Ind J Dermatol Venereol Leprol 1993; 59 : 251-3

CASE REPORTS

ACQUIRED REACTIVE PERFORATING DERMATOSIS

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Introduction

The classical four transepithelial elimination (TEE) disorders include reactive perforating collegenosis (RPC), elastosis perforans serpiginosa (EPS), perforating folliculitis (PF) and Kyrle's diseases (KD). These four classical entities have some specific differentiantig features, like RPC is characterised by Koebner's phenomenon and elimination of collagen fibres. EPS is characterised by annular or serpiginous eruption, elimination of elastic fibres and increased elastic tissue in the dermis. PF is distinguished by its follicular origin and often presence of a coiled hair, while Kyrle's disease is less precisely defined. It has a greater mangitude of epithelial hyperplasia, sometimes with follicular involvement and may or may not exhibit a Koebner's phenomenon or elimination of elastic or collagen fibres. RPC and EPC have an inherited as well as an acquired form while PF and KD are in most cases acquired.

Besides these four conventional TEE disorders, transepidermal elimination of many diverse substances like products of keratinisation, connective tissue components, foreign bodies and micro-organisms have been reported in a wide variety of cutaneous disorders. There have been an increasing number of reports of perforating dermatoses in association with systemic diseases like diabetes mellitus, chornic renal failure, malignancies and liver disorders. It has also been observed that all the lesions may not show the same histological pattern and lesions representative of more than one type of TEE phenomenon may be present in the same patient. Besides, combined elimination of both elastic and collagen fibres has been reported.[1] Thus it was recommended that all these four entities be grouped together under the term'Acquired Perforating Dermatoses.[1] We are reporting a cases of ARPD resembling RPC more closely.

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Case Report

A 22-year-old male was admitted in Nephrology unit with high grade fever, cough with expectoration, pedal oedema, ascitis, epistaxis, haemoptysis, hematemesis and malaena. Investingation revealed anaemia (Hb: 3.2 gm%), high blood urea (205 gm%) and creatinine (11.8 gm%) and pneumonitis. He was diagnosed to have chronic renal failure and put on peritoneal dialysis. A week after the first dialysis sitting, he developed severely itchy, small papular eruption over dorsa of hands which gradually spread to forearm, arm, both lower limbs and abdomen. For these lesions he was referred to Dermatology department. There was no history of such lesions in childhood or in any other family members.

Cutaneous examination revealed bilaterally symmetrical, discrete, horny, papular lesions, 2-5 mm in diameter, predominantly around knuckles, elbows, thighs and lower abdomen. Some lesions were arranged in a linear fashion suggesting a positive Koebner's phenomenon. The lesions were severely itchy with excoriation marks. On expression of some of the lesions, a cheesy material expressed out.

Biopsy from the left arm revealed a crater in the centre of the lesion filled with parakeratotic keratin and neutrophil debris. The base of the crater showed break in the continuity of epidermis. Adjacent epidermis was thickened with compact hyperkeratosis. Strands of collagen were seen to project in the crater. Stains for elastic tissue were negative. Serial sectioning of the specimen did not establish continuity with hair follicle, thus confirming the diagnosis of RPC. Patient was given topical steroids and oral vitamin A preparations without much benefit.

Comments

A case of ARPD in association with chronic renal failure and dialysis is reported here. The classical morphology and histopathology of the lesion and demonstration of collagen fibres in the crater contributed to the diagnosis of RPC in the present case. ARPD especially those simulating KD and RPC have been reported in association with chronic renal failure.[1,2] In majority the lesions developed after dialysis, although they may develop before dialysis. It has been estimated that 5-10% of all patients undergoing heamodialysis develop lesions of APRD.[1] In the present case the lesions appeared following peritoneal dialysis.

The pathogenesis of TEE disorders is still under investigation. Inherited forms of RPC and EPS result from an altered response of dermal connective tissue to trauma in predisopsed individuals.[3,5] TEE disorders are named so, as the activity is mainly in the epidermis which grows and surrounds the entity to be eliminated. Elimination from epidermis then occurs spontaneously as epidermal maturation proceeds.[1] What triggres epidermal activity is under speculation. In RPC, histochemically altered collagen may be recognised as foreign resulting in its elimination. Prostaglandins may contribute to epidermel hyperplasia.[6] In ARPD, an overlap of the classical TEE disorders is seen. Hence a common denominator to the pathogenetic process must exist. Widespread metabolic changes found in diabetes mellitus, liver diseases and chronic renal failure might per say cause ARPD. Associated pruritus and xerosis on the other hand may be the necessary predisopsing factors.[7]

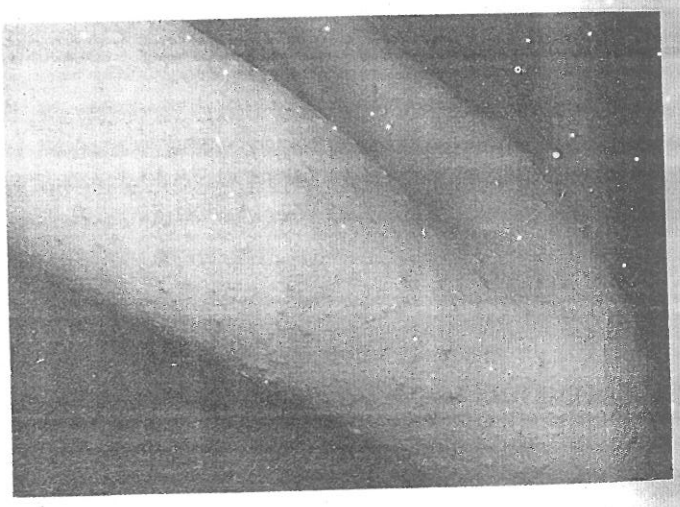

Fig. 1. Discrete, horny, papular lesions over back of upper arm and elbow

The various treatment modalities that have been used include topical corticosteroids and oral vitamin A preparations,[5] but were not found beneficial. H1 and H2 blockers and prostaglandin inhibitors were also not found useful.[6] A cases of RPC responded well to PUVA therapy and a direct effect of psoralens on fibroblasts and collagen was proposed as the possible mode of action.[8] Retinoids have been found useful in some cases of APRD. Both topically applied tretinoin and orally administered isotretinoin were effective. Retinoids probably act both on hyperplastic epidermis and dermal fibroblasts and collagen.[7] Further therapeutic trials are needed to establish proper guidelines of therapy.

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