Reactive Eccrine Syringofibroadenomatosis in the Setting of Venous Insufficiency

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ABSTRACT

Reactive eccrine syringofibroadenomatosis describes multiple lesions of a subtype of the benign adnexal tumor eccrine syringofibroadenoma, which may be difficult to diagnose due to its rarity. This case illustrates a classic presentation of reactive eccrine syringofibroadenomatosis occurring within sites of a patient’s previous leg ulcers in the setting of chronic venous insufficiency. Recognition of this condition is essential for patient quality of life as well as monitoring for potential associated malignancy.

INTRODUCTION

Eccrine syringofibroadenomas (ESFAs) are rare adnexal tumors presenting clinically as multiple hyperkeratotic nodules. They are diagnosed histopathologically with findings of anastomosing epithelial cords surrounding fibrovascular stroma. Based on associated clinical characteristics, ESFAs are further classified into five different subtypes. One of these subtypes is reactive ESFA, which has been reported to occur in the setting of chronic ulceration. Reactive ESFA is a result of inflammatory damage to eccrine ducts, with subsequent dysregulation of adnexal growth and differentiation suspected as the underlying cause.

CASE REPORT

An 88-year-old male patient presented with a bilateral lower extremity rash of one year duration. He had a past medical history significant for type II diabetes mellitus and venous insufficiency, previously complicated by large, bilateral venous ulcers.

Physical examination revealed a well-appearing man with monomorphic, hyperkeratotic, papillomatous papules coalescing into large, well-circumscribed, cobblestoned plaques of the bilateral distal medial lower extremities (Figure 1). Importantly, the plaques were painless, and the patient confirmed that the presenting cutaneous findings had formed within the sites of previous ulceration. Furthermore, he stated that the lesions were limited to the boundaries of the ulcers, and denied associated fever or dyspnea. Histopathologic examination revealed numerous anastomosing strands of small, benign epithelial cells with occasional duct formation enclosing a fibrovascular core (Figure 2), consistent with reactive eccrine syringofibroadenoma.
Eccrine syringofibroadenomas (ESFAs) are rare adnexal tumors most commonly occurring in elderly patients in the 7th and 8th decades of life. Typically presenting as isolated, hyperkeratotic nodular lesions with predilection for acral sites and extremities, ESFAs are comprised of five distinct clinical...
subtypes, including a class designated reactive ESFA. A rare phenomenon, reactive ESFA is a process hypothesized to arise from dysregulated adnexal growth and differentiation, as a result of repeated inflammatory damage to eccrine structures, particularly in the setting of chronic ulceration\(^1\). When involving multiple sites, “eccrine syringofibroadenomatosis” has been utilized to describe the lesions and degree of involvement\(^1\). Thus far, over 40 cases have been described in the literature, with the majority occurring in the setting of non-healing, chronic wounds and neuropathy\(^2\). Although benign lesions, ESFAs have been associated with neoplasms such as squamous cell carcinoma and pilomatrixoma\(^2\). Additionally, malignant transformation to eccrine syringofibroadenocarcinoma has been described in non-reactive subtypes\(^3\).

Clinically presenting as multiple verrucous nodules coalescing to form a characteristic “Streusel-bread” appearance, reactive eccrine syringofibroadenomatosis carries a differential diagnosis that often includes elephantiasis verrucosa nostra, tuberculosis verrucosa cutis (or other mycobacterial infections), atypical deep fungal infections, and retention hyperkeratosis related to poor wound care. The diagnosis is made histopathologically, with pathology demonstrating reticulated, anastomosing, epithelial cords with abundant surrounding fibrovascular stroma. Because of its rarity and potential association with malignancy, clinical recognition and subsequent diagnosis is important for affected patients, meaningfully impacting both treatment and quality of life\(^4\).

Thus far, a standard treatment has not been delineated for reactive ESFA\(^2\). Primarily following a benign course, reactive ESFA has been described to resolve spontaneously, although close monitoring is recommended given risk for concomitant malignancy\(^1\). Isolated reports have described successful treatment of lesions with surgical excision, but recommend targeting specific areas with the highest potential for malignant transformation within plaques covering large surface areas\(^3\). Multiple other modalities have also been trialed with variable efficacy, and include CO\(_2\) laser ablation, liquid nitrogen therapy, systemic retinoids, and scouting biopsies of larger, untreated lesions for serial monitoring\(^1,2\).

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