Calcinosis Circumscripta of the Hand in Scleroderma

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ABSTRACT: In a series of sixty-five patients with scleroderma, eleven had calcinosis circumscripta. Four other patients in the series had calcifications in the soft tissues elsewhere than in the hand. Nine of the eleven patients were women. The diagnosis of calcinosis in the hands was made on the average of seventeen years after the onset of Raynaud’s phenomenon. In only two of the patients was surgical excision of the calcifications performed.

Calcinosis circumscripta is characterized by localized calcific deposits in the skin and subcutaneous tissues particularly of the hands and wrists. In 40 to 60 per cent of cases, it is associated with collagen disease, especially scleroderma 27. At times, no underlying disease can be found. Our interest in this disease started with a patient who appeared with localized calcification in a finger. Ten additional cases of calcinosis circumscripta in the hand were found by reviewing the charts of sixty-five patients discharged from University Hospitals of Cleveland during the past ten years with the diagnosis of scleroderma.

Case 1. A fifty-five-year-old black woman, a housekeeper, was admitted for evaluation of progressive enlargement and loss of motion of the left index finger of two years’ duration. On physical examination, the left index finger was enlarged in diameter and very firm to palpation (Fig. 1). Although the skin over this finger lacked pliability, it was not shiny, depigmented or ulcerated. Flexion at the interphalangeal joints was limited so that the tip of the left index finger approached the palm no closer than five centimeters. A roentgenogram of the left index

Fig. 1

Dorsal surface of the left hand showing enlargement of the index finger.

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finger revealed many nodular calcifications in the soft tissues over the radial and palmar aspects of the proximal and distal phalanges. Resorption of the tuft of the distal phalanx was present (Fig. 2). Clinically no other abnormalities of the hand were noted. Laboratory studies showed: hematocrit, 45 per cent; white blood cell count, 4,500; sedimentation rate, 26; uric acid, 6.5; calcium ranged between 10.1 and 10.3; phosphate ranged between 3.7 and 5.2 milligrams per 100 milliliters; alkaline phosphatase, normal; blood urea nitrogen, 10; creatinine, 0.8; and antinuclear factor and latex fixation, negative.

![Fig. 2](image)

Roentgenograms of the left index finger. Conglomerate calcific densities are visualized over the radial and palmar aspects.

No definite diagnosis was made and surgical exploration was undertaken. Under tourniquet ischemia, a longitudinal incision was made over the radial side of the index finger. A pastelike grayish yellow material was found just beneath the epidermis which extended throughout the subcutaneous tissues, and surrounded but did not involve the tendon sheaths. Much of this paste-like material and associated mineralized fibrous tissue was excised from the volar aspect of the finger while preserving the digital nerves and vessels. Microscopic and chemical examination of the excised material revealed sparsely cellular poorly vascularized connective tissue containing deposits of calcium and phosphate. A definite diagnosis could not be made on the basis of the biopsy. More clinical information was needed.

On careful questioning the patient admitted to having vasospastic symptoms consistent with Raynaud's phenomenon about four years prior to admission. She was left-handed, and the Raynaud's phenomenon had always been worse on the left side. Three and one-half years prior to admission, pain and swelling developed in both hands and wrists which subsided over the next three months. The increase in the size of the left index finger started two years prior to admission and seemed to be unrelated to the past history of Raynaud's phenomenon.

Closer physical examination revealed scattered pointlike and polyangular telangiectasia over the palms, volar aspects of the fingers, lips, buccal and oropharyngeal mucosa (Fig. 3). Except for the loss of pliability of the skin of the distal portion of the left index finger, no ab
es over the radial and palmar aspects of the distal phalanx was present. Laboratory studies showed: hematocrit, 26; uric acid, 6.5; between 3.7 and 5.2 milligrams per nitrogen, 10; creatinine, 0.8; and

The typical telangiectasia associated with the calcinosis cutis. Raynaud's phenomenon, scierodactyly and telangiectasia syndrome are seen.

ormalities in skin texture or pigmentation were noted. Roentgenograms of both hands were taken and small foci of calcification were found in the soft tissues around the distal phalanges of both thumbs and the right index finger. In addition, roentgenographic evidence of calcification of the soft tissues in the left thigh was found. The esophagogram and intravenous pyelogram were normal. On reviewing the specimen excised from the left finger, increased collagen tissue consistent with scleroderma was found.

After surgery, some marginal necrosis of the skin flaps of the left index finger developed. A free full thickness skin graft was applied to this area. One year after the first operation the function of the left index finger was improved. Some limitation of flexion of the interphalangeal joints persisted such that the tip of the index finger barely touched the palm.

As a result of the more complete history and physical examination, the diagnosis of calcinosis cutis, Raynaud's phenomenon, sclerodactyly and telangiectasia syndrome was made. Winterbauer 32 who first described the calcinosis cutis, Raynaud's phenomenon, sclerodactyly and telangiectasia syndrome indicated that it was closely related to scleroderma. Our case emphasizes the importance of recognizing telangiectasia as a cutaneous manifestation of scleroderma. Braverman 5 considered that telangiectasia was as important as cutaneous sclerosis in making the diagnosis of scleroderma and was not only characteristic of the calcinosis cutis. Raynaud's phenomenon, sclerodactyly and telangiectasia syndrome but was also seen along with other cutaneous manifestations in the more diffuse forms of scleroderma (progressive systemic sclerosis). Telangiectases, indistinguishable from those seen in scleroderma, can be found in Osler-Weber-Rendu disease 30,32 and dermatomyositis 8.

Among our sixty-six cases of scleroderma, fifteen had calcifications in soft tissues and eleven had calcinosis in the hand. Forty-six patients were women and twenty were men (2.3 to 1 ratio). Among those with ectopic calcifications, thirteen were women and two, men.

In six of the eleven patients with calcinosis in the hands, all four manifestations of the calcinosis cutis, Raynaud's phenomenon, sclerodactyly and telangiectasia syndrome were present. In three patients, the telangiectases were not present, and in two, both the Raynaud's phenomenon and telangiectases were absent. Nine of the eleven were women, and the average age was fifty-four years. The diagnosis of calcinosis in the hands was made on the average of seventeen years after the onset of Raynaud's phenomenon.

CASE 2. This patient appeared with pain and swelling of the distal end of the left long finger of two weeks' duration. A deposit of calcium was visualized roentgenographically in the
pulp space of this finger. It was excised through a small incision over the volar aspect of the distal part of the finger. The incision healed well and no further symptoms developed during a ten-month follow-up period. In this case, the diagnosis of scleroderma was well established on the basis of cutaneous manifestations and visceral involvement.

Surgical excision of the calcifications in the hand was performed in two patients. In both, the function of the hand was improved. In nine patients cutaneous sclerosis was a prominent feature and none of them required excision of the deposits of calcium. In several patients, periodic spontaneous ulceration of the overlying skin with extrusion of the calcified material occurred, which was often associated with surprisingly little disability. In some patients, the calcification was asymptomatic and was found incidentally on roentgenograms of the hands.

The roentgenographic picture of calcification in the soft tissues of the hand varied from powdery punctate densities to large discrete densities. As the tissues became more involved by calcification, the densities became confluent. In general, the calcification was more extensive over the palmar and radial aspects of the distal phalanx, that is, over pressure-bearing areas. Resorption of the tuft of the distal phalanx was characteristic of the scleroderma. This occurred in seven of the eleven patients with calcinosis in the hands.

Although scleroderma can affect joints producing a synovitis and resulting in narrowing of the joint spaces, seldom are there deformating changes as in rheumatoid arthritis. Rheumatoid arthritis is related to scleroderma and the two diseases can occur in the same individual. A deforming rheumatoid arthritis of the hands was associated with scleroderma in two of the eleven patients with calcinosis in the hands.

Discussion

Thibierge and Weissenbach, in 1911, firmly established the association of calcinosis circumscripta with scleroderma, and, in 1959, in a study by Muller and associates, 86 per cent of the cases of calcinosis reported involved the hands. In our study, of the 23 per cent of patients with calcinosis, 73 per cent had it in the hands. Since calcinosis of the hands is relatively frequent in scleroderma, roentgenograms of the hand are recommended routinely in patients in whom this diagnosis is considered.

In our patient the severe calcinosis in the hand was associated with minimum cutaneous sclerosis. Calcinosis usually is found in patients with severe sclerodactyly. Muller and associates found a 27 per cent prevalence of calcinosis in patients with severe sclerodactyly and a 4 per cent prevalence in those with moderate sclerodactyly. At one time, all the changes in scleroderma were thought to be secondary to tightening of the skin.

Other diseases which can be associated with calcinosis circumscripta include dermatomyositis, rheumatoid arthritis, lupus erythematosus, and acrodermatitis atrophicans. Dermatomyositis, especially in children, can cause widespread calcification which involves muscles as well as subcutaneous tissues. Considerable disability can result from the extrusion of these deep-seated deposits. Appearance of the calcifications has been reported in dermatomyositis but not in scleroderma.

The mechanism for calcinosis circumscripta in scleroderma and other collagen diseases is poorly understood. Local tissue factors are probably more important than any systemic derangement in the calcium or phosphorus balance. The possibility that some form of hyperparathyroidism contributes to the calcinosis has been studied but no improvement in the calcinosis has been reported in patients with scleroderma who underwent parathyroidectomy even if there was chemical improvement.
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Calcification over the volar aspect of the thenar symptoms developed during scleroderma was well established on 1 and was performed in two phases. In nine patients cutaneous excision of the deposits ulceration of the overlying skin, which was often associated with, the calcification was asymptomatic of the hands.

In the soft tissues of the hands, there were no discrete densities. As the tissues became confluent. In general, the radial aspects of the duration of the tuft of the digit. This occurred in seven of the fingers. A synovitis and resulting a firming change as in rheumatoid scleroderma and the two diseases rheumatoid arthritis of the hands. In patients with calcinosis in the established the association of calcinosis, in a study by Muller and reported involved the hands. In calcinosis, 73 per cent had it in the one or the other. The patients with scleroderma, roentgenographically in whom this diagnosis is associated with minimum symptoms with severe sclerodactyly, 15% of calcification in patients with calcinosis. These patients with moderate sclerodactyly, ought to be secondary to tightening.

calcinosis circumscripta includes pustular emphysema and, although in children, can cause widespread subcutaneous tissues. Considerable deep-seated deposits. Disappearance in dermatomyositis but not in scleroderma and other collagenoses is probably more important than in ankylosing spondylitis. The possibility to the calcinosis has been studied in patients with scleroderma as chemical improvement.

References


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