axillae, shoulders, abdomen, and pubic area [9]. Important associations of eruptive syringomas include Nicolau- Balus syndrome, Down syndrome, urticaria pigmentosa, interdermal nevus, basal cell carcinoma (BCC), carcinoid tumor, and epidermal cyst [9, 10].

The etiopathogenesis of syringomas is not clear. According to some authors, these may be the end-result of chronic inflammatory processes of adnexal structures, referred to as “syringomatous dermatitis” [11]. Evidence in support of this hypothesis includes the finding of syringomas in association with lichen planopilaris, prurigo nodularis, and radiation therapy [12, 13]. Another school of thought believes in the role of hormonal factors behind the development of syringomas. This is supported by features like the predilection for females, lesions occurring around puberty, and development of vulvar syringomas during pregnancy [14].

The treatment outcomes are quite frustrating. Options include retinoids (topical and systemic), topical atropine, electrocautery, laser ablation, cryosurgery, and dermabrasion [9, 15].

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