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Case Report

Idiopathic Universal Calcinosis Cutis: Case Report

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Abstract

Observation: Calcinosis cutis is a rare disease in which insolubl calcium and phosphate salts are abnormally deposited in the skin. We report a 21-year-old girl who has had multiple firm nodules on her body mainly around her large joints since she was 2 years old. Her laboratory studies were normal. X-ray studies showed round-shaped calcifications in soft tissues but there was no bone pathology. Histopathological findings showed calcinosis in subcutaneous tissue. On the basis of clinical, laboratory and histological data, the diagnosis of idiopathic universal calcinosis cutis was made.

Introduction

Calcinosis cutis is a metabolic disease in which insoluble calcium salts are abnormally deposited in the skin [1]. Calcinosis cutis is classified into four major subcategories according to their etiology: dystrophic, metastatic, idiopathic, and iatrogenic [2].The most frequent type is dystrophic type which occurs in the presence of normal plasma levels of calcium and phosphate in affected skin. Dystrophic calcinosis is common in connective tissue diseases [3, 4]. Metastatic calcinosis occurs in the presence of primary disorders of calcium-phosphate metabolism in normal tissues [3]. Idiopathic calcification occurs in the absence of evident tissue or metabolic abnormalities [5]. Iatrogenic calcinosis can occur as a complication of intravenous calcium chlorid and phosphate therapy when extravasation occurs as well as with calcium salt exposure from electromyography (EMG) and electroencephalography (EEG) electrode compounds [4, 5, 6].

Case Report

A 21-year-old female was admitted to our hospital with firm nodules especially around her large joints. The nodules first appeared on her knee joints when the girl was 2 years old.

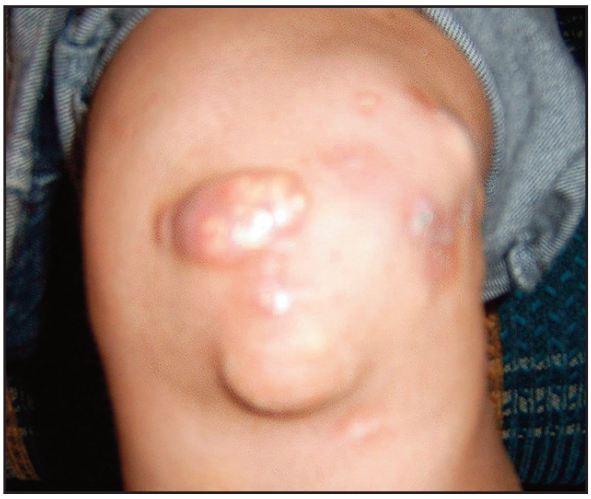

Figure 1. Cream-violet coloured, firm, roundish shaped, immobile, asymptomatic nodules around the patient's knees

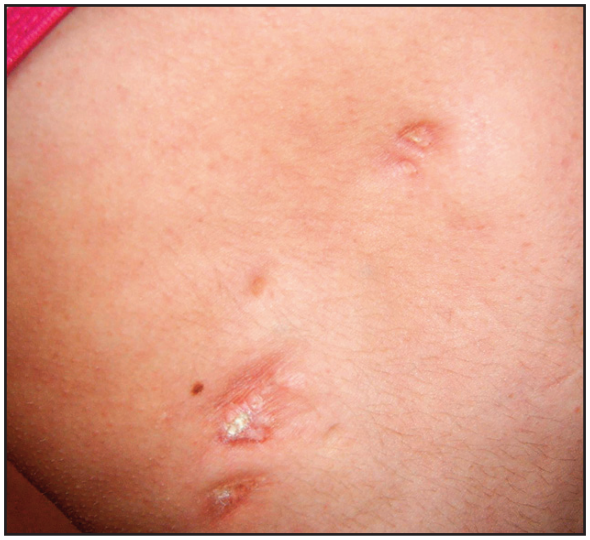

Figure 2. Scar tissue on pre-existing ulcerous nodules on the right upper thigh

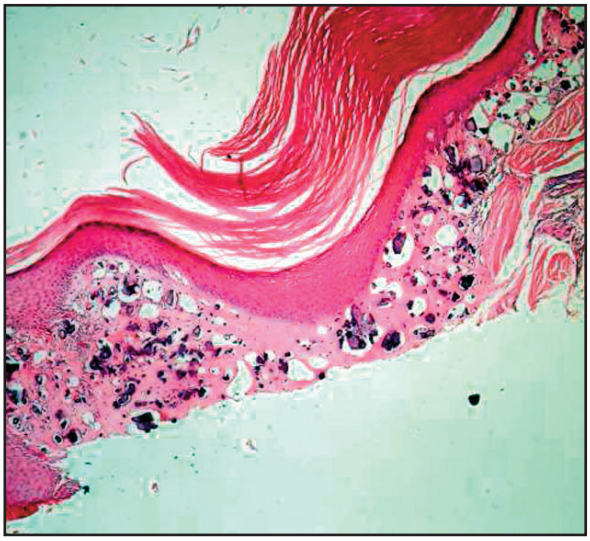

Figure 3. Hyperplasia in the epidermis and calcium deposits in the dermis and subcutaneous tissue (H&Ex10)

After a while nodules enlarged and other joints were affected. From time to time lesions became painful ulcers and a creamy material drained from these ulcerous lesions.Some were comprised of scar tissue. In her family history there were no patients with similar complaints.

On physical examination she had multiple cream-violet coloured, firm, roundish shaped, immobil, asymptomatic nodules approximately 2-3 cm in diameter around her knees, elbows and hip joints (**Figure 1**). Plaques which consisted of these nodules were seen on the pelvic area of the patient.

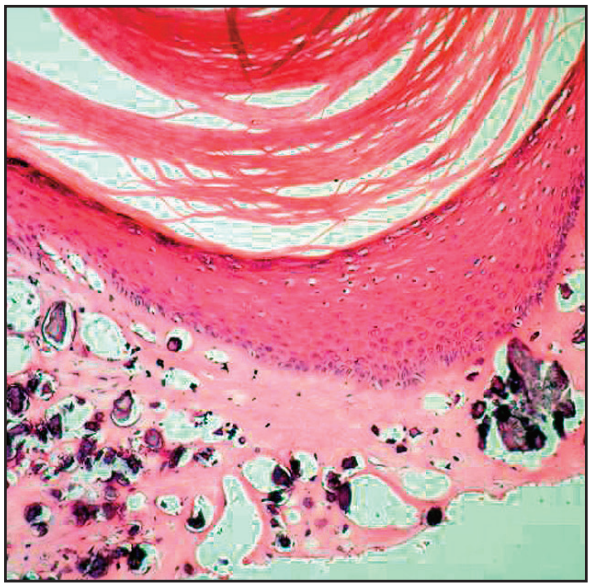

Figure 4. Hyperkeratosis, acanthosis, hyperplasia in the epidermis and calcium deposits in the dermis and subcutaneous tissue (H&Ex40)

There were scar tissue on pre-existing ulcerous nodules (**Figure 2**).

A punch skin biopsy was performed from a nodule on the knee. Histopathological examination revealed hyperkeratosis, acanthosis, hyperplasia in the epidermis and calcium deposits in the dermis and subcutaneous tissue (**Figure 3, 4**).

Routine laboratory tests and metabolic evaluation (calcium, phosphate and parathyroid hormone) were within normal limits. Calcium and phosphate excretion in 24 hour urine was normal. Plasma osteocalcin, vitamin D3, pancreatic amilase and lipase levels were within normal limits. Clinical observation and screening tests for collagen vascular disease were normal. There was no pathology in bone syntigraphy and abdominal ultrasonography.

Radiological examination showed that there was no bone pathology but widespread calcification in the soft tissues.

On the basis of the clinical, radiological, laboratory and histopathological data the diagnosis of calcinosis cutis universalis was made.

Discussion

Calcinosis cutis is a rare metabolical disease that is characterized by abnormal deposits of calcium salts in the dermis or hypodermis or in both. Based on pathophysiologic mechanism, it has been classified as metastatic, dystrophic, idiopathic and iatrogenic [**5, 7**].

Metastatic calcinosis occurs in undamaged tissues and is associated with elevated serum phosphate or calcium levels or both. Renal failure, paraneoplastic hypercalcemia, hypervitaminosis D, hyperparathyroidism, milk alkali syndrome, sarcoidosis and destructive bone disease may develop metastatic deposits of calcium [**3,5,8**]. In our patient plasma calcium, phosphate, vitamin D3, parathormone and osteocalcin levels were normal, there was no pathology in calcium/phosphate metabolism. So metastatic calcinosis was eliminated.

Dystrophic calcinosis is localized deposition of calcium salts in dead or degenerated tissues amongst individuals in the presence of normal calcium and phosphate plasma levels [**1**]. Dystrophic calcinosis is common in childhood dermatomyositis, systemic scleroderma, CREST syndrome, morphea, subcutaneous fat necrosis of the newborn, cutaneous neoplasms (pilomatrixoma, basal cell epithelioma, epidermal cysts and melanoma), Ehler- Danlos syndrome and pseudoxantoma elasticum. Cutaneous calcification resulting from abdominal surgery, chronic ulcers and burn scars have also been reported [**3, 5**]. In our patient there were no tissue damage, infection and connective tissue disease, so we eliminated dystrophic calcinosis cutis.

Iatrogenic calcinosis can occur as a complication of intravenous calcium chloride and phosphate therapy as well as with calcium salt exposure from EMG and EEG electrode compounds [**4, 5, 6**]. Our patient's history and clinical findings were not consistent with iatrogenic calcinosis.

Idiopathic calcinosis cutis occurs in the absence of evident tissue or metabolic abnormalities. Serum calcium and phosphorus levels are within normal limits and there is no underlying cutaneous or systemic disease. Idiopathic calcification includes three distinctive disorders of childhood; milia like calcinosis cutis associated with *Down* syndrome (MICC), idiopathic calcinosis of the scrotum and subepidermal calcified nodule of *Winer* (SCN) [**9, 10, 11**]. MICC is a rare benign disorder and the majority of the cases are associated with Down syndrome. Clinically the lesions appear as smooth, firm, whitish papules resembling milia and usually disappear before adulthood. The disease affects primarily the hands, the feet and rarely the face [**10**]. SCN most occurs in children. It is characterized by a solitary or rarely multiple, small, firm, well-circumscribed, slightly elevated, white-yellow nodule; most frequently localized on the face. The treatment of choice is surgical excision [**11**].

We classified our patient as having idiopathic universal calcinosis cutis because there was no underlying cutaneous or systemic disease; serum calcium and phosphate levels were within normal limits and she had multiple lesions widespread on her body since childhood. The pathophysiology of this condition remains unclear and no effective therapy is currently available. Our patient was sent to plastic surgery for excision of the lesions that discomfort her and for reconstruction of the deformations caused by existing scars.

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