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Eruptive Syringoma in the Elderly: A Case Report

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ABSTRACT Syringomas are benign tumors of adnexal origin, and eruptive syringoma is an extremely rare subtype. In this paper, we present a case of a unusual occurrence of eruptive syringoma in 66-year old woman that includes clinical and pathohistological findings and the review of the clinical picture, diagnosis, and treatment options.

KEY WORDS: eruptive syringoma, diagnosis, clinical presentation, therapy

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INTRODUCTION

Syringomas are benign tumors of adnexal origin. They were initially described by Kaposi in 1872 as lymphangioma tuberosum multiplex (1). Syringomas typically present as multiple soft or firm, skin-colored or yellow papules measuring 1 to 5 mm in diameter (2), that tend to appear on the eyelids in middle-aged women (3).

A classification of syringomas was proposed by Friedman and Butler. According to their clinical features and associations with other medical conditions, syringomas can be classified as one of four principal variants: localized form, familial form, form associated with Down syndrome, and generalized form that encompasses eruptive syringomas (4).

Eruptive syringoma is a rare variant of syringoma that usually appears before or during puberty (5).

Herein we present an unusual case of eruptive syringoma in elderly female patient.

CASE REPORT

A 66-year old woman presented with multiple skin lesions appearing on the face, neck, and axillar and cubital areas in successive crops that developed over a 15-year period. She reported pruritus in the affected areas following perspiration as well as aggravation of lesions after exposure to heat. She was initially treated with local corticosteroid cream, which made lesions less erythematous and less pruritic but still persistent. She denied a familial history of such or similar lesions. The patient's past medical history included hypothyreosis and hypertension, for which she had been treated with lisinopril and levothyroxine.

Physical examination revealed multiple flat-topped papules 1-3 mm in diameter and symmetrically distributed, on the forehead, lower third of the face, neck, and axillary and cubital areas, ranging in color from flesh color to pink. The lesions on the forehead and neck were surrounded by erythematous halos. The lesions were symmetrical in all parts of the body (Figure 1).

The initial clinical diagnosis was flat warts, and skin biopsy of a small papule on the cubital area was obtained. Histological examination of the specimen showed multiple ducts in superficial dermis. The ducts were formed by two layers of epithelial cells and were filled with amorphous eosinophilic material. Some of ducts had elongated tails of epithelial cells, producing the characteristic comma-shaped appearance (Figure 2). Clinical and histological findings were compatible with diagnosis of eruptive syringoma.

An additional patch test was performed using a baseline series of patch test allergens (European Standard Series), which yielded a negative result for all allergens tested.

The patient refused all suggested treatment options, including topical retinoids and cryosurgery.

DISCUSSION

Syringomas are commonly classified as benign neoplasms derived from the eccrine duct (3). The presence of eccrine enzymes such as succinic hydrogenase, phosphohorylase, and leucine aminopeptidase provides further evidence of their eccrine rather than apocrine differentiation, as originally thought (6). Eruptive syringoma is a rare variant first described by Jacquet and Darier in 1887. It occurs more frequently among women.

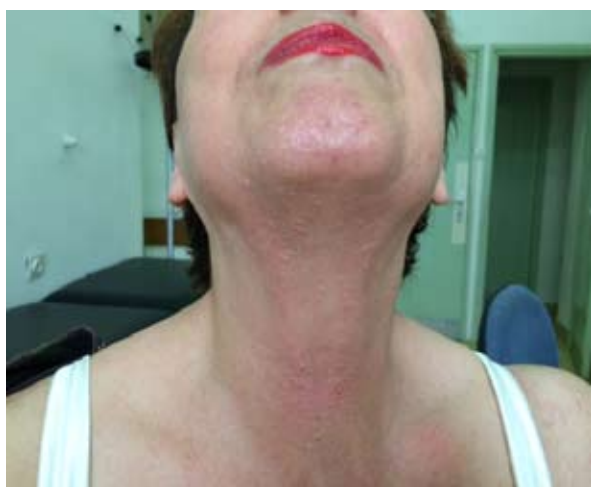


Figure 1. Multiple pink papules with erythematous halos in the neck region, 1-3 mm in diameter.

Clinically they appear as multiple, small, firm, and skin-colored or slightly yellow soft papules, 1-3 mm in diameter. They typically present in a bilateral, symmetrical distribution, but there have been reports of unilateral, unilateral nevoid, bathing trunk, and generalized distribution. Eruptive syringomas most commonly involve the face, anterior surfaces of the neck, chest, abdomen, upper extremities, and genitalia. There are some differences between prepubertal and postpubertal distribution. The neck and anterior trunk are more frequent sites before the age of 15 years. After 15 years of age, the apocrine localization is infrequent (7).

The eruptions are generally asymptomatic, although pruritus has been reported in some cases, especially in conjunction with perspiration. Vulvar syringomas were reported as an unusual cause of pruritus (8,9). The pathogenesis of eruptive syringomas still remains unclear. Syringoma is currently believed to be a neoplastic process.

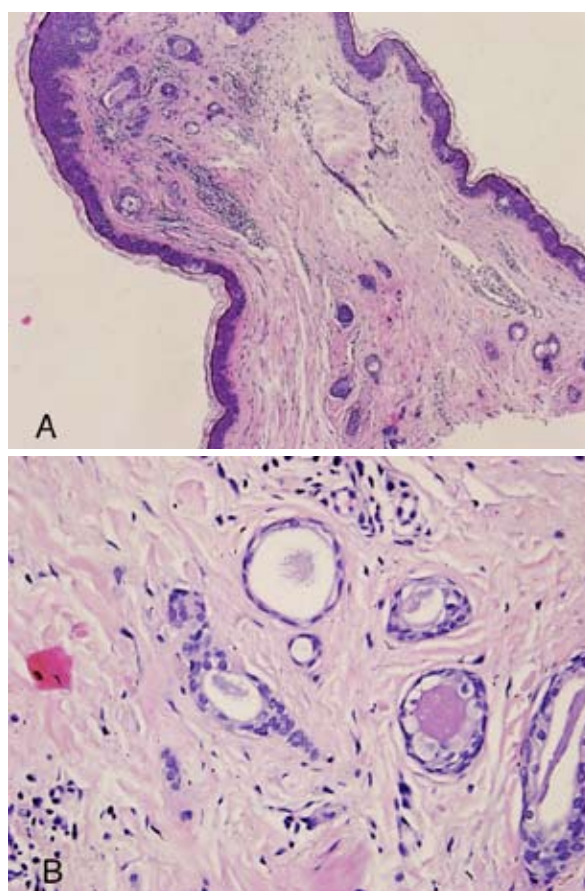


Figure 2. Eruptive syringoma demonstrating ducts formed by two layers of epithelial cells and filled with eosinophilic material, with elongated tails of epithelial cells; (a) hematoxylin and eosin (H&E) stain $\times 40$; (b) H&E stain $\times 120$.

Guitart *et al.* proposed the new term “syringomatous dermatitis”. They reported 2 cases of maculopapular eruptions with histologic findings of lymphocytic infiltrate involving intraepidermal and dermal portions of the eccrine ducts which evolved during next few months into tortuous syringomatous proliferation of ductal structures (10). Garrido-Ruiz *et al.* reported a distinct eruptive syringoma over the pubic and inguinal area which had developed after intensive waxing (11).

Similar syringomatous changes have been reported in association with alopecia areata, prurigo nodularis, and radiation dermatitis. In these cases, the initial inflammatory processes may precede the appearance of eruptive syringomas (12,13,14,15).

The higher incidence of syringoma in women, proliferation of lesions at puberty, exacerbation during pregnancies and enlargement during the premenstrual period may suggest a hormonal role in some cases (8).

The clinical diagnosis of eruptive syringoma is relatively difficult. Clinically they may be mistaken for acne vulgaris, sebaceous hyperplasia, milia, lichen planus, eruptive xantoma, urticarial pigmentosa, hidrocystoma, plane warts, granuloma annulare, and hystiocytosis (16,17).

Establishing the diagnosis of syringoma is based on histological examination. The haematoxylin and eosin stain shows the presence of multiple small ducts and epithelial cords within the dermis. The ducts are lined by two rows of flattened epithelial cells, the outer layer bulging outward to create a comma-like tail (7). Clear cell syringoma is a rare histological variant of syringoma which cannot be clinically separated from common variants. This variant is formed by cells that have a pale cytoplasm as a result of glycogen accumulation and is strongly associated with diabetes mellitus (18,19).

Our case is unique for several reasons. The localization of syringoma in our patient was the face, neck, axillae, and antecubital area, which is characteristic for prepubertal localization. As mentioned earlier, apocrine localization in postpubertal patients is infrequent. We have found only one report of eruptive syringoma onset in the seventh decade of life (20). Our patient also had symptomatic eruptive syringoma with pruritus and erythema around the lesions. We did not find any inflammatory infiltrate in our biopsy or signs of contact sensitization in a patch test. However, we took only one biopsy and it was performed at a “late stage” regarding a possible inflammatory phase.

Some authors reported a possible relationship between medications and eruptive syringoma occurrence (20,21). In our case, the patient started her medication (lisinopril, levothyroxine) after the skin changes developed, so we could not establish such a connection.

The treatment of syringoma is performed for cosmetic reasons. The goal of therapy for syringomas should be destruction of the tumor with minimal scarring and no recurrence. Treatment modalities have included dermabrasion, various methods of excision, cryosurgery, electrodesiccation, chemical peeling, and oral and topical retinoids (22,23,24).

A recent report described a successful treatment of symptomatic eruptive syringomas with 1% topical atropine. The use of topical atropine resulted in the disappearance of the pruritus and a discrete reduction in the size of the lesions without any side effects (25). However, because syringomas are embedded within the dermis, complete removal is often impossible. Any method of chemical or surgical destruction is associated with the risk of scarring.

CONCLUSION

Our case represents a distinctive clinical presentation of eruptive syringoma that should be considered in the differential diagnosis of papular dermatosis at any age.

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