

An Unusual Case: Intensely Pruritic Eruptive Syringoma

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Abstract

Eruptive syringoma is a generalised variant of syringomas. They are rare benign tumours of the eccrine sweat ducts typically presenting asymptotically, but occasionally with pruritus when associated with perspiration or in individuals with a background of atopy. Although treatment is usually mostly oriented to cosmetic improvement, it often proves difficult and unsatisfactory.

We present a diagnostically challenging case of eruptive syringoma in a young woman who presented atypically, with an intensely pruritic dermatosis.

Case Report

An 18-year old Afro-Caribbean woman presented with a 2-year history of a gradual onset, severely pruritic eruption affecting her upper chest and arms. The eruption started as crops of itchy papules on her upper chest, which were slowly spreading to her abdomen and inner aspect of both forearms. Emollients and topical steroids had been ineffective. She had no past medical history or family history of note. In addition to the pruritus the patient had significant cosmetic concerns regarding the rash.

On examination there were multiple smooth, monomorphic, 2-3mm hyperpigmented papules of firm consistency and symmetrically distributed on the inner aspect of forearms, upper chest and abdomen (Figure 1). The remainder of her skin was spared.

The histopathological examination revealed a focal dermal proliferation of small cystically dilated eccrine ducts in the mid-reticular dermis, some of them cystically dilated or comma-shaped. No infiltrative edges, necrosis or cytological atypia was found (Figure 2). These findings were in keeping with eruptive syringoma.

Discussion

Syringomas (from the Greek word syrinx, meaning pipe or tube) are benign ductal tumours which differentiate toward the intraepidermal portion of the eccrine duct. There are four main clinical variants recognized: the localised, familial, Down's Syndrome-associated and generalised variants [1]. Eruptive syringoma is included within the generalised variant.

Although syringomas are relatively common, the eruptive form is rare, with less than one hundred cases having been reported since the first report in 1887 by Jacquet and Darier [2]. Young women are more frequently affected [3-4]. The papules commonly occur in localised crops over the chest, neck, axilla and upper abdomen [2]; however there have been cases reported of more widespread involvement [5]. Most commonly they are asymptomatic, but have rarely been reported to be pruritic particularly in the setting of perspiration [6] or in patients with a background of atopy [4].

The histology typically shows multiple small ducts and epithelial cords within the superficial dermis. The ducts are lined by two rows of flattened epithelial cells, the outer layer bulging outward to create a "comma-like or tadpole tail" [4].

Although the pathophysiology of eruptive syringoma is not fully understood, they may represent a reactive response to an inflammatory reaction rather than a true adnexal neoplasm, inducing a hyperplastic response of the eccrine duct. For this reason some authors have proposed the name of 'syringomatous dermatitis' [7]. In some cases, syringomas can be associated with alopecia areata [8], prurigo nodularis [9] and radiation therapy [10]. An isolated case of pubic syringomas triggered by waxing of the area has also been described [11]. The authors postulate that the inflammatory reaction induced by trauma may cause hyperplasia of the eccrine ducts. A hormonal association has also been suggested [12].

Furthermore, there are recent reports of syringomas related with immunomodulating drugs

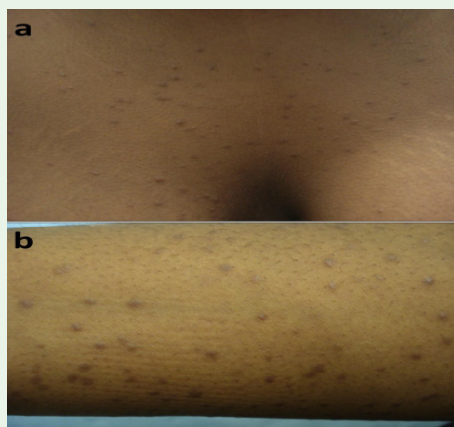


Figure 1: Multiple small hyperpigmented papules on (a) upper chest and (b) arms.

[13], antiepileptic medication [14] and carbamazepine [15]. The role of these drugs in the development of these adnexal tumours is still unknown.

The differential diagnosis of eruptive syringoma is wide and includes lichen planus, papular eczema, sarcoidosis, eruptive xanthoma, steatocystoma multiple and milia. Papular eczema is common in dark skinned races and can be intensely pruriginous, representing the most important clinical simulators of syringomas. Histologically, it is characterised by superficial perivascular spongiotic dermatitis. Micropapular sarcoidosis can also mimic syringomas, but it is usually asymptomatic and typically shows non-necrotising granulomas. Lichen nitidus can also be presented as small monomorphic pinhead papules which are usually asymptomatic, revealing lymphohistiocytic lichenoid infiltrate enclosed within collaret's of epidermal acanthosis and multinucleated cells.

The clinical course of eruptive syringoma varies from spontaneous resolution to, more commonly, remaining stable [5]. However it has not been found to have any long term morbidity associated. The treatment modalities for this benign dermatosis include cryotherapy, dermabrasion, electrodesiccation and cautery, and most recently CO₂ laser resurfacing therapy [16-17]. The latter has shown promise in

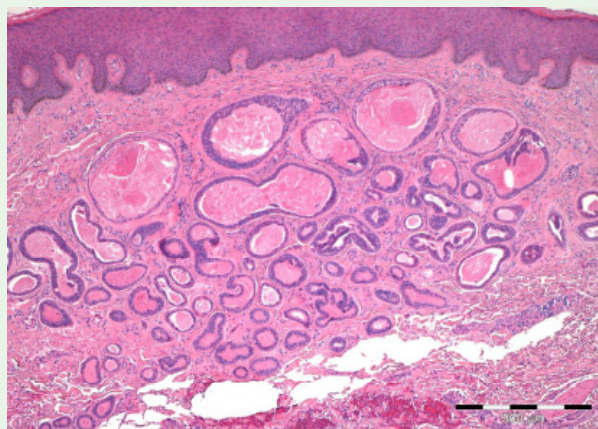


Figure 2: Histology findings of eruptive syringoma: Dermal proliferation of small cystically dilated eccrine ducts in mid-reticular dermis, some of them cystically dilated or comma-shaped.

providing a safe and non-scarring method of treatment for periorbital syringomas [17].

Although treatment in eruptive syringoma is usually oriented to meeting cosmetic demand, this case was particularly challenging given the unusual symptom of intense pruritus. Although we offered the patient a course of narrow band UVB light for itchy relief, it was declined due to her university commitments. Furthermore, antihistamines made little difference to her pruritus. Laser treatment was also recommended, however she was concerned by the potential risk of recurrence and pigment changes, and therefore also declined this.

In conclusion, the presence of an itchy papular eruption in a young woman should raise the clinical suspicion of eruptive syringoma. This condition should be included among the differential diagnosis of itchy papular dermatoses.

Conclusion

Learning points

- Syringomas are benign sweat gland tumours, which may present as a localised form, familial, Down's syndrome associated and generalised.
- Eruptive syringoma is more common in young women, typically presenting as multiple, asymptomatic, flesh coloured papules on trunk and upper limbs
- More rarely they can present as a pruritic papular dermatosis
- The differential diagnosis is extensive therefore biopsy is usually required
- Treatment of eruptive syringoma is often challenging and unsatisfactory.

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