Cutaneous Lesions in Acroosteolysis

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A patient with idiopathic acroosteolysis had unique papular skin lesions. To our knowledge, similar lesions have been previously mentioned only in connection with acroosteolysis seen in vinyl chloride processing, and a detailed description of these has not been noted in the literature.

The term acroosteolysis refers to lytic changes of the shafts of the distal phalanges, with preservation of the tufts and bases. In most cases, other bones are also involved. Three types have been described in the literature: (1) familial,[1,2] (2) idiopathic or nonfamilial,[3-6] and (3) those associated with vinyl chloride processing.[7,8]

We are describing a case of idiopathic acroosteolysis, manifesting unique skin lesions. Similar lesions have been described only briefly in some cases associated with industrial vinyl chloride processing. To our knowledge, these cutaneous findings have not been discussed in the dermatologic literature.

Report of a Case

In August 1969, a 36-year-old white woman was examined at US Walson Army Hospital. Her condition had apparently started in 1960. During cold weather, she began to develop symptoms

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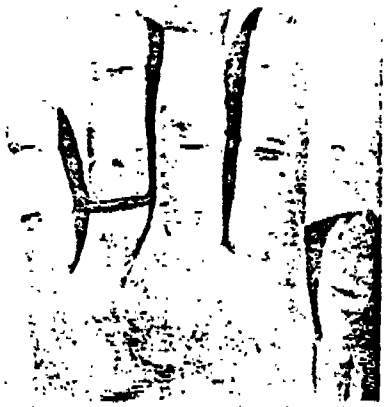
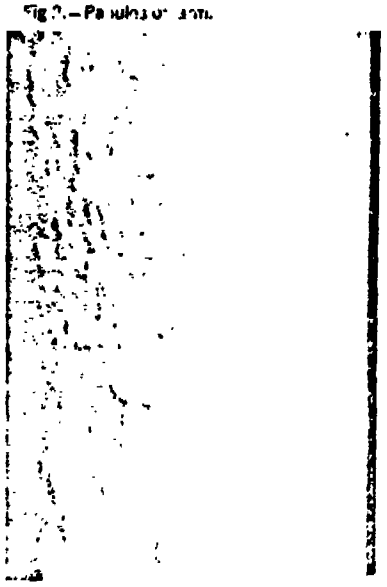


Fig 1.—Note short, thick fingertips and nail changes.

Fig 2.—Papules on arm.



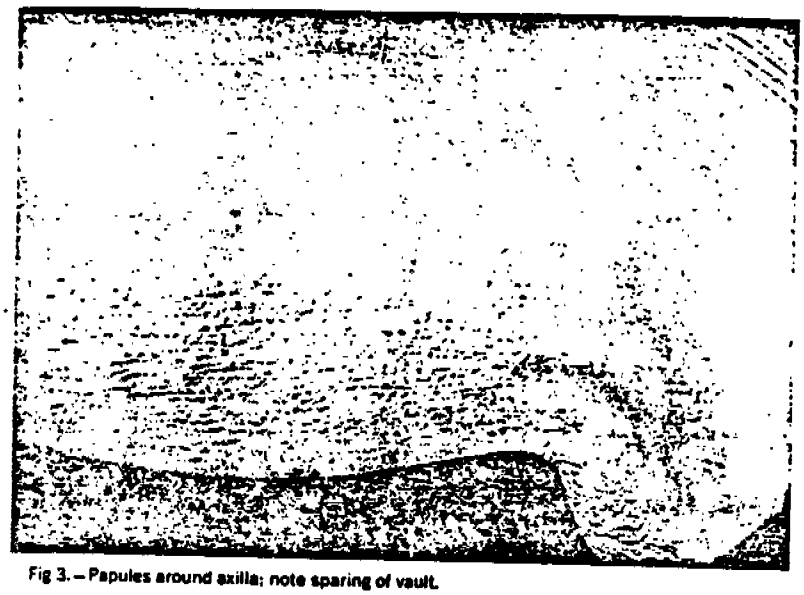


Fig 3.—Papules around axilla; note sparing of vault.

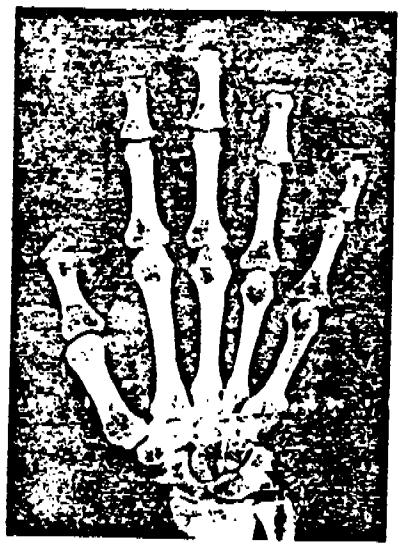


Fig 4.—X-ray film of right hand showing acroosteolysis.

Fig 5.—Thickened dermis with enlarged collagen fibers (separation of fibers is an artifact). Note preservation of eccrine sweat gland and duct (hematoxylin-eosin, ×40).

Fig 6.—Increase in elastic fibers, with fraying and fragmentation (elastic stain, ×400).

typical of Raynaud's phenomenon. Early in 1961, the right fifth finger became painful, slowly shortened, and the fingernail became shorter and thicker. At the same time, cutaneous lesions appeared on both wrists. By May 1961, most of the fingers had become similarly involved, and the cutaneous lesions spread to involve the arms, axillae, neck and trunk. The patient was hospitalized at that time, and x-ray films of the hands showed destruction of all distal phalanges, with preservation of the tufts. A diagnosis of scleroderma was made.

Despite the administration of prednisone from 1961 to 1966, the changes in the distal fingers and nails gradually progressed, and the skin lesions enlarged and spread. The findings of a skin biopsy obtained in 1965 were reported to be consistent with the diagnosis of scleroderma, and the patient was placed on a regimen of potassium aminobenzoate with no effect. From 1966 on, the patient developed progressive difficulty in the use of her jaw, with pain involving the temporomandibular joints, leading to progressive malocclusion of the teeth. X-ray films of the jaws in 1968 revealed osteolysis of the articular surfaces of the mandible. This condition progressed, and in 1970 she developed a pathologic fracture through the neck of the right mandibular condyle. Therapy for this aspect of her condition was conservative with the use of various dental appliances for support and partial correction.

In 1968, vitamin E was prescribed, and in the patient's opinion, the skin lesions decreased somewhat in size. However, no improvement was noted in the other symptoms or on subsequent x-ray films. The patient denied any sensory changes in the hands or feet. She was never employed in the processing of vinyl chloride or any other chemical. She primarily worked in offices of various financial institutions.

There is no family history of a similar condition. Her father, aged 68, has arthritis and a "straight spine," and her mother, aged 69, has rheumatoid arthritis. Her maternal great-great-grandfathers were closely related, but it is not certain whether they were brothers or first cousins.

Physical examination revealed shortening of all the distal phalanges of the hands and short, thickened fingernails (Fig 1). Similar changes were noted on the fourth toe of the right foot. The fingertips were broad with no tapering or ulcerations. Skin of the hands and arms was somewhat tighter than normal, but there appeared to be normal range of motion in the joints. The facial skin similarly was tighter than normal.

The most striking abnormality of the skin was the presence of multiple yellowish, cutaneous papules, 2 to 4 mm in size, appearing in linear distribution or in patches, and in areas becoming confluent. The most involved area was the skin of the forearms, primarily the flexor surfaces. Here, the papules were large and tended to form linear bands extending up the arms (Fig 2). Similar, though less prominent, papules surrounded both axillae, but spared the vaults (Fig 3). Patches of small papules were also found on the anterior and posterior aspects of the neck, the upper parts of the chest and back, beneath both breasts, around the waist, and the upper thighs. No involvement of the face, lower part of the back, abdomen or lower part of the legs was noted.

The patient was seen in consultation by an oral surgeon who noted an inability of the mandible to return into a normocentric, occlusal relationship with the maxilla. Due to this malocclusion, the brunt of the occlusal forces was borne by the posterior teeth which resulted in an anterior open bite.

The remainder of the physical examination showed no abnormalities.

The results of the following laboratory studies showed values within normal limits: complete blood cell count, sedimentation rate, VDRL test for syphilis, blood glucose, bilirubin, total protein and protein electrophoresis, calcium, phosphorus, alkaline phosphatase, uric acid, serum glutamic oxaloacetic transaminase, serum glutamic pyruvic transaminase, lactic dehydrogenase, blood urea nitrogen, rheumatoid arthritis latex fixation, cryoglobulins, antinuclear antibodies, LE preparation, and urinalysis.

X-ray films showed acroosteolysis of all distal phalanges of the hands (Fig 4) and of the right fourth and fifth and the left second toes. X-ray films of the mandible showed a marked resorptive process of the articular surfaces of the condyles and a complete pathologic fracture through the neck of the right condyle. X-ray films of the chest, skull, spine, and long bones were all normal.

Pathological Findings.—Biopsies taken from the right axilla and the right wrist revealed identical findings. Histologic examination showed flattening of the epidermis with loss of the rete ridges. The dermis was thickened with enlarged collagen fibers and decreased cellularity (Fig 5). The adnexal structures appeared intact, and there was no inflammatory reaction. An elastic tissue stain showed an increase in the number of elastic fibers with fraying and fragmentation (Fig 6). The results of stain tests for mucopolysaccharides were negative.

Comment

Familial acroosteolysis[1,2] is probably autosomal dominant and occurs more commonly in males. The disease usually begins in early childhood or adolescence with a slightly painful ulceration on the sole of the foot. The ulcer slowly grows over several weeks or months, discharges bony fragments, and then heals. The process continues with recurrent episodes associated with slight fever. The feet gradually become deformed, swollen, and mutilated. Sensory disturbances of both the upper and lower extremities are always associated. X-ray films show osteomyelitic changes in the areas of ulceration as well as acroosteolysis of the phalanges.

Idiopathic, nonfamilial acroosteolysis[3-6] usually starts in early adulthood. The hands are more severely involved than the feet, and there is no associated neurologic abnormality. Raynaud's phenomenon has been reported in some cases. No ulcerations appear, and no consistent, cutaneous lesions have been described. Radiologic findings include acroosteolysis of the phalanges, defects of the metatarsal-phalangeal joints, and resorption of the alveolar process in the jaw. Changes have also been reported in the skull, spine, and other bones.

Classification of these first two types is by no means consistent or totally accepted in the literature. Some reported cases of acroosteolysis have been secondary to osteomalacia[9,10] or hyperparathyroidism.[11] Giaccai[2] mentioned a sporadic "nonfamilial type of neurogenic acroosteolysis." Cheney[12] described a family in which several members had features of both familial and idiopathic acroosteolysis.

A new type of acroosteolysis was first reported in the English literature by Wilson et al[7] in 1967. They described 31 cases involving the hands of workmen engaged in vinyl chloride polymerization. (Vinyl chloride is made into polyvinyl chloride, a widely used synthetic resin.) The

ndition is apparently specifically ociated with the hand cleaning of lymerizers.[7-13] A survey of more an 1,000 persons who handled the nished resin, or processed it into stic products, failed to find any ditional cases.[7]

The great majority of patients described by Wilson et al had Raynaud's phenomenon and all had acroosteolysis of the distal phalanges of the hands. According to a written communication from R. H. Wilson, M.D. on Jan 21, 1970, three or four of their 31 cases "had external skin lesions on the dorsal surfaces of the hands and forearms, with a ropelike appearance resembling changes sometimes seen in scleroderma."[7] Harris and Adams[8] described two cases associated with vinyl chloride processing. One had "puffiness of the face and thickening of the skin of the hands, fingers, and left forearm," and the condition had previously been diagnosed as scleroderma. The other patient had "raised nodules in the skin, described as 'xanthomatous-like' patches" around his wrists.

The skin lesions described in these patients are almost identical with those found in our case. Furthermore, the histopathologic findings reported by Harris and Adams are essentially the same as in our patient. We reviewed the slides from two cases of Wilson et al and found these too to have the same histopathologic evidence. All had thickened collagen fibers and fragmented elastic fibers in the dermis. The adnexae were normal, and there was no significant inflammation present.

Because of the similarities to our patient and because we could not find such skin lesions described in other types of acroosteolysis, we carefully questioned our patient about any possible contact with vinyl chloride processing. She denied any such contact and has never been employed in any type of chemical factory. Thus, our patient apparently represents the first reported case of idiopathic acroosteolysis to manifest these peculiar skin lesions.

The cause of this disease is obscure. Some of the industrial cases were initially thought to represent atypical scleroderma,[8-14] and our patient had this diagnosis for some time. However, the skin lesions do not really resemble scleroderma, and the histopathological findings are also different. Furthermore, the x-ray film changes in scleroderma almost always show destruction of the phalangeal tufts, and not acroosteolysis, which has relative sparing of the tufts.

Only a small fraction of workers engaged in cleaning polymerizers develop the disease. Therefore, there must be some predisposition in these people which is brought out by contact with certain chemicals. Similarly, our patient may have had such a predisposition and had her disease triggered by some unknown factor or factors.

John Creech, MD, lent us the pathological slides from the cases of Wilson et al.

Louis J. Brunelle made the photographs and photomicrographs.

Nonproprietary and Trade Names of Drug

Potassium aminobenzoate—*Potaba*.

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