Generalized eruptive syringomas

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Abstract

Eruptive syringoma is a rare variant of syringoma, benign neoplasms of the eccrine sweat ducts that appear on the face, neck, chest, and axillae of predominately Asian and African American women before or during puberty [1, 2]. Lesions appear as small skin-colored or slightly pigmented, flat-topped papules [2]. The condition can be cosmetically disfiguring and difficult to treat, especially in dark-skinned patients. The investigators report a 52-year old Guyanese woman who presented with widespread, chronic, non-pruritic and nontender, skin-colored papules that arose approximately 20 years earlier. A punch biopsy of affected skin was obtained and the histological diagnosis was eruptive syringoma. The patient pursued no further treatment, after discussion of costs and risks.

Keywords: eruptive syringomas, syringomas, benign adnexal neoplasm

Introduction

Syringomas are benign adnexal tumors of eccrine sweat ducts that affect 1% of the general population and are more common in women [3-5]. The name is derived from the Greek word syrinx, meaning pipe or tube. They appear as asymptomatic, flat-topped, skin-colored to slightly pigmented papules, 1-4mm in size, that classically develop in the periorbital area, but can also affect other parts of the body. Syringomas are classified into four clinical variants: localized, familial, associated with Down syndrome, and generalized/eruptive. Eruptive syringomas, which were first described in 1887 by Jacquet and Darier, are quite rare, affecting mainly Asian and African-American patients and females during puberty or the third and fourth decade of life [2]. The eruptive variant can be aesthetically disfiguring and difficult to treat.

On histologic examination, syringomas are composed of single- to double-layered cuboidal epithelial cells with pale, eosinophilic cytoplasm that form discrete nests, cords, or tubules in the upper dermis, and rarely, in the deep reticular dermis [6]. The surrounding stroma is often sclerotic. Syringomas often have periodic acid-Schiff–positive, eosinophilic material in the tubular lumen and short epithelial tails, often compared to a "tadpole" or "paisley tie" appearance [3].

Case Synopsis

A 52-year-old Guyanese woman presented to the dermatology clinic for evaluation of numerous skin lesions on the face, trunk, and extremities. The non-pruritic, non-tender papules arose approximately 20 years earlier; the papules had been stable in recent years. The patient was otherwise healthy; she denied any known medical conditions and did not take any prescription or herbal medications. She did not have Down syndrome and her parents and siblings did not have similar skin lesions. The lesions did not improve with emollients.

Physical examination revealed innumerable skin-colored papules on Fitzpatrick type VI skin, distributed periorbitally and diffusely throughout the face and neck (**Figure 1**a). Similar small papules

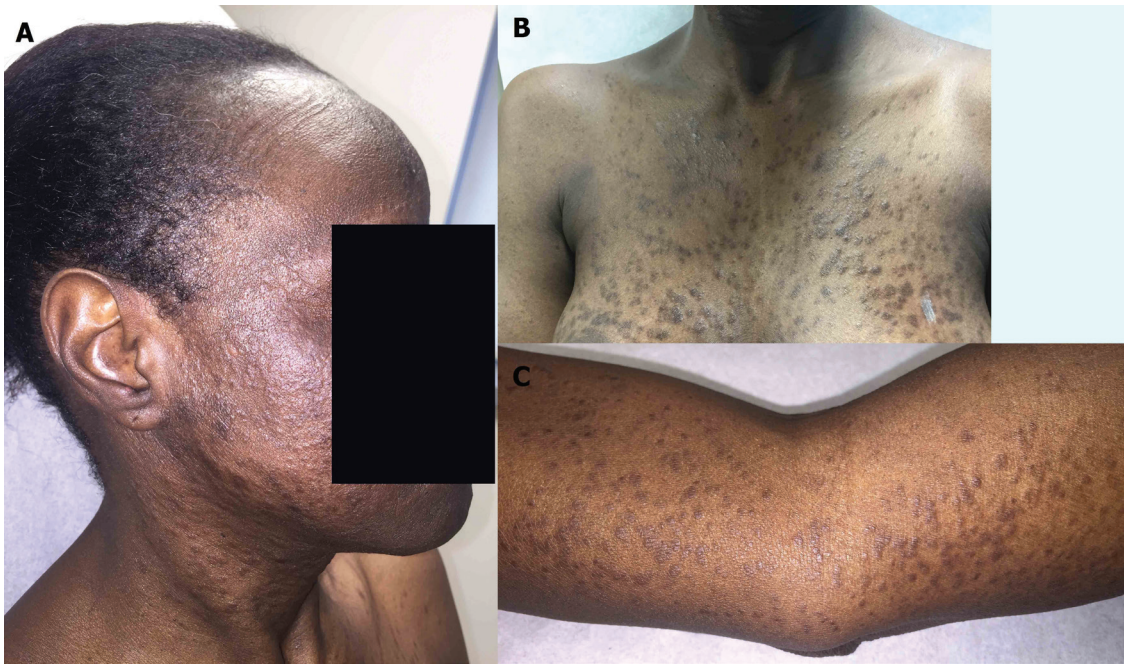

Figure 1. *A) Numerous skin-colored to slightly pigmented papules are shown on the face and anterior neck. B) Larger, distinct hyperpigmented oval brown papules on the anterior chest. C) Smaller hyperpigmented papules on the arms.*

were also found on the arms and legs, though more sparsely distributed (**Figure 1**c). She had larger distinct hyperpigmented oval brown papules on the anterior trunk (**Figure 1**b). Darier sign was negative. The lower extremities were relatively spared, as were the ears, nasal rim, oropharynx, and lips. A biopsy was performed.

Histopathologic examination of the punch biopsy revealed an upper dermal proliferation of small sweat ducts lined by one or two layers of cuboidal epithelium with a tadpole-like tail, consistent with syringoma (**Figure 2**). Parts of the stroma surrounding the lesions were fibrotic and the lesions were well-demarcated from the stroma. No mitotic figures or atypia were observed. After discussion of costs of treatment and risks of scarring and hypopigmentation, the patient decided against further treatment of her condition.

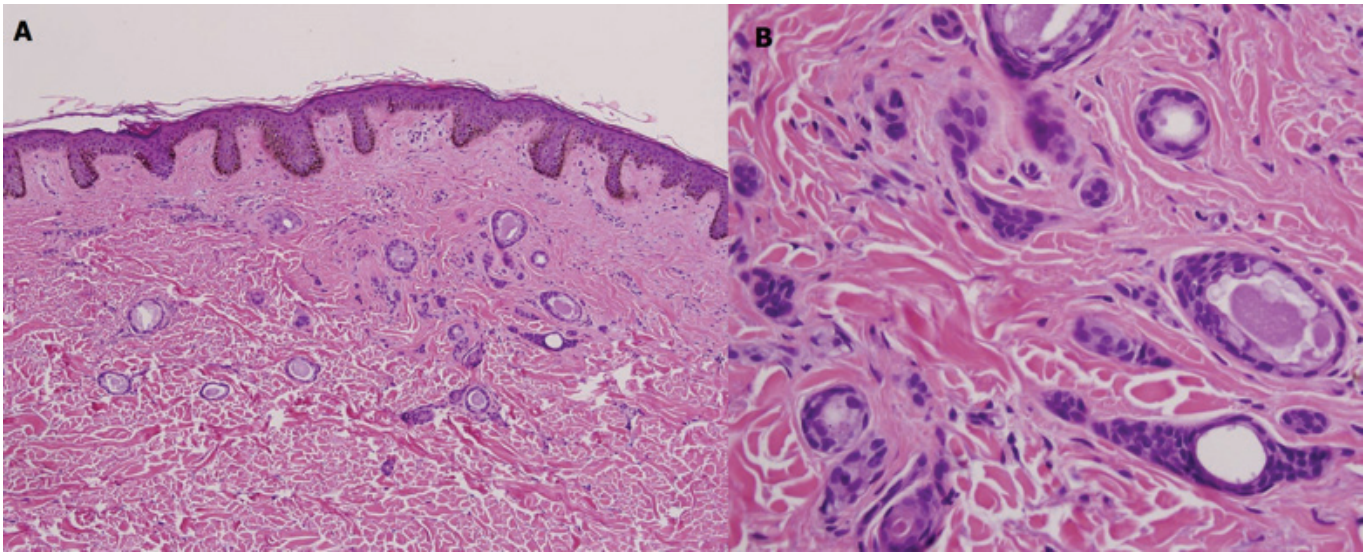

Figure 2*A-B. A) Dermal proliferation of sweat ducts within a fibrotic stroma 10x. B) In detail, showing single- to double-layered cuboidal epithelial cells with pale, eosinophilic cytoplasm that form nests, cords, or tubules, H&E 40x. Periodic acid-Schiff-positive, eosinophilic material are seen within the tubular lumens. The comma-like tails of epithelial cells contribute to the tadpole' or 'paisley-like' shape of the adnexal neoplasm.*

Case Discussion

Although syringomas are benign, the lesions can be aesthetically disfiguring, especially in patients presenting with the eruptive variant. Lesions appear on the extremities, abdomen, neck, and anterior trunk, in addition to the face. The pathophysiology of eruptive syringomas is unknown, but some speculate hormonal influence to be a major cause [7], whereas others cite an inflammatory trigger in response to autoimmune conditions, trauma from waxing, radiation, or picking, and heat stimuli [8-10]. Although they have been reported to spontaneously regress in rare cases, they commonly remain stable [3]. Eruptive syringomas have also been described to occur mainly over the anterior trunk, including the anterior neck, chest, axilla, and inner aspect of the upper arm; such predominately anterior lesions were observed in our patient [11].

Treatment modalities are cosmetic and may include topical or systemic retinoids, laser ablation, electrodessication, cryosurgery, trichloracetic acid, dermabrasion, chemical peels, and a variety of other destructive measures. Treatment is often unsatisfactory, particularly in the eruptive variant, as the tumors are deep in the dermis and many of these modalities may lead to scarring and reoccurrence [2, 12]. Treatment in dark-skinned patients presents an additional management challenge, as laser resurfacing and cryosurgery may cause unwanted pigmentary changes [12]. One case study demonstrated good cosmetic outcome with minimal side-effects in trichloroacetic acid (TCA) and CO2 laser ablation of facial syringomas in an African-American patient [13]. However, many other case reports showed scarring with such ablative procedures [14]. The adverse effects, uncertain outcomes, and high out-of-pocket costs of such cosmetic therapies may potentially deter many patients from seeking treatment.

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