Generalised Eruptive Syringomas: a Diagnostic and Cosmetic Conundrum

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Abstract

Observation: Syringomas are benign, adnexal tumours that are derived from the intraepidermal portion of the eccrine sweat ducts. Syringomas are usually sporadic but familial forms have been seen rarely. Lesions are in the form of bilaterally symmetrical firm skin colored papules which are commonly seen in periorbital areas and are cosmetically a concern. Even though periorbital is the most common location, rare forms have been described. Generalised eruptive syringomas is a rare form. Here, we report a patient with sudden eruptive syringomas that included the whole body posing a difficulty in the diagnosis.

Introduction

Syringomas are common, benign, adnexal tumours that are derived from the intraepidermal portion of the sweat ducts. The word syringoma is derived from the Greek word 'syrinx' that means a tube [1]. These are more commonly seen in females and periorbital area is the most common site involved. Friedman and Butler described four types of syringomas- localised, generalised, familial and those associated with Down Syndrome [2]. Syringomas present as asymptomatic, multiple, firm, skin colored papules that are bilaterally symmetrical in distribution [3]. Various treatment modalities have been tried that may include chemical cautery, electocautery, cryotherapy, excision, dermabrasion, topical and oral retinoids and CO2 lasers, with variable results [4]. Diagnosis is clinical and biopsy is only required in case of atypical presentation. Such an atypical presentation with generalised eruptive syringomas in an adult female patient is being presented.

Case Report

A 23 year-old female presented to the skin OPD with 15 years old history of multiple papules over whole body. The lesions were asymptomatic. These initially developed over face and then gradually spread over to rest of the body in one year. After that the lesions remained stable. There was no family history or any history of similar complains in the past. Mucocutaneous examination revealed bilaterally symmetrical, multiple, firm, skin colored and flat topped papules with varying sizes and well defined irregular margins which were present over face, trunk, bilateral upper and lower limbs (Figures 1a and b) (Figure 2). Examination of scalp, nails and mucous membranes was found to be normal. There were no associated cutaneous or systemic diseases. Based on history and clinical examination, a differential diagnosis of verruca plana, epidermodysplasia verruciformis and eru-
tive syringomas and generalised lichen planus was kept. After taking informed consent, skin punch biopsy was taken from lesion on arm which reported an epithelial neoplasm involving upper reticular dermis and made up of solid epithelial islands and ductal structures which were lined with two or three layers of cuboidal cells. These cells present in solid cords showed attempt towards ductal differentiation. Stroma was made up of thickened collagen bundles with impression of syringoma. All other routine laboratory investigations were normal. Based on clinical and histopathology findings, a final diagnosis of generalised eruptive syringomas was made and patient was treated with isotretinoin 20mg daily. The degree of response to treatment could not be seen as the patient did not come for follow up.

Discussion

Syringomas are common benign adnexal tumours. These arise from intraepidermal portion of eccrine sweat glands. They usually arise during puberty and are more common in females [5,6] Periorbital location is the most common, but other forms may also be seen. Ours was one such unusual case with generalised eruptive syringomas in an adult female. Syringomas may resolve spontaneously or may require treatment which includes chemical cautery, electrocautery, cryotherapy, dermabrasion, excision, topical and oral retinoids and CO2 laser. The results have been variable. The only concern is that the lesions are cosmetically concerning for the patient. Our patient was started on isotretinoin 20 mg daily keeping in mind the generalised nature of the disease.

To conclude we can say that syringomas are benign but they can be cosmetically unappealing and a cause of concern for the patient and when the presentation is rare, they pose a diagnostic challenge for the doctor. Treatment is usually unsatisfactory and should aim at counselling the patient regarding benign nature of the disease.

References


