

## Syringoma: A Review of Twenty-nine Cases

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**The purpose of our study was to identify the clinical characteristics, epidemiologic data and histologic features in 29 cases of syringoma with a duration of lesions prior to the observation between 1 and 25 years. Only one patient complained of moderate itching. In two cases the lesion was solitary, in another the papules formed a lichenified plaque. In six patients only the eyelids were involved and in two patients a symmetrical localization on the forearms was observed. The other 18 patients showed generalized syringoma, 16 with an eruptive onset, 6 of which were familial. One of our cases showed lesions mimicking urticaria pigmentosa and two patients were affected by Down's syndrome. In two cases, histopathology showed association between syringoma and a melanocytic naevus and in one patient with a solitary lesion a clear cell syringoma was observed.**

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Syringoma is a benign tumour of eccrine origin which derives from eccrine ducts. Although the variety localized on the eyelids in middle-aged women is the most frequent, many other clinical variants differing in age of onset, location and clinical aspect have been reported in the literature (1–14).

Friedman & Butler (1) proposed a classification of syringoma according to clinical features and associations. This consisted of four principal clinical variants of syringoma: a localized form, a familial form, a form associated with Down's syndrome and a generalized form which encompasses multiple and eruptive syringoma. In eruptive syringoma, a rare variant first described by Jacquet & Darier in 1887 (15), the lesions occur in large numbers and in successive crops on the anterior

chest, neck, upper abdomen, axillae and periumbilical region at puberty or during childhood (6, 16–19). More rarely, cases of eruptive syringoma with wider involvement of the body have also been reported.

The aim of our study is to report our experience regarding syringoma by reviewing the clinical and histopathologic features of a series of 29 patients with histologically diagnosed syringoma observed over a 12-year period.

### PATIENTS AND METHODS

All cases with a histopathologic diagnosis of syringoma observed in our Dermatology Department between January 1983 and January 1995 were selected for this retrospective study and the multiple section specimens stained with haematoxylin-eosin and PAS were reviewed. On the basis of iconographies, medical case notes and anamnesis we tried to arrange our patients following the classificational criteria proposed by Friedman & Butler (1).

#### Clinical features

Twenty-nine Italian patients (16 females and 13 males; ratio 1.2:1) were included in this study. The mean age of the patients was 40 years (range 11–78 years). There was a slightly higher incidence in the 5th and 6th decades. The duration of the cutaneous lesions prior to observation varied from 1 to 25 years. In 11 patients the age of onset was in the 2nd decade, in 7 patients in the 4th, while in the other patients the age of onset was equally distributed among the 1st, 3rd, 5th and 6th decades. None of our patients revealed subjective symptoms and only one complained of moderate itching.

The clinical data relative to our patients are summarized in Table I.

#### Histopathologic features

In our series, all the histologic features reported in the literature were present, including sclerosis of the collagen, lymphomonocytic perivascular infiltrates of varying degrees and a granulomatous foreign-body-like reaction. In a 26-year-old woman with a widespread papular eruption, a close association of a junctional cellular naevus and a syringoma was found without intermingling of melanocytes and



Fig. 1. Eruptive syringoma with multiple slightly hyperpigmented papules ranging in size between 2 and 5 mm localized on the neck.

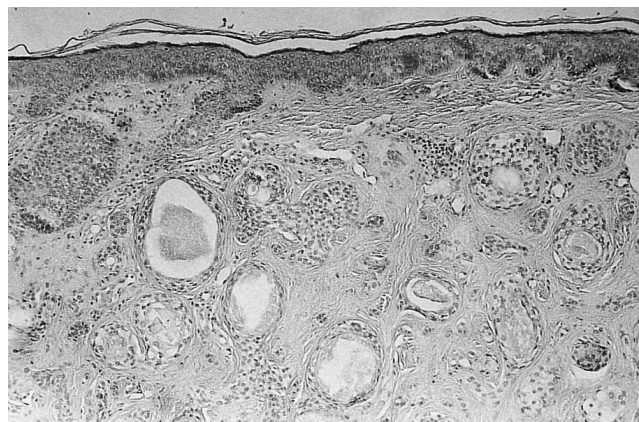


Fig. 2. Haematoxylin Eosin 100×. Histologic examination of a biopsy specimen taken from a typical papule of eruptive syringoma. The overlying epidermis shows hyperpigmentation.

Table I. Clinical and associated data of 29 patients with syringoma

	Sex/age (years)	Clinical variant	Onset	Morphologic features	Sites involved	Associations and familial cases
Case 1	M/78	Localized solitary		Non-pigmented plaque	Right temple	
Case 2	M/52	Localized solitary		Dome-shaped nodule 7 mm in diameter with a smooth translucent surface	Right upper eyelid	
Case 3	F/47	Localized multiple unifocal unilateral		Brownish papules confluent in a lichenified plaque	Volar surface of the right wrist and forearm	
Cases 4–9	F/23–59 (n=5)	Localized multiple unifocal (infraocular)	Eruptive in a 39-year-old female	Multiple papules	Eyelids symmetrically	Down's Syndrome in a 22-year-old male
Case 10	M/43	Localized multiple unifocal (acral?)	Eruptive	Multiple papules	Forearms and wrists symmetrically	
Case 11	F/60	Localized multiple unifocal (acral?)	Eruptive	Multiple papules	Forearms symmetrically	Tubular breast adenoma
Cases 12–27	M/14–57 (n=6) F/11–60 (n=10)	Generalized	Eruptive (14 cases) Unknown (2 cases)	Multiple flesh coloured, yellowish or slightly hyperpigmented papules 2–5 mm in size (Fig. 1)	Widespread involvement (2 cases) Trunk (14 cases)	Down's Syndrome in a 15-year-old male 6 familial cases
Case 28	M/36	Generalized	Eruptive	Multiple lichen planus-like papules	Neck (11 cases) Axillae (3 cases) Abdomen (3 cases)	
Case 29	M/27	Generalized	Eruptive	Multiple maculo papular, urticaria pigmentosa-like lesions Darier's sign +	Upper limbs (3 cases) Eyelids (1 case) *	

\* The total number of affected areas is higher than the number of patients because a single patient may have multiple involved areas.

syringoma cells. On the contrary, in patient no. 1 in Table I, histology showed the intermixing of melanocytes and syringoma cells. In patient no. 2 in Table I, the histologic examination revealed an unusual preponderance of clear cells. Moreover, we noticed the presence of hyperpigmentation, sometimes very marked, in the basal layers of the overlying epidermis in 12 of our patients (9 with eruptive syringoma, 2 with syringoma localized on the eyelids, 1 with clear cell syringoma) (Fig. 2) which was not necessarily associated with a brownish or a yellowish colour of the lesions.

## DISCUSSION

Although syringoma is quite common, studies relative to large series of patients have, to our knowledge, rarely been reported in the literature (2, 16). It should be pointed out that our series involved a double selection: a primary selection on clinical grounds (patient consulting a dermatologist) and a second (patients whose syringoma was excised and sent to a pathologist), so this is not an epidemiologic study. Moreover, our patients were all Caucasian and attended the same dermatologic department. All this, in our opinion, may explain the particular distribution of our patients regarding age, sex and variety of observed syringoma. In fact, the overrepresentation of adolescent onset in this series is due to several patients (16 cases) with eruptive form, 9 of which with onset at under 20 years. The other seven patients, however, did not develop the disease until later in life, even in the fifth and sixth decades. The onset of eruptive syringoma in adults or in elderly patients, though rarely reported, can be found in the literature (10, 24, 26, 28, 29).

Two of our patients presented a solitary syringoma (cases 1

and 2, Table I). In one case it was found in close association with a melanocytic naevus. This could be merely a chance association, even if other cases have been reported previously (20–23) and the hypothesis of a circumscribed dysembryoplasia involving both the melanocytic cells and the sudoral adnexa could be made (22). In the other patient, histology showed an unusual microscopic preponderance of clear cells. The well-established association of clear cell syringoma with diabetes mellitus was not present in our patient and, in addition, unlike the other clear cell syringoma cases reported (24–26) the lesion was solitary. We have been able to find in the literature only one case of solitary syringoma localized on the ankle (27). Solitary syringoma, although rare, is probably underestimated.

One case of unilateral syringoma where the papules formed a lichenified plaque was observed (case 3, Table I). Other cases of unilateral syringomas have been described (7–9), one with a similar aspect to our case (9).

We observed two patients with syringomas symmetrically distributed on the wrists and forearms (cases 10 and 11, Table I), but not on the hands, as reported in all the cases of acral syringoma (11, 14, 28). We suggest classifying cases with an exclusive involvement of the wrists and forearms in the variant "acral syringoma" and in so doing all our cases could be included in the clinical classification proposed by Friedman & Butler (1).

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