Skeletal Radiol (1987) 16:312–315

Skeletal Radiology

Distal phalangeal brachydactyly secondary to healed renal osteodystrophy

Andrew C. Wu, M.D., and Louis A. Gilula, M.D.
Mallinckrodt Institute of Radiology, Washington University School of Medicine, St. Louis, Missouri, USA

Abstract. Six cases of distal phalangeal brachydactyly of the hands in patients with healed renal osteodystrophy are reported. Severe osseous changes of renal osteodystrophy were seen in all cases. These cases present healed renal osteodystrophy as another consideration in the differential diagnosis of distal phalangeal brachydactyly.

Key words: Brachydactyly phalangeal – Hyperparathyroidism – Renal osteodystrophy, healed

Renal osteodystrophy is characterized by excessive bone resorption and abnormal bone formation. In the hands, typical changes are cortical, subperiosteal, and subchondral bone resorption. Subperiosteal resorption along the radial aspect of the middle phalanges of the hand has been recognized as a sensitive radiographic sign for hyperparathyroidism [7]. Many authors have described phalangeal tuft resorption, which reflects substitution of poorly mineralized fibrous bone for lamellar bone [8], as an early and sensitive indicator of hyperparathyroidism [4, 10].

Numerous studies have documented the reversal of the skeletal changes of renal osteodystrophy following hemodialysis and/or parathyroidectomy [2, 3, 9, 11, 12]. In the hands, osseous improvement after appropriate therapy allows the restoration of a nearly normal appearance of the phalanges (Fig. 1 A, B).

Because of our clinical realization that distal phalangeal brachydactyly could follow treatment

Address reprint requests to: Louis A. Gilula, M.D. Mallinckrodt Institute of Radiology, Washington University School of Medicine, 510 South Kingshighway Boulevard, St. Louis, MO 63110, USA

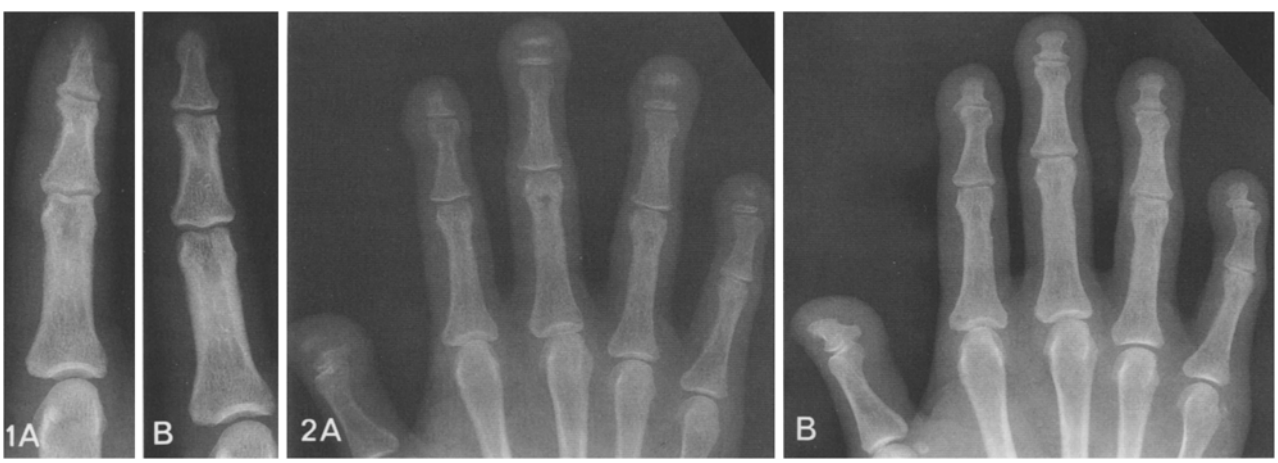

Fig. 1 A, B. Twenty-eight-year-old man with renal osteodystrophy. **A** Phalangeal tuft resorption is present and subperiosteal resorption is seen along the radial aspect of the proximal and middle phalanges of the right index finger. **B** One year after parathyroidectomy, remineralization of the phalanges has occurred with restoration of normal-appearing phalanges

Fig. 2 A, B. Twenty-nine-year-old woman with severe renal osteodystrophy. **A** Extensive cortical and subperiosteal resorption associated with acro-osteolysis of distal phalanges of right hand. **B** Fourteen months after parathyroidectomy, healing has resulted in distal phalangeal brachydactyly. Also the thumb has healed with a "beaked" configuration

Table 1.

	PTH (units/ml) Pre-operation ($n=2$–10)	PTH (units/ml) Immediate post-operation	Number of parathyroid glands removed at surgery	PTH (units/ml) follow-up
Case 1	85	2.3	3-1/2	19–25 (8 years)
Case 2	415	25	3-3/4	24–29 (4 years)

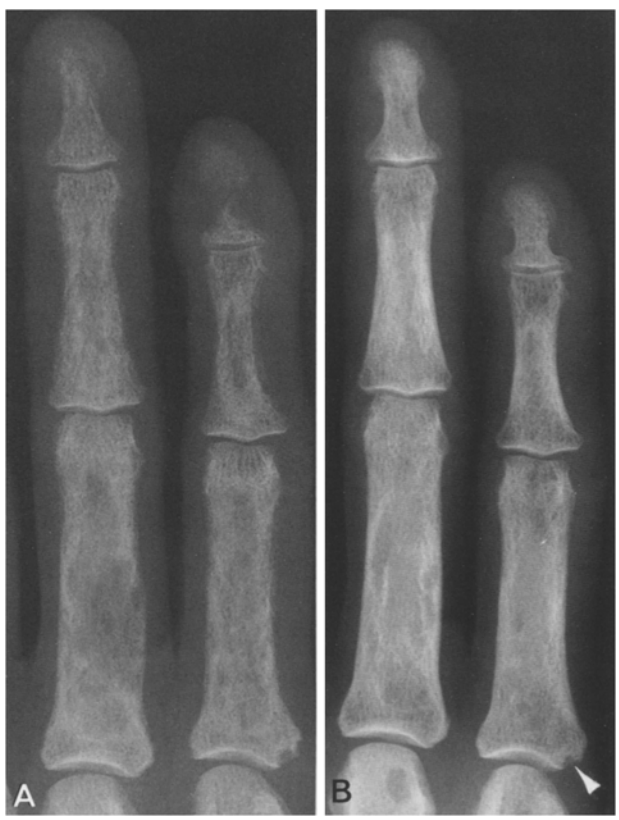

Fig. 3A, B. Forty-seven-year-old woman requiring chronic hemodialysis secondary to renal failure due to chronic pyelonephritis. **A** Severe changes of renal osteodystrophy in left index and middle fingers. **B** Radiograph 5 years after parathyroidectomy demonstrates healing with distal phalangeal brachydactyly of the index finger, narrowing of the diaphyses of the middle phalanges, slight widening of the diaphyses of the proximal phalanx of the long finger from a possible healed brown tumor, and normal appearance of the proximal phalanx of the index finger except for a healed erosion at the base of this phalanx (*arrowhead*)

of advanced renal osteodystrophy, we undertook a study to determine the frequency of this finding and, more important, to document its occurrence. We present six patients having particularly severe findings of renal osteodystrophy, including marked phalangeal tuft resorption in the hands with resultant distal phalangeal brachydactyly. Although briefly mentioned by Destouet and Murphy [1], we could not find any other previous publication reporting healed renal osteodystrophy as a cause of brachydactyly [6].

Case reports and results

Two groups of postparathyroidectomy patients treated since 1974 at the Barnes Hospital, Washington University School of Medicine, Department of Surgery, were evaluated retrospectively. The first group of patients had parathyroidectomy for clinical indications and abnormal laboratory values. There was no history of renal disease in this group. The second group of patients had parathyroidectomy because of severe complications associated with recognized renal disease. Many of the patients in the second group were on renal dialysis. A total of 132 patients in these two groups had radiographs available for review, and 58 of these had hand radiographs. Of the patients in the first group, three showed osseous abnormalities in the hands, but no follow-up studies were available. Of the second group, five patients had destruction of the distal phalanges of the hands with resultant brachydactyly following healing. A third source of radiographic material was the teaching file of the Musculoskeletal Section at the Mallinckrodt Institute of Radiology. Review of these files produced one additional case, yielding a total of six cases of brachydactyly following healing of renal osteodystrophy. Complete clinical and laboratory records including parathyroid hormone levels were available for correlation with radiographs in two of our cases. In two other cases, incomplete laboratory data (with no parathyroid hormone levels) were found. No medical records were available in the final two cases.

In the two cases with complete clinical and laboratory records, parathyroid hormone (PTH) levels were obtained both before and after parathyroidectomy (Table 1). Both patients had markedly elevated parathyroid hormone levels before surgery and radiography of the hands demonstrated the severe changes of renal osteodystrophy (Figs. 2A and 3A). Follow-ups of 4 (case 1) and 8 years (case 2) showed a significant decrease in parathyroid hormone levels. Postparathyroidectomy hand radiographs (14 months after surgery in case 1, 5 years in case 2) demonstrated distal phalangeal brachydactyly after healing of the renal osteodystrophy (Figs. 2B and 3B). Also, in Fig. 2B, healing resulted in a curved or "beaked" appearance of the distal phalanx of the thumb.

The patient illustrated in Fig. 4 developed chronic renal insufficiency because of chronic glomerulonephritis. Radiographs of the hand from August, 1971, showed only minimal subperiosteal bone resorption along the radial margins of the middle phalanges of the hands (Fig. 4A). Similar examinations from August, 1973, and September, 1974, (Fig. 4B, C), demonstrated development and progression of the skeletal changes of renal osteodystrophy, including cortical tuft resorption, subperiosteal and intracortical resorption, and acro-osteolysis (transverse lytic band through the distal phalangeal shaft). No

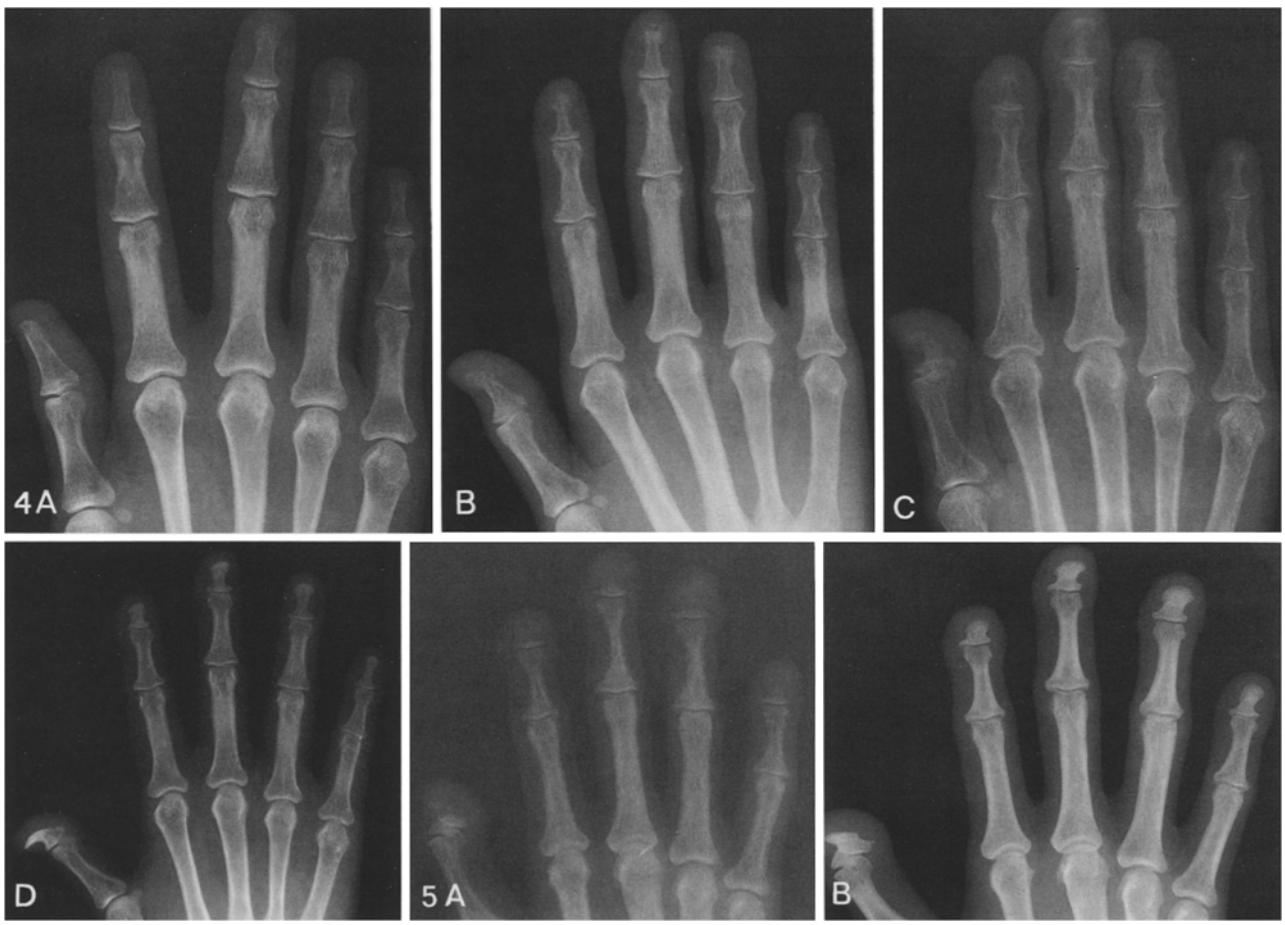

Fig. 4A–D. Nineteen-year-old man with chronic glomerulonephritis. **A** Minimal subperiosteal resorption is present along the radial margins of the middle phalanges of the right hand. **B and C** Progression of renal osteodystrophy with extensive cortical and subperiosteal resorption and acro-osteolysis. **D** Seventeen months after parathyroidectomy, healing with resultant distal phalangeal brachydactyly is demonstrated. Note again the "beaked" appearance of the thumb distal phalanx

Fig. 5A, B. Twenty-five-year-old woman with severe renal osteodystrophy. **A** Extensive cortical and subperiosteal resorption involves the right hand digits. Transverse lucent zones in shafts of tufts indicate acro-osteolysis. **B** Healing with distal phalangeal brachydactyly in subsequent study. The thumb has healed with a "beaked" appearance

parathyroid hormone levels were available from the patient's medical records before surgery. Radiographs obtained 17 months after parathyroidectomy showed distal phalangeal shortening (Fig. 4D). Again, healing of the distal phalanx of the thumbs resulted in a "beaked" appearance.

In our fourth case, a patient with a diagnosis of lobular glomerulonephritis, follow-up hand radiographs 5 months after beginning hemodialysis demonstrated healing of the osseous changes of renal osteodystrophy identified on predialysis radiographs. However, healing resulted in a decrease in the length of the distal phalanges. Serum calcium and phosphate levels were normal during the period over which the radiographs were obtained. No parathyroid hormone levels were obtained during this time.

In our final two cases, clinical and laboratory records were not available. In these patients, severe changes of renal osteodystrophy were manifested by acro-osteolysis, subperiosteal bone resorption, and cortical tuft resorption (Fig. 5A). In follow-up studies, distal phalangeal shortening after healing was again noted (Fig. 5B).

Discussion

We feel that distal phalangeal brachydactyly of the hand after healing results from loss of the poorly mineralized fibrous bone as described in the literature [8]. Our cases illustrate that after it heals, the remaining bone will act as a scaffolding for the remineralization process. Since the ultimate shape and size of a phalanx will be determined by the amount of bone remaining at the time of healing, extensive phalangeal periosteal resorption may result in a thinned or deformed phalanx (Figs. 3B, 4D, 5B). Although we do not have laboratory values for all patients, we believe that remineralization of the remaining shortened distal phalanges following restoration of the calcium phosphate balance and parathyroid hormone values to near-nor-

Table 2. Disorders with short distal phalanges and eroded or hypoplastic tuft[a]

Acquired disorders	Buerger disease
	Burns
	Chemical acro-osteolysis
	Congenital insensitivity to pain
	Frostbite
	Hyperparathyroidism
	Juvenile hyaline fibromatosis
	Leprosy
	Neurotrophic disorder
	Psoriasis
	Raynaud radiation disease
	Scleroderma
	Trauma
Primary disorder	Brachydactyly B
	Osteolysis (Hajdu-Cheney)
Others[b]	Cranioectodermal dysplasia
	Epidermolysis bullosa
	Lesch-Nyhan
	Mandibulo-acral
	Pachydermoperiostosis
	Porphyria
	Progeria
	Pseudoxanthoma elasticum
	Pycnodysostosis
	Rothmund-Thomson
	Scalp defects
	Werner syndrome

[a] Modified from reference [5]
[b] See Poznanski, Chapters 12 and 13, for associated syndromes [5]

mal levels would produce healing accompanied by these abnormalities.

Poznanski has elaborately classified brachydactyly into entities affecting various rows or rays of the hands [5]. This differential diagnosis is extensive and encompasses a broad spectrum of abnormalities. Although renal osteodystrophy and hyperparathyroidism may result in distal phalangeal tuft resorption and acro-osteolysis, the alterations following healing have not been included in the differential diagnosis of distal phalangeal shortening and brachydactyly. A list of disorders associated with distal phalangeal brachydactyly is shown in Table 2 [5].

Since it is impossible to determine the exact frequency of cases of distal phalangeal brachydactyly (since most were collected over several years), it is obvious that this finding is not common. However, when extensive destruction of phalanges occurs, deformity after healing can be expected. We therefore also believe that, although previously unreported, this finding is probably more common worldwide than the literature would imply.

In conclusion, six cases of distal phalangeal brachydactyly in patients with healed renal osteodystrophy and secondary hyperparathyroidism are reported. In all cases, severe osseous changes of renal osteodystrophy were present. After treatment, the shortened distal phalanges and tufts of the hands healed, but there was a significant decrease in length, resulting in brachydactyly. Deformed and thinned phalanges were also seen. These cases present healed renal osteodystrophy as another etiology in the differential diagnosis of distal phalangeal brachydactyly and proximal and middle phalangeal thinning and deformity in the hands.

Acknowledgements. We express our appreciation to Drs. Samuel Wells, Charles Anderson, and Gregorio Sicard for access to their patient information, and to Sharon Keathley and Julie Imo for their secretarial assistance in the preparation of this manuscript

References

1. Destouet JM, Murphy WA (1983) Acquired acroosteolysis and acronecrosis. Arthritis Rheum 26:1150
2. Lehman CA, Schreiber MH (1976) Autonomous hyperparathyroidism in patients on maintenance home dialysis. AJR 127:377
3. Ogg CS (1967) Total parathyroidectomy in treatment of secondary (renal) hyperparathyroidism. Br Med J 4:331
4. Parfitt AM (1977) Clinical and radiographic manifestations of renal osteodystrophy. In: David DS (ed) Calcium metabolism in renal failure and nephrolithiasis, 1st edn. John Wiley, New York, p 167
5. Poznanski AK (1984) The hand in radiologic diagnosis, 2nd edn. WB Saunders, Philadelphia, p 210
6. Poznanski AK (1985) Personal Communication
7. Resnick D, Niwayama G (1981) Diagnosis of bone and joint disorders, 1st edn. WB Saunders, Philadelphia, p 1804
8. Ritz E, Mallucke HH, Krempien B, Mehls O (1977) Bone histology in renal failure. In: David DS (ed) Calcium metabolism in renal failure and nephrolithiasis, 1st edn. John Wiley, New York, p 220
9. Stevens LE, Bloomer HA, Lower GD, Maddock R, Reemtsma K (1969) Indications for subtotal parathyroidectomy in patients with renal transplantation. Ann Surg 169:578
10. Sundaram M, Joyce PF, Shields JB, Riaz MA, Sagar S (1979) Terminal tufts of the hands – site for earliest changes of renal osteodystrophy in patients on maintenance hemodialysis. AJR 133:25
11. Sundaram M, Phillipp SR, Wolverson MK, Riaz MA, Rao BJ (1980) Ungual tufts in the follow-up of patients on maintenance hemodialysis. Skeletal Radiol 5:247
12. Talwalkar YB, Puri HC, Hawker CC, Tseng C, Campbell JR, Campbell RA (1979) Parathyroid autotransplantation in renal osteodystrophy. Am J Dis Child 133:901