Acro-osteolysis in a Filipino Male with Vinyl Chloride Exposure: A Case Report

Karl Babe G. Tagomata, MD and Therese Eileen B. Lladoc-Natividad, MD

Section of Rheumatology, Department of Internal Medicine, East Avenue Medical Center, Quezon City, Philippines

ABSTRACT

Occupational acro-osteolysis pertains to bone resorption of the distal phalanges of the hands and feet among workers with vinyl chloride exposure. We report the case of a Filipino man with osteolysis of the distal phalanges of the hands initially considered to have systemic sclerosis. The patient had gradual shortening of the fingers, thickening of the skin over the extremities, and hypopigmented patches over a span of more than 20 years. His lower extremities presented with non-pitting edema, skin thickening, and neuropathy, without shortening of the digits. Difficulty of ambulation was apparent due to the development of feet inversion. Radiographic findings of the hands and feet included resorption of distal phalanges, erosive and sclerotic changes, and narrowed joint spaces. Other conditions considered were Hansen's disease, skeletal tuberculosis, and diabetic neuropathic arthropathy, which were eventually ruled out. The final diagnosis was occupational acro-osteolysis secondary to vinyl chloride exposure. The patient underwent serial total contact casting of the bilateral lower extremities to relieve bipedal edema and to reposition the feet. This case emphasizes the significance of investigating a patient's occupational history and highlights a rare sequela of exposure to a commonly used chemical agent in the manufacture of polyvinyl chloride products.

Keywords: occupational acro-osteolysis, bone resorption, vinyl chloride, Filipino

INTRODUCTION

The term acro-osteolysis refers to bone resorption of the distal phalanges of the hands and feet and can occur at the terminal tuft or on the shaft (also called transverse or band acro-osteolysis).[1] Conditions associated with acro-osteolysis include genetic disorders, infections, occupational activities, and rheumatic disorders such as psoriatic arthritis and systemic sclerosis.

Occupational acro-osteolysis has been described in Western countries as early as the 1960's. These cases attributed the bone resorption to exposure to vinyl chloride among workers in polyvinyl chloride (PVC) production. We report the case of a patient with shortening of digits of the hands initially referred to Rheumatology service for consideration of systemic sclerosis.

This report aims to add knowledge on the occupation-related diseases that may be observed among Filipinos with previous or current exposure to vinyl chloride.

CASE

The patient was a 52-year-old Filipino man from Tacloban City. He was admitted at the East Avenue Medical Center last January 2021 for undocumented fever, cough, and exertional dyspnea. The initial impression was sepsis secondary to moderate-risk community-acquired bacterial pneumonia with acute kidney injury. The nasopharyngeal

Poster presented at Philippine Rheumatology Association Annual Convention 2021 and Asia Pacific League of Associations for Rheumatology Congress 2021.

eISSN 2094-9278 (Online)
Published: July 27, 2023
https://doi.org/10.47895/amp.vi0.5224

Corresponding author: Karl Babe G. Tagomata, MD
Section of Rheumatology
Department of Internal Medicine
East Avenue Medical Center
East Avenue, Quezon City 1100, Philippines
Email: drkbabe2014@gmail.com
ORCiD: https://orcid.org/0009-0003-8992-9819

swab for severe acute respiratory syndrome coronavirus-2 (SARS-CoV-2) reverse transcriptase-polymerase chain reaction was negative. During his prolonged hospital stay, he had episodes of hospital-acquired pneumonia which were treated with piperacillin-tazobactam then meropenem. He was referred to the Rheumatology service with the impression of systemic sclerosis because of the shortening of fingers.

More than 30 years ago, the patient noticed "map-like" hypopigmented patches on his both hands and wrists associated with tingling sensation usually during cold weather. The patient then observed gradual shortening of his fingers over the past 20 years. These did not prompt consult as there was no associated pain, pruritus, or ulceration. He also reported one episode of bluish discoloration of a finger on the left hand which resolved after a few days. It was not associated with cold exposure and never recurred. One year prior to admission, he had bilateral ankle and feet swelling associated with difficulty in ambulation, a sensation of thickening of skin over the extremities and occasional tingling sensations of both hands (for which he would apply a herbal concoction of guava leaves). He did not take any medications or seek medical attention.

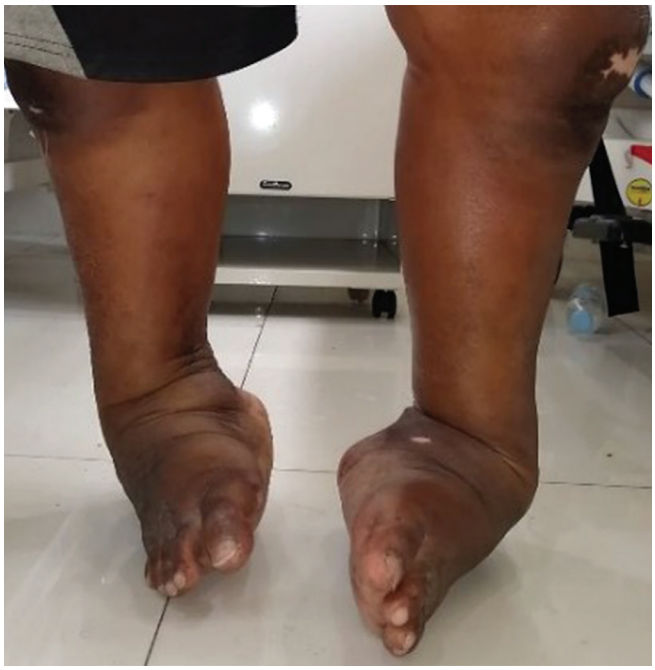

Figure 1. Inversion of feet on standing position.

About three months prior to admission, he noted that his feet would spontaneously invert, causing difficulty in ambulation. One month prior to admission, he noticed similar swelling and thickening sensation on the left arm. He denied fatigue, weight loss, morning stiffness, gastrointestinal or genitourinary symptoms, and claudication. He denied any history of hypertension, diabetes, bronchial asthma, pulmonary tuberculosis. He sustained a burn injury on his left hand and right arm from previous work. He had never undergone any surgeries. In his family, hypertension was prevalent, but no one had similar extremity abnormalities. He was a four pack-year smoker, having stopped smoking several years ago, and drank alcohol in moderation. He previously used marijuana and methamphetamine for less than a year.

For the past 30 years, he has worked as a painter, laborer and cleaner in several construction sites; including a five-month stint at a polyvinyl chloride (PVC) factory . He used safety boots but not hand gloves when cleaning containers and when transferring industrial chemicals. At work, he noticed that a coworker had similar "map-like" hypopigmented patches but no shortening of digits.

His vital signs were stable, but he had difficulty with standing, balance, gait (inversion of both feet, Figure 1), and bed transfers. His skin had areas of hypopigmentation on the hands, wrists, knee, and ankle, without telangiectasias or "salt-and-pepper." His oral aperture could admit three fingerbreadths with no vertical furrowing of the perioral skin. He had bibasal crackles on chest auscultation. Cardiovascular and gastrointestinal examination were unremarkable. He had good pulses on all extremities; non-pitting edema was noted on both lower extremities. Contractures on the thumb, fourth and fifth fingers of the right hand as well as third and fifth fingers of the left hand were observed (Figure 2). The skin on his hands was

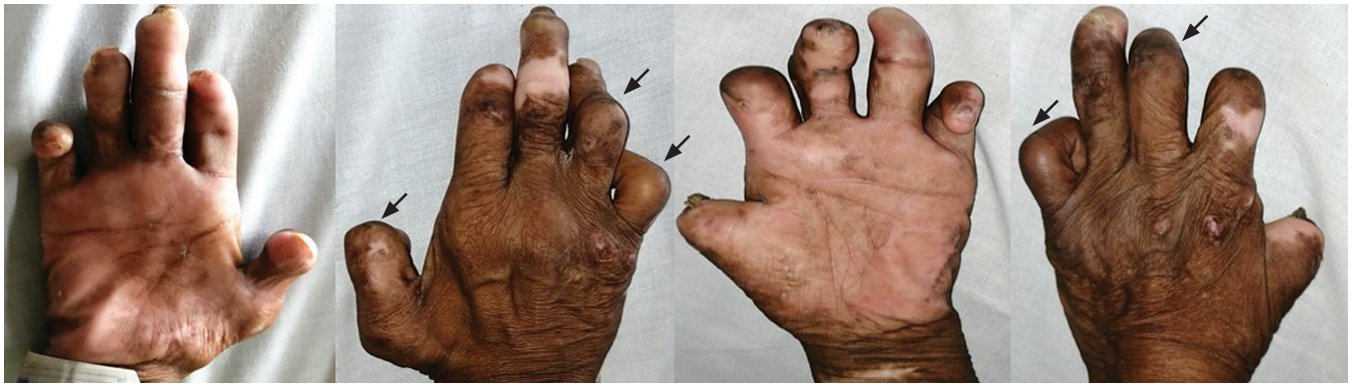

Figure 2. Acro-osteolysis, contractures *(black arrows)*, and hypopigmented patches.

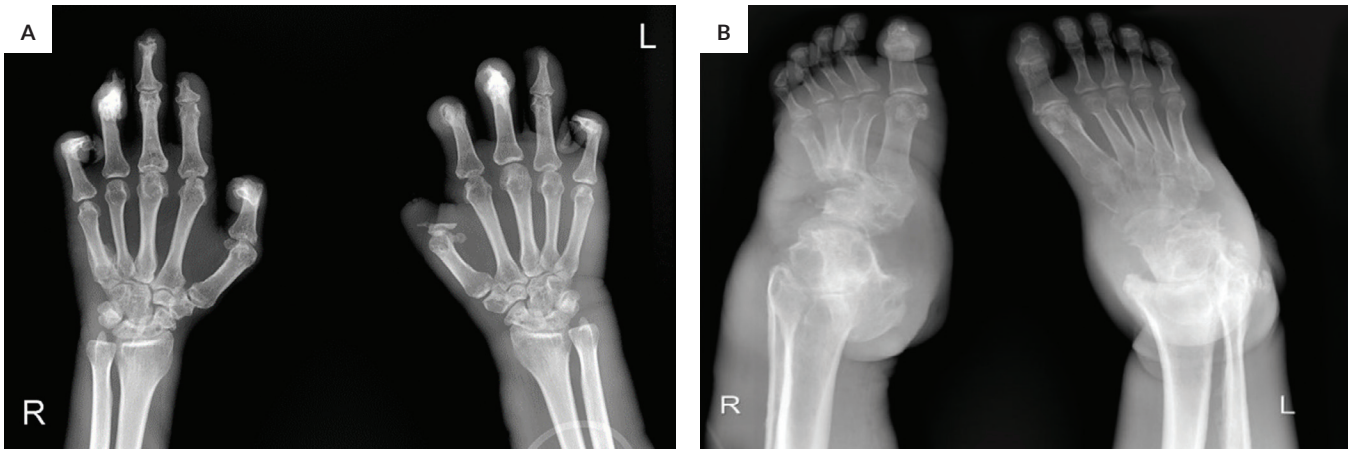

Figure 3. Resorption of phalanges of the hands and erosion with lytic changes on the feet.

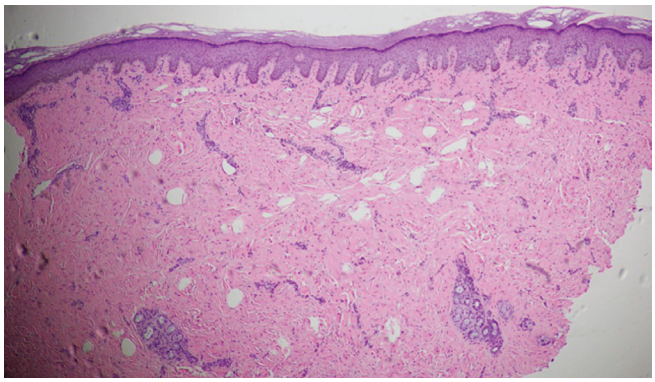

Figure 4. Section *(site: left foot)* showing markedly thickened, closely packed collagen bundles throughout dermis with eccrine glands entrapped by collagen, and replacement of subcutaneous fat by collagen. Observed under Scanning Magnification. Stain used was Hematoxylin and Eosin.

not thickened nor was there sclerodactyly of the remaining fingers. There were no ulcerations, but hypoesthesia was noted on the lower extremities with 50% sensory deficit up to the knees. Muscle atrophy on both palmar and dorsal aspects of the hands were apparent.

The initial working impression was Hansen's Disease, probably lepromatous type, with secondary neuropathic arthropathy; systemic sclerosis was less likely.

Initial laboratories showed a hemoglobin of 83 g/L, a white blood cell count of 13.3 x10^9/L, neutrophils of 85%, a platelet count of 173 x10^9/L, a creatinine level of 86.70 umol/L, urea nitrogen level of 4.70 mmol/L, a serum sodium level at 135 mmol/L and a serum potassium level at 2.44 mmol/L. Initial chest radiograph showed cardiomegaly with pulmonary congestive changes and infiltrates observed more in the right lower lung field than the left, suggestive of pneumonia. This was initially managed with ceftriaxone and azithromycin. A chest CT scan confirmed the presence of pneumonia bilaterally, with loculated pleural effusion on the right more than the left, and bilateral emphysematous and bronchiectatic changes, suggestive of pulmonary tuberculosis. The renal ultrasonography showed bilateral chronic kidney disease, most likely secondary to hypertensive kidney disease.

Radiographs of the upper extremities showed decreased density in the osseous structures (Figure 3). X-rays of the right hand showed complete resorption of the distal phalanges of the first, second, and third digits, with fusion of the fourth middle and distal phalanges, erosive and sclerotic changes in the fifth distal phalanx, and cortical sclerosis and subchondral cystic lucencies at the scaphoid, lunate, and distal end of the radius. X-rays of the left hand showed complete bone resorption of the distal phalanges of the first, second, third, and fourth digits, almost complete bony resorption involving the proximal phalanx of the first digit, and cortical sclerosis and subtle subchondral cystic lucencies at the scaphoid and distal end of the radius. Radiographs of the feet showed lytic and erosive changes in the proximal ends of the second to fifth metatarsals of the right foot, and marginal erosion, lytic changes, and narrowed joint spaces seen involving the tarsal bones of both feet (Figure 3).

To investigate his exertional dyspnea, fatigue, and bipedal edema, a 2D-echo was done, revealing dilated cardiomyopathy with an ejection fraction of 38% and moderate pulmonary hypertension. This was managed with telmisartan, spironolactone, amlodipine, and sildenafil.

The patient was referred to Dermatology with the consideration of lepromatous leprosy. Clofazimine was started with plans to start dapsone once glucose-6-phosphate dehydrogenase (G6PD) deficiency was ruled out. Slit-skin smear performed was negative for acid-fast bacilli (AFB) and skin punch biopsy obtained from the left foot reported sclerosing dermatitis, probably morphea or scleroderma (Figure 4), and was considered nonspecific. A repeat skin biopsy of the right leg showed superficial perivascular dermatitis with basal layer hyperpigmentation, which did not match the diagnosis of Hansen's Disease. Regimen for

leprosy was then discontinued. Nail-fold capillaroscopy revealed few microhemorrhages on the fourth digit of the right hand but no dilated or tortuous capillaries and telangiectasias. Screening tests for diabetes and connective tissue disease were negative (ANA 1:10).

Percutaneous Achilles tendon lengthening, and serial total contact casting of bilateral lower extremities were performed by Orthopedics to relieve bipedal edema and reposition the feet. The patient was discharged as a case of occupational acro-osteolysis and advised to follow-up with Orthopedics for change of total contact cast to fiberglass cast.

DISCUSSION

The first reports of work-related acro-osteolysis were published in Western journals in the 1960's to 70's. Occupational or industrial acro-osteolysis was described by Walker (1976) as the triad of Raynaud's phenomenon, sclerodermatous skin change and lytic bone lesions among workers involved in the polymerization of vinyl chloride monomer (VCM) to polyvinyl chloride (PVC).[2] The incidence of occupational acro-osteolysis on earlier epidemiologic surveys was approximately 3%, ranging from 1 to 6%.[2,3] Wilson et al. (1967) also observed 31 cases of hand disorders out of 3,000 personnel working in PVC manufacture and found no ethnic or racial tendency for the syndrome.[4] In an earlier report by Walker (1975), acute Raynaud's phenomenon and lytic bone lesions were manifested by two workers who either packed the dry PVC powder (called 'baggers'), or hosed and cleaned the reactors (called operators), consistent with the first reported case described by Cordier and colleagues in 1966.[3] Wilson et al. (1967) also mentioned 'polycleaners' to refer to those who manually clean the reactor by using hand scraping techniques.[4] The VCM, a colorless, gaseous, and sweet-smelling organochloride, is used in the production of PVC resins, which are used commonly in building materials, construction, and home furnishings.[5]

Several conditions can present as acro-osteolysis. Botou et al. (2017) reviewed the possible etiologies, including rheumatic diseases, infections, endocrinopathies, genetic and lysosomal storage diseases, and occupational hazards.[1] Scleroderma was the initial diagnosis for a case reported by Harris and Adams (1967) on a man who presented with puffiness of the face, thickening of the skin and Raynaud's phenomenon. He was later assessed to have occupational acro-osteolysis upon eliciting a 7-year history of working in a PVC plant.[6] Niamane et al. (2005) documented three patients having acro-osteolysis out of 47 patients with psoriatic arthritis.[7]

Skeletal tuberculosis may also be considered given our endemic setting and the patient's clinically-diagnosed pulmonary tuberculosis; it can cause arthritis, osteomyelitis, and sometimes acro-osteolysis with reactive sclerosis.[8] Leprosy may also present with resorption of distal phalanges as reported by Chu et al. (2017).[9] Skeletal tuberculosis and leprosy were ruled out with the absence of AFB on skin smear and biopsy.

Baer et al. (2012) showed that a poorly controlled type I diabetes mellitus can also cause shortening of fingers–radiographically described as resorption of the distal phalanx of the left little finger and loss of bone in the distal phalanx of the right little finger.[10] Diabetes and leprosy are common causes of neuropathic arthropathy, a condition that presents with joint destruction due to underlying disorder of the nervous system.[11] This was ruled out via history and diagnostic tests.

Familial acro-osteolysis syndromes, such as Hajdu-Cheney syndrome, are reported genetic etiologies. In our patient, systemic sclerosis was ruled out clinically, utilizing the 2013 American College of Rheumatology/European League Against Rheumatism criteria.

Occupational acro-osteolysis after repetitive trauma is rare, with reports of two dancers, a surfer, and a guitar player.[1]

Our final diagnosis is occupational acro-osteolysis secondary to vinyl chloride exposure, which occurred 30 years ago.

In the survey among 37 PVC workers (reactor operators, maintenance men, 'baggers' and warehousemen) with length of employment ranging from 9 months to 5 years, Walker (1976) showed that the most common presenting symptoms are excessive fatigue and coldness of the hands. A noteworthy feature specified by the author was the feeling of being 'unsteady' or 'slightly drunk' on at least one occasion with exposure to vinyl chloride fumes.[2] Wilson et al. (1967) noted that majority of the patients had two common findings: Raynaud's phenomenon and acro-osteolysis of the distal phalanges.[4] Most of the reports revealed that patients did not have bothersome symptoms to warrant medical consultation. This was also the case with our patient.

Upon repeated history-taking, our patient was able to recall a spontaneously resolving Raynaud-like phenomenon but could not recall its association with cold exposure.

Preston et al. (1976) reported a wide radiological spectrum ranging from no radiographic abnormality to extensive acro-osteolysis of the upper and lower extremities and even erosion of the sacroiliac joints and the mandible.[12] Aside from terminal phalanges of the fingers and feet, Harris and Adams (1967) also reported bony erosions at the sacroiliac joints and the patellae.[6] Wilson et al. (1967) classified acro-osteolysis into the mild, advanced, and healing stages. The mild stage presents as a loss of the cortex of at least one of the tufts of the distal phalanges. The advanced stage presents with complete loss of the tuft and a part of the shaft of at least one of the distal phalanges. The healing stage was described when there is fragmentation of the remaining tuft or a filled-in area that was previously lytic.[4] Our patient is most likely in the advanced stage.

Hypoesthesia was not mentioned in case reports; however, Walker (1976) observed paresthesia or tingling sensation in 16 out of 37 patients.[2] Hypoesthesia and paresthesia are most

likely part of the neuropathic arthropathy from the chemical exposure. Although hypopigmentation was not mentioned in the existing literature, it could be caused by other chemical agents in the factory or by the degree of chemical exposure of the patient. Our patient only noted inversion of both feet in the past few months, but it could have started gradually at an earlier time. This is a new reported finding caused by lysis and erosion of the tarsal bones.

In the survey of Walker (1976), some patients can also have skin thickening, limitation of finger extension and mild 'clawing' of the hands while others would have swelling of the face, particularly in the periorbital area that tends to improve in warmer conditions. Patients also manifested with skin lesions resembling scleroderma while a few had clubbing of the fingers, called pseudoclubbing.[4,5] The affected extremities may also have soft tissue thickening.[12]

On histopathology, the epidermis may be normal, but the dermis may have some destruction of elastic tissue, or medial thickening of dermal arteries in severe Raynaud's phenomenon.[2] Severe narrowing of small dermal arteries has also been reported by Harris and Adams (1967).[6]

The pathophysiology of the acro-osteolysis from vinyl chloride exposure is unclear. Being a highly volatile compound, inhalation is the main route of exposure to VCM. The chemical agent distributes widely throughout body compartments, is metabolized in the liver, and is excreted by the kidneys. Acro-osteolysis is putatively mediated through obstructive lesions of small peripheral arteries.[5]

The duration of exposure is a significant factor in this occupational disease. Wilson et al.'s (1967) cases had at least 12 months of cleaning experience. It is believed that this condition is the result of three main factors: chemical insult (exposure to the solid or vapor state of the monomer, catalyst, and reaction products), physical insult (prolonged hand scraping), and personal idiosyncrasy (involving the individual's vascular system, nerves, or collagen).[4] Our patient was exposed for less than six months. He recalled a coworker having the same hypopigmented patches on the hands but no shortening of digits. Other coworkers had no similar symptoms. The interplay of these three factors resulted in our patient's extensive acro-osteolysis despite a short vinyl chloride exposure.

Personal idiosyncrasy may be an important concept as two of the four cases presented by Preston et al. (1976) with work exposure for two to five years did not show acro-osteolysis or any other abnormality.[12] Moreover, Wilson et al. (1967) did not observe acro-osteolysis among more than 1,000 individuals who were handlers of the finished PVC resin or who processed the polymers into plastic products.[4]

Early recognition and cessation of exposure could prevent severe skin change.[2] Bone changes can also regress after removing the worker from the hazard.[12] Harris and Adams (1967) observed that workers removed at the time of diagnosis may have no further radiological change or may have healing with partial ossification after a year.[6] Stewart et al. (1975) documented a worker who was removed after six years of work in a PVC factory. After 14 months, there was minimal clinical improvement, with partial radiographical healing.[13] Heart failure may not be related to the vinyl chloride exposure as no study has associated heart failure to the chemical agent. Carreón et al. (2014) reported elevated mortality from hepatobiliary cancer but not from non-Hodgkin lymphoma or coronary artery disease.[14] Therefore, screening for hepatobiliary malignancy should be included in the workup.

A thorough review of literature of unpublished case reports from the Philippine Rheumatology Association database and locally available journals did not yield the prevalence of occupational acro-osteolysis among Flipinos. The limitations of this report include the accuracy of the patient's account of duration and manner of chemical exposure, and the absence of diagnostic test to confirm and directly attribute the patient's manifestations to vinyl chloride exposure. We recommend that data on the prevalence of this condition should be gathered due to its rare but significant morbidity.

CONCLUSION

We report the case of a patient with occupational acro-osteolysis who was initially considered to suffer from a rheumatic condition. Consistent with early Western reports of industrial acro-osteolysis, systemic sclerosis and Hansen's disease were among the early differentials. The importance of an accurate occupational history cannot be overemphasized in rare conditions associated with chemical exposure. This is the first reported case of occupational acro-osteolysis in a Filipino worker who handled vinyl chloride.

Acknowledgments

The authors would like to thank the EAMC Department of Dermatology, especially with Dermatopathology

Statement of Authorship

Both authors contributed in the conceptualization of work, acquisition and analysis of data, drafting and revising, and approved the final version submitted.

Author Disclosure

Both authors declared no conflicts of interest.

Funding Source

This study has no funding support.

REFERENCES

1. Botou A, Bangeas A, Alexiou I, Sakkas LI. Acro-osteolysis. Clin Rheumatol. 2017 Jan; 36(1):9-14. doi: 10.1007/s10067-016-3459-7. Epub 2016 Oct 29. PMID: 27796661.
2. Walker AE. Clinical aspects of vinyl chloride disease: skin. Proc R Soc Med. 1976 Apr; 69(4):286-9. PMID: 131318; PMCID: PMC1864532.

3. Walker A. Occupational acro-osteolysis (two cases). Proc R Soc Med. 1975 Jun; 68(6):343-4. PMID: 1208525; PMCID: PMC1863762.
4. Wilson RH, McCormick WE, Tatum CF, Creech JL. Occupational acroosteolysis. Report of 31 cases. JAMA. 1967 Aug 21; 201(8):577-81. doi: 10.1001/jama.201.8.577. PMID: 5006758.
5. Fralish MS, Downs JW. Vinyl chloride toxicity. 2020 Jun 25. In: StatPearls. Treasure Island (FL): StatPearls Publishing; 2021 Jan. PMID: 31335054.
6. Harris DK, Adams WG. Acro-osteolysis occurring in men engaged in the polymerization of vinyl chloride. Br Med J. 1967 Sep 16; 3(5567):712-4. doi: 10.1136/bmj.3.5567.712. PMID: 6038365; PMCID: PMC1843033.
7. Niamane R, Bezza A, El Hassani S, Bensabbah R, Hajjaj-Hassouni N. Apport des critères doigts--orteils dans le diagnostic précoce du rhumatisme psoriasique [Value of the radiographic criteria "fingers and toes" in the early diagnosis of psoriatic arthritis]. J Radiol. 2005 Mar; 86(3):321-4. French. doi: 10.1016/s0221-0363(05)81361-7. PMID: 15908872.
8. Burrill J, Williams CJ, Bain G, Conder G, Hine AL, Misra RR. Tuberculosis: a radiologic review. Radiographics. 2007 Sep-Oct; 27(5):1255-73. doi: 10.1148/rg.275065176. PMID: 17848689.
9. Chu BBR, Falcão Brandão Côrtes Gobbo G, Copês R, Gutjahr G, Cavalcanti Cossa E, Dos Santos Paiva E. Leprosy simulating systemic sclerosis: a case report. Rev Bras Reumatol Engl Ed. 2017 Nov-Dec; 57(6):630-632. English, Portuguese. doi: 10.1016/j.rbre.2016.09.010. Epub 2016 Oct 18. PMID: 29173702.
10. Baer AN, Zahr ZA, Khan S, Polydefkis M. Acroosteolysis in diabetes mellitus. J Rheumatol. 2012 Dec; 39(12):2364-5. doi: 10.3899/jrheum.120662. PMID: 23204315.
11. Chan RLS, Chan CH, Chan HF, Pan NY. The many facets of neuropathic arthropathy. BJR Open. 2019 Jul 29; 1(1):20180039. doi: 10.1259/bjro.20180039. PMID: 33178926; PMCID: PMC7592473.
12. Preston BJ, Jones KL, Grainger RG. Clinical aspects of vinyl chloride disease: acro-osteolysis. Proc R Soc Med. 1976 Apr; 69(4):284-6. PMID: 1265039; PMCID: PMC1864528.
13. Stewart JD, Williams DM, McLachlan MS. Acro-osteolysis in a polyvinyl chloride worker with an atypical industrial history. J Soc Occup Med. 1975 Jul; 25(3):103-9. doi: 10.1093/occmed/25.3.103. PMID: 162645.
14. Carreón T, Hein MJ, Hanley KW, Viet SM, Ruder AM. Coronary artery disease and cancer mortality in a cohort of workers exposed to vinyl chloride, carbon disulfide, rotating shift work, and o-toluidine at a chemical manufacturing plant. Am J Ind Med. 2014 Apr; 57(4):398-411. doi: 10.1002/ajim.22299. Epub 2014 Jan 24. PMID: 24464642; PMCID: PMC4512282.