

Late-onset of eruptive syringomas: a diagnostic challenge*

Cristina Resende¹

Rui Santos¹

Celeste Brito¹

Catarina Araújo¹

Teresa Pereira¹

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Abstract: Syringoma is a benign, adnexal tumor of the eccrine sweat gland ducts. Eruptive syringomas are a rare variant, occurring before or during puberty in most cases. A 57-year-old man was observed in our department, with a 10-year history of multiple brownish papules (1-4mm in diameter), localized on the neck, shoulders, trunk and axillae. The clinical diagnosis was cutaneous mastocytosis. Histopathological examination from a papule in the trunk was compatible with the diagnosis of syringoma. The patient was treated with isotretinoin, without any improvement. The clinical diagnosis of eruptive syringoma is difficult and histological examination is crucial for its diagnosis. Long-term morbidity is not associated with syringomas; they are treated for cosmetic reasons with unsatisfactory results.

Keywords: Exanthema; Mastocytosis, cutaneous; Syringoma

INTRODUCTION

Syringoma is a common, benign, adnexal tumor derived from the intraepidermal portion of the eccrine sweat ducts, most frequently found in the periorbital area.^{1,3} Syringomas are usually sporadic but some familial cases of eruptive syringomas have also been reported.³ The incidence of syringomas appears to be higher in Asians and African-Americans.^{1,2} Clinically, they appear as small papules and are skin-colored or slightly pigmented.^{1,4}

Syringomas are clinically divided into four types: localized, familial, a type associated with Down's syndrome, and generalized (encompassing eruptive syringomas).^{1,2,5,6} Eruptive syringomas are a rare variant, typically occurring as multiple, small, skin-colored to reddish brown papules, in large numbers, in successive groups, localized on the neck, chest, anterior trunk, axillae, or on the extremities.^{1,5,7} Syringomas are more frequent among women and the eruptive form also has a predilection for women.^{2,3,6} However, in contrast to classical syringoma, the

eruptive type occurs mostly before or during puberty.^{1,3} Eruptions are generally asymptomatic but pruritus has been reported in some cases.^{1,5} They are usually incidental, though some cases may be familial.^{5,6}

The clinical and histologic characteristics do not differ from those of classic syringomas.^{1,5}

We presented a case of late-onset, eruptive syringomas in a 57-year-old man who was first diagnosed as having cutaneous mastocytosis.

CASE REPORT

A 57-year old man was admitted to our department with a 10-year history of papular dermatosis, involving the trunk. The lesions appeared at the same time and were asymptomatic, except for occasional, mild pruritus during perspiration. He denied any medical problems and using medications. No other family members had been affected by similar skin lesions.

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¹ Hospital de Braga - Braga, Portugal.

Physical examination revealed multiple, brownish erythematous, flat-topped papules 1-4mm in diameter, distributed on the neck, shoulders, anterior chest, abdomen and axilla (Figure 1). The lesions were monomorphic, bilateral and symmetrical. Darier sign was negative. The remainder of the physical examination was unremarkable. Laboratory examinations did not show abnormal findings. He was initially suspected of having cutaneous mastocytosis.

A skin biopsy was obtained from a lesion on the trunk. Histological examination of this specimen revealed a normal epidermis and a dermal tumor, which was filled with multiple small ducts embedded in a fibrous stroma. The ducts were lined by two rows of epithelial cells and some had amorphous, keratinous material in their lumina (Figure 2).

Taken together, the clinical and histological findings were compatible with the diagnosis of eruptive syringoma. The patient was treated with low-dose isotretinoin (5 mg/day, alternate days), without any improvement.

DISCUSSION

The clinical diagnosis of eruptive syringoma is relatively difficult, as it can be confused with other skin diseases, including acne vulgaris, sebaceous hyperplasia, milia, lichen planus, xanthelasma and urticaria pigmentosa.^{1,5,6} Thus, histological examination is crucial for its definitive diagnosis.¹

Our patient was initially suspected of having cutaneous mastocytosis, based on dermatologic examination. Moreover, diagnosis was complicated by the late age of onset of multiple syringomas in our patient, since the majority of eruptive syringomas described in the literature occurred before or during puberty.^{1,5} The definitive diagnosis of syringomas in our patient was made by histological examination because syringomas exhibit distinctive, histological features.^{5,6}

Histological examination of syringomas revealed a dense, fibrous stroma, with a proliferation of eccrine-type ducts, lined by rows of cuboidal cells, forming dermal strands and cords, while the lumen was filled with eosinophilic, amorphous, periodic acid-Schiff-positive material.^{1,5,6}

Consequently, eruptive syringomas must be considered in the differential diagnosis of eruptive papular dermatosis at any age, even in adult and elderly patients (like ours).^{2,5} Our patient was diagnosed with generalized eruptive syringoma, as he had multiple, skin-colored to brownish papules, symmetrically distributed on the upper extremities, chest, abdomen, and anterior aspect of the thighs.

Syringomas are benign lesions and may spontaneously resolve, or more commonly, remain stable.^{4,5} Regression of the lesions in adulthood has been observed but it is exceptional.^{3,5} Treatment of syringomas is performed only for cosmetic reasons and no long-term morbidity is associated with the condition.^{4,5} Therapeutic options for syringomas are abundant



FIGURE 1: Multiple, brownish erythematous, flat-topped papules of 1-4mm in diameter, distributed on the neck, shoulders, anterior chest, abdomen and axillae

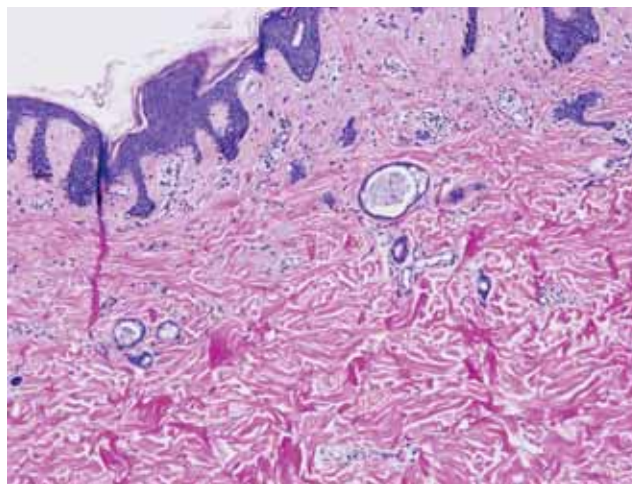


FIGURE 2: Histological examination revealed a normal epidermis and a dermal tumor, which was filled with multiple, small ducts embedded in a fibrous stroma (haematoxylin and eosin, magnification 400x)

but generally unsatisfactory. They also carry a risk of scarring because syringomas are localized in the dermis.^{4,5} Treatment modalities have included surgical excision, dermabrasion, electrocautery, cryosurgery, chemical peeling, topical atropine and topical and oral retinoids.^{4,6} Treatments for syringomas are generally inefficient and no treatment can eliminate the risk of recurrence.^{1,4}

We discussed this case due to the rarity of eruptive syringomas, and to emphasize that they must be considered in the differential diagnosis of eruptive papular dermatosis at any age. □

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MAILING ADDRESS:

Cristina Isabel Pinho Resende
Department of Dermatology and Venereology
Hospital de Braga
Sete Fontes - São Victor.
PT-4710-243 Braga, Portugal.
E-mail: cristinapresende@gmail.com

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