

Reactive Eccrine Syringofibroadenomatosis Associated with Venous Stasis and Plaque Psoriasis

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Sir,

Eccrine syringofibroadenoma (ESFA) is a rare benign tumour with differentiation towards the acrosyringium. There are five subtypes, one of which is known to occur in reaction to inflammatory and neoplastic dermatoses. We describe here a patient who presented with two nodules over his right shin of 10 years' duration against a background of chronic plaque psoriasis and venous stasis.

CASE REPORT

A 75-year-old Chinese man with a 30-year history of chronic plaque psoriasis for which he had been treated with topical steroids and coal tar was referred to our clinic by his family physician after an exacerbation of psoriasis. He had a medical history of ischaemic heart disease, hypertension and hypercholesterolemia. There was no family history of skin disease.

Examination revealed scattered typical psoriatic plaques involving approximately 15% of his body surface area. Over his right shin was a papillomatous nodule 1.8 cm in diameter, with a second smaller, firm and smooth nodule on the upper lateral aspect (Fig. 1). These nodules had been present and stable in size for about 10 years. Psoriatic plaques were present over the location of the nodules and they had also involved the same area over the shin in a relapsing manner during the disease course. In addition, there were features of brownish haemosiderin pigmentation over both his lower limbs, which had been present for about 15 years. Notably there was no pitting oedema, ulceration, dilated veins, or cellulitic changes present on the lower limbs. No lymphadenopathy or organomegaly was detected.

A punch biopsy of the papillomatous nodule was performed to exclude a squamous cell carcinoma. Histology showed anasto-

mosing strands of basaloid epithelial cells forming a net-like pattern connected to the undersurface of the epidermis (Fig. 2A). Neovascularization was observed, with thick-walled small vessels present throughout the dermis surrounded by fibrous bands of thickened collagen and deposits of haemosiderin (Fig. 2B). The histological changes were interpreted as eccrine syringofibroadenomatosis with venous stasis changes. Subsequent ultrasound duplex scan of the venous systems of the legs revealed incompetence of the left lesser saphenous vein.

Our diagnosis was therefore reactive eccrine syringofibroadenomatosis secondary to venous stasis and chronic plaque psoriasis. The option of surgical excision of the two nodules was offered, but the patient declined. He was treated with topical steroids and coal tar for his psoriasis. Measures to improve the underlying venous hypertension, including graduated compression bandaging, leg elevation and skin care were instituted. He was still on follow-up at the time of reporting.

DISCUSSION

ESFA is a neoplasm of the acrosyringium related to eccrine poroma and hidroacanthoma simplex, which are both neoplasms of the intra-epidermal sweat duct. It was first described by Mascaro in 1963 (1). Most of the patients are in their seventh and eighth decades of life. The clinical presentation is variable and non-specific, ranging from solitary lesions to multiple papules and nodules in a symmetrical or linear nevoid pattern (1, 2). The site of occurrence varies widely and the extremities are commonly affected.

Starink (3) classified ESFA into four subtypes: solitary ESFA, multiple ESFA with ectodermal dysplasia, multiple ESFA without associated cutaneous findings, and non-familial unilateral linear ESFA. French (4) subsequently proposed an additional subtype known as reactive ESFA. This subtype represents an epithelial change in relation to other inflammatory and neoplastic dermatoses, such as chronic ulceration and multiple trauma in diabetic and lepomatous neuropathy (5), burn scars (6), venous stasis (7), nail trauma, bullous pemphigoid (8, 9), erosive palmoplantar lichen planus (10), peristomal dermatopathy, nevus sebaceous, epithelioid hemangioma, and squamous cell carcinoma. To date, approximately 71 cases of ESFA have been repor-



Fig. 1. (A) Two nodules on the right lower limb on a background of treated psoriatic plaques and stasis pigmentation. (B) A nodule on the right shin with a papillomatous surface. The central defect is post-biopsy.

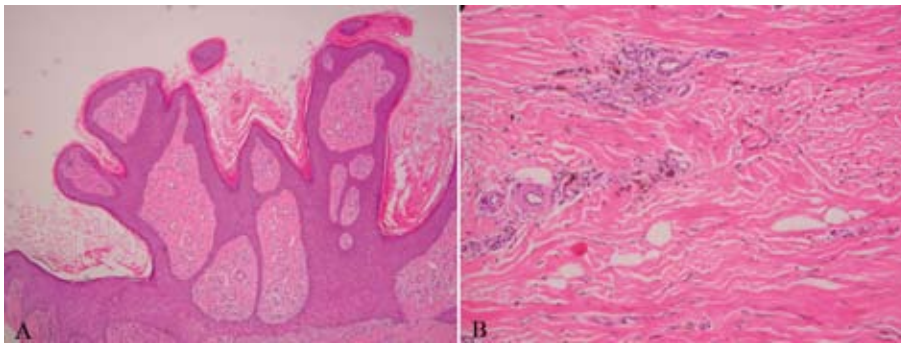


Fig. 2. (A) Anastomosing strands of basaloid epithelial cells form a netlike pattern and are connected to the undersurface of the epidermis. (H&E $\times 20$). (B) Neovascularization of thick-walled small vessels is present throughout the dermis, surrounded by fibrous bands of thickened collagen and deposits of haemosiderin (H&E $\times 40$).

ted in the literature and, of these, approximately 16 are reactive ESFA. To our knowledge, there is only one previous reported case of reactive ESFA secondary to venous stasis (7) and one previous case that was related to psoriasis (11).

In our patient, venous stasis was evident from the haemosiderin hyperpigmentation over his legs and the histological features, with venous incompetence demonstrated on duplex scan. Chronic venous hypertension and chronic plaque psoriasis would have resulted in chronic inflammation in the skin of the lower limbs. The increased mitotic and metabolic activity would have created an opportunity for aberrant proliferation of tissues and provided a basis for neoplastic influences. The development of the ESFA changes in our patient is as likely to be attributable to chronic venous stasis as it is to plaque psoriasis, as both conditions were present at the sites of the nodules. The use of coal tar may be a concern, but there is no evidence to date to suggest an increased incidence of skin cancers in patients treated with therapeutic doses of topical coal tar.

It is widely debated whether ESFA is a true neoplasm, a hamartoma, or a form of reactive hyperplasia. The manifestation of ESFA as multiple symmetrical lesions in ectodermal dysplasia (Schopf syndrome) (3) and the occurrence of ESFA in various inflammatory conditions suggest that it is a hamartoma (4, 8–10). On the other hand, malignant transformation into squamous cell carcinoma and porocarcinoma has been suspected to occur in eight cases in the literature (3, 12–14), and this may favour the classification of ESFA as a true neoplasm. Of note, all these cases involved the non-reactive forms of ESFA; five had occurred in solitary lesions, two had occurred against a background of ectodermal dysplasia, and one developed from a nevoid lesion.

ESFA is a histological entity and the subtypes probably should not be considered as a homogenous condition. The different subtypes have distinctly different clinical manifestations, with probable different pathogenesis and biological behaviour. Solitary ESFA behaves like a neoplasm; multiple ESFA associated with ectodermal dysplasia and ESFA without associated cutaneous findings appears hamartomatous, while non-familial unilateral linear ESFA is a nevoid condition.

Multiple authors have considered reactive ESFA to be a form of pathological hyperplasia (5, 6, 11); however, there is no preservation of the original sweat duct and glandular architecture, which should be present in hyperplastic conditions. Ohnishi et al. (15) had attempted to differentiate reactive ESFA from solitary ESFA immunohistologically, but did not find any significant differences. We consider reactive ESFA to be a form of hamartoma with a demarcated overgrowth of architecturally disorganized mature cells of eccrine ductal origin. Reactive ESFA may be more appropriately known as reactive eccrine syringofibroadenomatosis.

Bjarke et al. (13) studied five cases of non-reactive ESFA suspected to have undergone carcinomatous transformation. They commented that malignant potential is present in ESFA and that ESFA should be completely removed. Considering that there are about 55 cases of non-reactive forms of ESFA being reported, a reported incidence of eight cases of probable malignant transformation is high. We would agree with the approach of excising non-reactive forms of ESFA when feasible, particularly for the solitary forms. Although there have been no reports of carcinomatous transformation in reactive ESFA, it is also likely that this may occur, but with a lower risk compared with non-reactive ESFA. In our patient, considering that a flap or graft will be required following excision of the lesion on the bony part of the shin, we feel that observation and follow-up is a reasonable approach.

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