A Case of Idiopathic Calcinosis Universalis

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Accepted July 19, 2004.

Sir,
Calcinosis is the result of disorganized amorphous deposition of calcium and phosphate in organic matrices of the biological tissues with no tendency to normal bone formation. The process starts with deposition of a small focus of hydroxyapatite and amorphous calcium phosphate. Once formed, it increases in size by growth and may result in disorganized masses as nodules or plaques (1, 2). We report here on a patient affected by idiopathic calcinosis universalis.

CASE REPORT

A 43-year-old man exhibited many yellow-white nodules, symmetrically distributed over the backs of the hands and palms. They were hard, of different sizes and could become ulcerated, discharging chalk-like material (Fig. 1). The patient had no history of inflammatory disease, trauma or metabolic disorder. The dermatosis had begun 10 years earlier and was preceded by diffuse pains in the joints. Later on, nodules increased in size and number, causing an important limitation of movement due to stiffening of the skin and pain especially on finger joints. Some skin lesions became infected by *Staphylococcus aureus*, causing an abscess which required surgical drainage. Laboratory tests showed only mild decrease in neutrophils (38.9%) and lymphocytosis (51.6%), with no abnormalities in the calcium/phosphorus metabolism or parathyroid hormone levels. Erythrocyte sedimentation rate, C-reactive protein, complement and fibrinogen were within normal limits, as well as urine analysis. All

DOI: 10.1080/00015550410023491

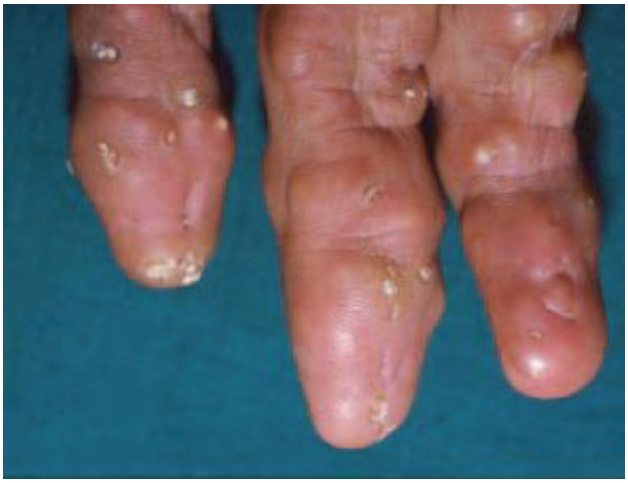

Fig. 1. Hard nodules of the fingers become ulcerated, discharging chalk-like material.

specific immunologic tests to evaluate autoimmune disease (organ- and non-organ-specific autoantibodies and complementary activity) or infections (VDRL, TPHA, HBV, HCV, CMV) were within normal limits. Tests for *Chlamydia pneumoniae*, trachomatis and psittacosis and common paraneoplastic markers were negative.

Radiographs and echography showed diffuse granular calcification in the soft tissue and muscle insertions of hands, feet, arms and legs. EMG of limbs showed polyneuropathy, probably due to compression from near calcifications. X-rays of the oesophagus and dental arches were normal. Histopathological examination of a biopsy of a nodule from the dorsal surface of the metacarpophalangeal junction in the second finger of the left hand showed widespread deposition of calcium salts in dermis and hypodermis.

DISCUSSION

Clinically, calcinosis cutis is characterized by small white papules, subcutaneous nodules or wide plaques, of stone-like consistency, symmetrically distributed to extremities and less often to the trunk. Lesions may become tender and ulcerated, discharging chalk-like material consisting mainly of calcium phosphate with a small amount of calcium carbonate. After ulceration, a slowly healing wound remains. Finger lesions are particularly painful, causing an important limitation of movement due to stiffening of tissues.

The pathogenesis of idiopathic calcinosis is unknown (4). This group includes calcinosis circumscripta (of scrotum, vulva, penis and serous burses) with calcification in localized areas of skin and subcutaneous tissues and calcinosis universalis involving skin, muscles, tendons and soft tissue (5, 6).

In our patient the presence of many deposits of calcium in dermis, hypodermis, soft tissues, muscles and tendons, unassociated with tissue damage or demonstrable metabolic disorder, led us to the diagnosis of idiopathic calcinosis universalis. This is an uncommon form because many cases reported in the literature under this diagnosis were later recognized as suffering from undetected dermatomyositis, systemic lupus erythematosus or scleroderma (4, 7). For this reason, our patient is periodically controlled with clinical evaluations and immunologic tests.

The therapy of calcinosis of the skin is still not very effective. Only one case has been reported with a complete resolution over a period of 1 year with continued steroid therapy (8). Treatment trials have included biphosphonate, etidronate disodium, aluminium hydroxide, warfarin, probenecid, intralesional injections of corticosteroids and finally surgical excision and laser therapy (7, 9).

REFERENCES

1. Walsh JS, Fairley JA. Calcifyng disorders of the skin. J Am Acad Dermatol 1995; 33: 693–706.
2. Chaves Alvarez AJ, Herrera Saval A, Marquez Enriquez J. Metastatic calcinosis cutis in multiple myeloma. Br J Dermatol 2000; 142: 820–822.
3. Cambiaghi S, Imondi D, Gangi S, Vegni C. Fingertip calcinosis cutis. Cutis 2000; 66: 465–467.
4. Bhatia S, Silverberg NB, Don PC, Weinberg JM. Extensive calcinosis cutis in association with systemic lupus erythematosus. Acta Derm Venereol 2001; 81: 446–447.
5. Oh CK, Kwon KS, Cho SH, Jang HS. Idiopathic calcinosis of the areola of the nipple. J Dermatol 2000; 27: 121–122.
6. Cecchi R, Giomi A. Idiopathic calcinosis cutis of the penis. Dermatology 1999; 198: 174–175.
7. Eddy MC, Leelawattana R, McAlister WH, White MP. Calcinosis universalis complicating juvenile dermatomyositis: resolution during probenecid therapy. J Clin Endocrinol Metab 1997; 82: 3536–3542.
8. Weinberger A, Kaplan JG, Myers AR. Extensive soft tissue calcification (calcinosis universalis) in systemic lupus erythematosus. Ann Rheum Dis 1979; 38: 384–386.
9. Matsuoka Y, Miyajima S, Okada N. A case of calcinosis universalis successfully treated with low-dose warfarin. J Dermatol 1998; 25: 716–720.