

Extensive skin papular lesions

Wednesday, 02 November 2005

A 44-year-old white man presented with a 12-month history of slightly pruritic, papular lesions on his face, upper and lower extremities. No other complaints were mentioned. No similar skin dermatosis was reported from his family history including first-degree relatives. The patient had no history of kidney disease, diabetes mellitus or liver disease.

Physical examination was unremarkable except for the skin findings. Specifically, skin lesions were 8 in number located on the middle of the chin, on the right angle of the mouth and on the extensor surfaces of the forearms, elbows, thighs and knees. There was no mucous membrane involvement. Skin lesions were characterized by areas with diameter of 1 cm up to 8 cm consisting of multiple hyperkeratotic plaques and burrows. The large and old lesions of the extremities did not have signs of inflammation (Figure 1). In contrast, the small and new lesions on the face had clear signs of ongoing inflammation with an erythematous halo surrounding them (Figure 2). Mild tenderness and increased temperature was also evident on the face skin lesions.

Testing for anti-nuclear and anti-mitochondrial antibodies was negative, but it was weakly positive for anti-smooth muscle antibodies. There was also a slight elevation of the level of the haemolytic complement [CH50 = 556 U/ml (normal 300-510 U/l)]. The levels of the third and fourth component of complement (C3 and C4) were normal. Finally, a serologic test for syphilis [Venereal Disease Research laboratory (VDRL) slide test] was negative.

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Differential diagnosis Prurigo nodularis Multiple keratoacanthomas Verruca vulgaris Follicular atrophoderma Communicating fistulous sebaceous cysts Atypical mycobacterium infection Actinomycosis Perforating skin dermatoses

- Elastosis perforans serpiginosa
- Reactive perforating collagenosis
- Perforating folliculitis
- Kyrle's disease

Diagnosis

Kyrle's disease. Three hyperkeratotic skin lesions from the right lower extremity and one from the left forearm were surgically removed. Microscopic examination of the excised lesions showed invaginations of the epidermis in several points, hyperkeratosis of the epidermis and mild perivascular inflammatory infiltration of the dermis. Focal lichenoid type inflammatory infiltration was also found in the invaginations.

Treatment

In addition to the removal of the large hyperkeratotic skin lesions, which did not have findings of ongoing inflammation, the patient received treatment with clindamycin 300 mg, three times per day by mouth, for one month.

Teaching points

- Kyrle's disease is a hereditary skin disease (genodermatosis), although the mode of inheritance remains controversial (autosomal dominant or autosomal recessive).
- Kyrle's disease usually appears between 30 to 50 years of age. It predominately affects females with a 6:1 ratio compared to males. (1)
- Occurs commonly in the setting of diabetes mellitus, chronic renal failure, hepatic insufficiency or hyperlipoproteinemia and rarely may be a paraneoplastic disease. (2)
- Several hypotheses have been proposed about the aetiopathogenesis of Kyrle's disease. Diabetes mellitus related microangiopathy, microtrauma due to chronic pruritus and abnormalities of collagen, elastin and vitamin A or D metabolism in patients with renal disease are some of them. (3)
- The extracellular matrix proteins, particularly fibronectin, have been suggested to play a pathophysiological role in Kyrle's disease as well as in the other perforating dermatoses. (4)
- Management of patients with Kyrle's disease includes surgical removal of the lesions or destruction of them by cryo-, electro-, or CO2 laser surgery in addition to local and/or systemic treatment with vitamin A acid (retinoic acid). (5)

References

- Cunningham SR, Walsh M, Matthews R, Fulton R, Burrows D. Kyrle's disease. *J Am Acad Dermatol* 1987; 16:117-23.
- Harman M, Aytakin S, Akdeniz S, Deric M. Kyrle's disease in diabetes mellitus and chronic renal failure. *J Eur Acad Dermatol Veneteol* 1998; 11:87-8.
- Yanagihara M, Fujita T, Shirasaki A, et al. The pathogenesis of the transepithelial elimination of the collagen bundles in acquired reactive perforating collagenosis. *J Cutan Pathol* 1996; 23: 398-403.
- Morgan MB, Truitt CA, Taira J, Somach S, Pitha JV, Everett MA. Fibronectin and the extracellular matrix in the perforating disorders of the skin. *Am J Dermatopathol* 1998; 20:147-54.
- Baumer FE, Wolter M, Marsch WC. The Kyrle disease entity and its therapeutic modification by acitretin. *Z Hautkr* 1989; 64:286, 289-91.

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