Reactive Eccrine Syringofibroadenoma: An Emerging Subtype

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In this month’s issue of Dermatology, 2 reports of eccrine syringofibroadenoma (ESFA) associated with inflammatory dermatoses are reported. These new cases of ESFA, along with those previously reported [1-7, 12-16], all have the particularity of being closely associated with another specific dermatosis (most frequently inflammatory or neo-plastic), suggesting that in some instances ESFA may occur as a consequence of recurrent eccrine duct lesioning. As no classification proposed to date accounts for this particular type of ESFA, I suggest to consider this growing group of ESFA that appears to result from eccrine ductal remodelling as a distinct subtype, namely ‘reactive ESFA’.

ESFA is a rare benign cutaneous adnexal lesion [8] that predominantly occurs in patients over 40 years of age. It has a strikingly polymorphous clinical presentation ranging from a solitary papule or nodule to multiple lesions with a linear or palmoplantar distribution; the latter is often referred to as eccrine syringofibroadenomatosis [3, 9, 10]. Despite the variability of clinical presentation, ESFA is histologically unique. It is a distinct tumour composed of a proliferation of anastomosing cords of monomorphous epithelial cells harbouring eccrine ductal formations, all admixed within an inflammatory fibrovascular stroma.

Over the last few years several classifications of this clinically and nosologically disparate adnexal lesion have been proposed, the latest and in my opinion most appropriate classification being that recently described by Starink [11]. Based on the clinical and histological analysis of 8 new cases and a review of the 36 previously reported cases of ESFA in the literature, Starink proposed a classification consisting of the following subtypes: (1) solitary ESFA; (2) multiple ESFA with hidrotic ectodermal dysplasia (Schöpf syndrome); (3) multiple ESFA without associated cutaneous findings, also called eccrine syringofibroadenomatosis; (4) non-familial unilateral linear ESFA, sometimes referred to as naevoid ESFA.

Unfortunately however, the above classification does not include a group in which the cases reported in this issue and those previously described fit appropriately, given their specific clinical characteristics and proposed pathogenesis. It appears from the increasing number of recent reports that ESFA can be associated with inflammatory or neoplastic dermatoses, or appear in the context of a peripheral polyneuropathy [1-7, 12-16]. These pathologies are all characterized by repetitive damage and regrowth of skin structures within affected sites and suggest that ESFA observed in their vicinity may have occurred as a consequence of recurrent eccrine duct lesioning. Indeed, eccrine ductal proliferation as a consequence of prior ductal disruption is a common response observed during wound healing and in inflammatory or neoplastic skin disorders [17]. In addition, the increased mast cell numbers observed in several of these cases [4,
15, 18] are a characteristic feature of healing wounds and also suggest that in certain instances ESFA may be a response of the eccrine duct to ongoing tissue remodelling. Taken together, these observations suggest that this emerging subtype of reactive ESFA associated with tissue remodelling may represent a specific type of eccrine ductal repair.

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