Brachyonychia Associated with Acroosteolysis in Chronic Kidney Disease: How Phalange Shape Influences Nail Morphology

José David Rosales Figueroa^a Patricia Chang^b

^aCentro Clínico Reforma, Ciudad de Guatemala, Guatemala; ^bHospital General de Enfermedades IGSS, Ciudad de Guatemala, Guatemala

Keywords

Chronic kidney disease · Renal insufficiency · Brachyonychia · Racket nails · Transverse acroosteolysis · Longitudinal acroosteolysis · Radiographic changes of the phalanges · Nail morphology · Nails · Nail disease

Abstract

Brachyonychia is a rare manifestation in patients with chronic kidney disease. Longtime disease, secondary hyperparathyroidism, and hemodialysis are common conditions among those who present it. We evaluated 8 cases who presented brachyonychia in the nephrology department and compared the clinical versus the radiographic findings, and evaluated how the tissue adjusts to the underlying bone structure, giving different forms to the nails. We conclude that brachyonychia and acroosteolysis in chronic kidney disease suggest long-term disease, secondary hyperparathyroidism, and hemodialysis, besides it being a good model on how the bony structure defines the soft tissue morphology.

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We reviewed 8 cases from patients with chronic kidney disease (CKD) undergoing hemodialysis treatment at the Nephrology Department of “Instituto Guatemalteco de Seguridad Social.” These patients presented with brachyonychia and all of them had secondary hyperparathyroidism. The sample included 6 males and 2 females between 21 and 60 years of age (Table 1). Most patients had lived with the disease for more than 6 years. The parathyroid hormone (PTH) level was measured, with a lower obtained value of 429 µg/mL and a higher value of 2,611 µg/mL. All patients had X-ray evaluation of the hands that showed acroosteolysis (AO) in the phalanges presenting with brachyonychia. Other findings were loss of interphalangeal space, osteoporosis, osteonecrosis, calcifications, and periosteal sclerosis in the metacarpal.

Nail dystrophy has been described as a sign of phalangeal osteolysis, since the function and shape of the nail unit is in part dependent on the underlying bone [1, 2]. Clinically, nail dystrophy can present as pseudoclubbing, brachyonychia, koilonychia, pincer nails, and anonychia [1, 3, 4].

Brachyonychia was described by DuBois in 1926 as an altered shape of the nail plate, which is short, broad, and flat, secondary to the deformity of the underlying bone and soft tissue of the terminal phalanges [1, 5, 6]. Two

forms have been described: genetic and acquired. The former is believed to be secondary to changes in the cartilage, which could be obliteration, as proposed by Oluf Thomsen, or early closure of the cartilaginous line of the terminal phalanx, as proposed by Burrows [6].

According to Baran et al. [6], the principal cause for the acquired form of brachyonychia is hyperparathyroidism. This condition may be due to several causes including chronic renal failure, malabsorption, osteomalacia, and rickets [6, 7]. Radiographic findings vary depending on the stage of the disease. At early stages, subperiosteal demineralization can be noted in the phalanges, followed by resorption of the terminal phalange, and finally AO [6].

AO describes the partial or total lytic destruction of the finger or toe phalanges, characterized by resorption of the bones [2–4, 8, 9]. It can be classified as primary or familial, idiopathic or nonfamilial, and occupational or secondary, as a consequence of several conditions. The most common causes being inflammatory diseases, sensory neuropathy, vascular disease, metabolic diseases, exposure to vinyl chloride, and trauma (Table 2) [1, 2, 8–10].

There have been several physiopathologic mechanisms proposed for AO, which vary in function with the base disease [11].

1. Vascular: (a) Occlusion and stenosis leads to bone infarction and resorption. This is secondary to elevated levels of cytokines like vascular endothelial growth factor. Osteoclast formation is also induced by hypoxia. [1, 12, 13]. (b) Vasodilatation increases blood flow (hyperemia), which in turn elevates oxygen partial pressure, favoring modulation and activity of osteoclast, resulting in bone resorption. This has been proposed as the mechanism for cold induced AO [1].
2. Nervous phenomena: (a) Sensory neuropathy causes predisposition to physical or thermal trauma, resulting in osteolysis secondary to mechanical trauma [10]. (b) Autonomic neuropathy leads to vasomotor instability, leading to loss of vasoconstriction and hyperemia, which has been postulated as a mechanism for osteolysis [10, 12].
3. Metabolic: Mainly caused by abnormal levels in serum calcium, phosphate, PTH, and active vitamin D [14].

Radiographic appearance of phalangeal osteolysis can be divided into three groups: those with resorption of terminal tufts, resorption of the midportion of the phalanges, and those with periarticular resorption [8]. Only two variants orient the clinical manifestations: transverse variant of AO presents an osteolytic band in the midportion of the phalange preserving its base and tuft, whereas

Table 1. Patient study characteristics

Case number	Sex	Age, years	Fingers affected	Time on hemodialysis, years
1	M	44	thumbs, index	5
2	M	50	whole hand, R and L	7
3	M	57	thumbs	15
4	M	56	whole hand, R and L	24
5	F	26	1, 2, 3, 5, R and L	8
6	M	35	1, 2, and 3	3
7	M	21	whole hand, R and L	16
8	F	29	whole hand, R and L	10

R, right; L, left.

Table 2. Mechanism of acquired acroosteolysis in related diseases

Mechanism	Diseases
Vascular	Vascular occlusion a. scleroderma b. diabetes mellitus Frostbite
Nervous	Diabetic mellitus Leprosy
Metabolic	Hyperparathyroidism Diabetes mellitus
Inflammatory	Scleroderma Raynaud disease Psoriatic arthritis Juvenile chronic arthritis Epidermolysis bullosa Dermatomyositis
Trauma	Mechanical trauma Burns Electricity
Drugs	Phenytoin Ergot derivatives intoxication Polyvinyl chloride exposure
Tumors	Epidermal inclusion cyst Glomus tumor

those with tuft resorption present a pencil-like pattern, also known as longitudinal AO [1, 4] (Fig. 1).

CKD is associated with several skin and nail manifestations, which increase according to duration and severity of the renal disease. Common manifestations are xer-

Fig. 1. Brachyonychia in a patient with 16 years of CKD and 3 years of hemodialysis. Clinical (a) and radiographic (b) images. Longitudinal acroosteolysis (white triangles) (b). Brachyonychia secondary to transverse acroosteolysis (white arrows) in a patient on hemodialysis for 8 years (c, d). Also an osteophyte is seen (white-outline arrow) (d).

rosis, skin hyperpigmentation, pruritus, and nail changes [15–17]. Most nail changes in CKD appear or increase with dialysis and tend to disappear after kidney transplantation [15]. Nearly 70% of patients on dialysis have nail alterations such as half and half nails, splinter hemorrhage, or absence of lunula, among others [16, 17]. In Sakarya University, Turkey, 365 CKD patients were evaluated and 15.3% were found to have nail changes. However, there was no difference in the prevalence of nail changes between dialysis and non-dialysis patients, nor did they report any patients with brachyonychia [18]. According to these findings, brachyonychia remains an uncommon feature of CKD.

Hyperparathyroidism due to parathyroid gland hyperplasia and autonomic secretion is proposed as the mechanism of AO in patients with CKD [19]. Since the bony structure of the nail apparatus determines some nail changes, it has been hypothesized that patients with chronic high levels of PTH experience brachyonychia [19].

Abnormalities in bone turnover are the result of hyperphosphatemia, negative calcium balance, insufficient active vitamin D, vitamin D receptors, and calcium receptors, as well as the proinflammatory state in patients with CKD. High turnover bone disease results from PTH levels higher than 400 pg/mL, which induces osteoclastogenesis and a compensatory osteoblastogenesis. Yet there is

also inhibition of osteoblast function and stimulation of osteoblast apoptosis with the end product of bone resorption [14, 20].

Patients with CKD also show resistance to vitamin D. It has been noted that supplementation with active vitamin D diminishes the activity of 25-hydroxylase and 1-alpha-hydroxylase enzymes, which are essential for endogenous formation of vitamin D forms, calcidiol and calcitriol, resulting in lower levels of vitamin D in the kidney as well as in extrarenal tissues. Lower bone formation and trabecular mineralization have been shown to occur in PTH in an independent manner. All of these changes lead to a significant loss in the mechanical integrity and toughness of cortical bone [14].

Cortical bone resorption has been noted as the primary mechanism of bone damage in CKD patients. The cortical bone suffers a decrease in its thickness, whereas trabecular bone thickness increases but has lower quality, signifying reduced trabecular connectivity and increased trabecular perforations [20].

Conclusion

Acquired brachyonychia is a rare nail morphology abnormality, most frequently seen in patients with hyperparathyroidism, a main cause of bone disease in CKD.

This morphological change might be due to transverse AO secondary to hypertrophy of the phalange tuft, trabecular bone, which causes a broadening of the nail apparatus and the loss of the corresponding phalange midportion, or cortical bone, resulting in a shorter phalange. Similar and even more drastic nail changes are seen in advanced AO, when the distal and midportions of the bone have been completely resorbed.

In longitudinal AO, there is simply a thinning of the fingertip and nail plate giving a triangular appearance to the nail plate and fingers, reflecting the pencil-like radiographic findings.

According to this small series of cases, the shape of nails in CKD patients appears to suggest chronicity, high PTH levels, and an advance stage of the disease.

Statement of Ethics

All patients gave their informed consent about using their clinical information for educational studies.

Disclosure Statement

The authors declare no conflicts of interest.

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