Answer to "What is Your Diagnosis?" on p.198

Diagnosis: Adult colloid milium

Adult colloid milium (ACM) is a rare degenerative skin disorder characterized by asymptomatic, dome shaped, yellowish or skin-coloured, semitranslucent papules. The lesions are closely grouped on sun-exposed areas, sometimes producing a cobblestone appearance. Occasionally, small incision and pressure allows the expression of a gelatinous substance. Microscopic examination shows accumulation of dermal colloid material. The disorder is rare with approximately 100 published cases up to date. It is seen mostly in middle-aged, fair skinned people (1).

Adult colloid milium is caused by elastic fiber degeneration due to excessive sun exposure and petroleum products.

The pathogenesis is poorly understood, but it is believed to be due to excessive and chronic sun exposure causing elastin degeneration. In favor of this, the left side of the face and the left hand are usually more severely affected due to intensive sun exposure while driving, and the involved skin typically presents pronounced solar elastosis. Petroleum derivatives may have a role by enhancing the effects of UV light on the skin (1). Amyloid P is present in all types of amyloidoses in addition to normal or abnormal elastic fibers. Colloid substance shows positive immunostaining with amyloid P, too (2). It has been shown that components of colloid and elastic fibers have the same microfibrillar proteins. Both are rich in sulfur containing amino acids, but lack hydroxyproline, which is a major component of collagen (1). Our patient had clinical and histologic evidences of solar damage more severe than expected for his age. Because of the damage to the vascular connective tissue, skin fragility was prominent and the patient complained of delayed wound healing and purpura from trivial injuries.

The differential diagnoses of ACM mainly include other cutaneous deposition disorders.

Skin biopsy and histopathologic examination with special stains are necessary for differential diagnosis.

The first entity for differential diagnosis is amyloidosis. Traumatic purpura sometimes occurring in ACM may be an additional factor for misdiagnosis. The deposition of colloid materal within dermal blood vessels causes purpura that is analogous to purpura seen in systemic amyloidosis. The histopathologic distinction from amyloidosis can be very difficult since both conditions show fissured, homogenous eosinophilic material. Moreover, crystal violet, Congo red and PAS may be positive in both disorders (2). Two other stains, methyl violet and Pagoda red no 9, which are not routinely used in most of the laboratories, may help to differentiate these disorders, as they fail to stain colloid in contrast to amyloid (3). Electron microscopy is sometimes the only way for accurate diagnosis; amyloid shows straight filaments, whereas colloid has branching and wavy filaments (1). Lichen amyloidosis presents as intensely pruritic, redbrown hyperkeratotic papules most commonly

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seen on the pretibial surfaces, a clinical picture highly different from ACM (4). The clinical features of nodular amyloidosis with waxy, smooth papules, nodules or plaques on the face, trunk, or genitalia are also characteristic enough to distinguish it from ACM (2). Acral persistent papular mucinosis may be confused with ACM, because of the presence of multiple, skin coloured papules localized to the hands and wrists. Alcian blue staining is a definite marker for its diagnosis (5). Disorders of porphyrin metabolism may be considered in the differential diagnosis of ACM, due to the involvement of sun-exposed areas (1). However, in our patient lack of true blisters, milia, scarring, and the negative urine test in addition to positive staining with Congo red and amyloid P eliminated porphyria.

The lesions of ACM are static and do not resolve spontaneously. Treatment is unavailable since destructive therapies do not provide satisfactory results. In conclusion a good clinicopathologic correlation usually provide accurate diagnosis of ACM, avoiding unnecessary work-up for more serious metabolic disorders.

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