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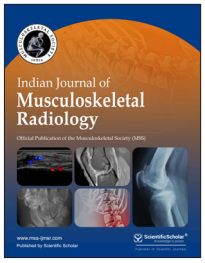

Pictorial Review

Lesions of the Distal Phalanx: Imaging Overview

Ramanan Rajakulasingam[1], Christine Azzopardi[1], Jennifer Murphy[1], Mark Davies[1], Andoni Toms[2], Steven James[1], Rajesh Botchu[1]

[1]Department of Musculoskeletal Radiology, Royal Orthopedic Hospital, Birmingham, West Midlands, [2]Department of Musculoskeletal Radiology, Norfolk and Norwich University, Norwich, England.

***Corresponding author:**
Dr. Rajesh Botchu,
Department of Musculoskeletal Radiology, The Royal Orthopedic Hospital Bristol Road South, Northfield, Birmingham, UK.

drbrajesh@yahoo.com

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ABSTRACT

Lesions of the distal phalanx often pose a radiological dilemma as the differential diagnosis is potentially broad. Particularly for lytic lesions, there is a concern whether an underlying primary tumor or a metastatic deposit is present. Bone tumors of the hand are infrequent when compared to soft tissue tumors, and those involving the distal phalanx are very rare. Lesions of the distal phalanx may arise secondary to benign or malignant pathologies and may reflect primary or secondary tumors. The most common benign lesion is an enchondroma while the most frequent primary malignant lesion is chondrosarcoma. Inflammatory, metabolic, autoimmune, and traumatic pathologies may also present with a wide spectrum of radiological changes involving the distal phalanx. It is these tumor-like mimics which can further complicate diagnosis. We hope to highlight distinguishing features between these entities, allowing the radiologist to generate a clinically useful differential diagnosis. Even though most lesions are benign, it is crucial to differentiate them as some may be the first presentation of more a systemic condition.

Keywords: Distal, Phalanx, Lesion, Imaging

INTRODUCTION

Many lesions affect the distal phalanx with overlapping imaging features. Apart from a lucent appearance, erosions are perhaps the most common feature, rheumatoid arthritis and gout can be wrongly attributed as the underlying cause. Soft tissue tumors ranging from benign to malignant lesions also cause erosions giving rise to a wide range of differential diagnoses. Lysis is traditionally associated with more serious pathologies such as malignancy and infection but is also seen in a variety of chronic systemic conditions. Through our pictorial review, we hope to demonstrate the diversity in type and number of pathologies affecting the distal phalanx and how they appear on imaging. We have classified these cases into different etiologies, including neoplastic and non-neoplastic lesions.

DEVELOPMENT

Lesions involving short tubular bones of the hand generally parallel those in long bones. The radiographic findings and periosteal response are similar. Due to the size of the tubular bones of the hands; however, the distinction between epiphyseal, metaphyseal, and diaphyseal is not precise and much overlap is seen.[1]

The phalanges have two ossification centers: A primary ossification center and an epiphyseal center. The latter appears in the 2nd year and fuses with the shaft around the 15th–16th years in

females and a few years later in males.[1] The distal phalanges are unique as their proximal end has only one articular surface while their distal end terminates as a terminal tuft. The distal phalanx epiphysis ossifies slightly later than the middle and proximal phalanges; however, fusion is usually complete before that of the middle phalanx.

We provide a pictorial review of the different pathologies effecting the distal phalanx of the hand. The different radiological characteristics will be reviewed as well as the demographics and clinically relevant information for each lesion.

NEOPLASTIC LESIONS

Primary bone tumors

Enchondroma

Enchondroma is a benign, typically intramedullary tumor composed of hyaline cartilage. It is by far the most common tumor involving the distal phalanx accounting for 50% of all primary bone tumors in this anatomical region.[2]

When in the phalanx, lesions typically arise in the proximal metaphysis. On radiographs, enchondromas are typically seen as a well-defined central, lucent, expansile lesion without periosteal reaction or cortical disruption [Figure 1]. Chondroid matrix calcification tends to be seen in enchondromas of the long bones and is normally absent in hand lesions. There is often cortical thinning with various degrees of endosteal scalloping, but this tends to be mild when compared to other tumors. Most are asymptomatic but can present with pathological fractures due to severe cortical thinning.

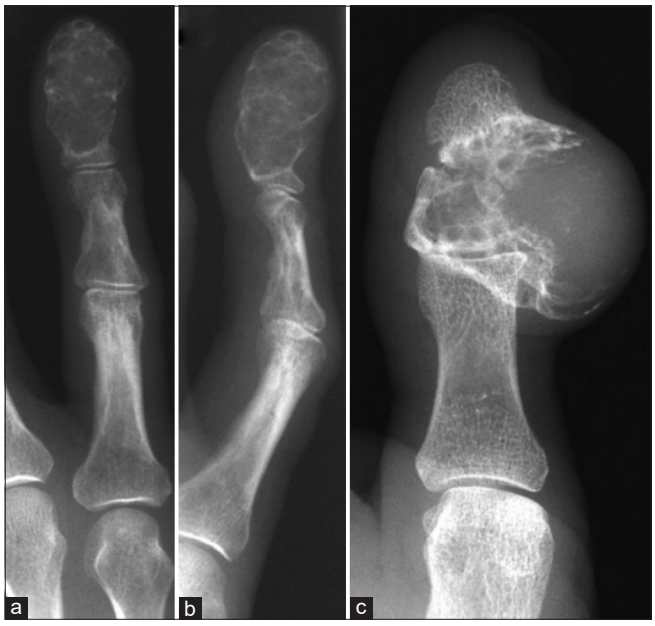

Figure 1: Enchondroma. Radiographs, AP (a, c) and lateral (b) demonstrate a lytic, expansile lesion in the distal phalanx.

Radiographic features suspicious for malignant transformation include cortical bone destruction, bone proliferation, and a soft tissue component. Differentiating enchondroma from low-grade chondrosarcoma can prove more troublesome, deep endosteal scalloping affecting at least two-thirds of the lesion is thought to be the most specific sign.[3] Magnetic resonance imaging (MRI) typically shows a low T1 and a very high fluid-like T2 signal with linear or globular enhancement pattern along the margins of the lesion. Most enchondromas are solitary and have a very low risk of malignant transformation, especially if in the hand. Multiple enchondromatosis, such as Ollier's disease, have a higher risk (up to 25%) of transformation into chondrosarcoma.[4]

As enchondromas are benign and asymptomatic, typically no follow-up is needed. For lesions that are persistently painful curettage and bone grafting are the main forms of treatment.

Subungual exostosis

A subungual exostosis is an osteocartilaginous tumor that typically affects the distal phalanx of the toes or fingers. As the name suggests, lesions arise in the subungual space beneath the nail bed. It is more common in females usually presenting between 10 and 20 years of age.[5] Patients are symptomatic with fingernail displacement, pain, soft tissue swelling, and potential for soft tissue infection.[5]

On radiographs, it presents as a broad-based osseous projection arising from the distal phalanx [Figure 2]. It may or may not have a well-defined bony cortex. Features distinguishing it from osteochondroma are a lack of continuity with the underlying medullary bone and lesions tend to have an overlying fibrocartilaginous rather than a hyaline cartilage cap. If symptomatic, MRI is recommended to look at how the exostoses affect neighboring structures. The fibrocartilaginous cap in subungual exostosis is hypointense on all MRI sequences, in contrast to the T2 hyperintense hyaline cartilage cap in osteochondroma.[6] Aggressive features have not been described and no cases of malignant transformation have been reported to our knowledge in literature.[5]

Soft tissue chondromas are a rare differential but they have a typical soft tissue appearance on USS with no contact with the underlying bone.[7] Keratoacanthoma is another differential, which radiographically presents as a soft tissue mass with osteolysis but with no periosteal reaction.[7]

Keratoacanthoma

Keratoacanthoma of the distal phalanx is typically seen in the subungual space. It is a rare but benign lesion consisting

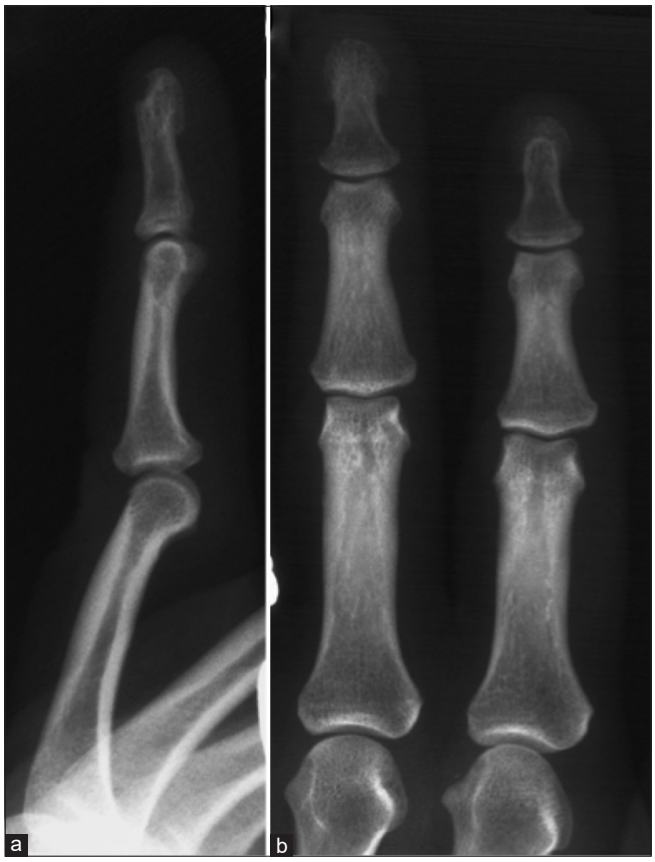

Figure 2: Subungual exostosis. Radiographs lateral (a) and AP (b) showing lack of continuity with the medulla distinguishes it from an osteochondroma.

of proliferating squamous epithelium filled with a keratin center.[8] It presents as a painful lump under the nail bed, which can increase in size. Unlike other forms of keratoacanthoma, the subungual variant shows little or no tendency to involute over time.[9] The main differential is squamous cell carcinoma; indeed keratoacanthoma can undergo malignant transformation to squamous cell carcinoma with subsequent bony invasion. Although radiologically both are virtually indistinguishable, squamous cell carcinoma is usually seen in older patients, typically in the seventh decade compared to the fifth decade in keratoacanthoma.[9] The latter also tends to grow quickly over a few weeks/months, whereas squamous cell carcinoma exhibits a chronic growth pattern.[10]

On plain radiographs, keratoacanthoma is seen a crescent-shaped soft tissue mass abutting the underlying bone.[11] There may be bony erosion with an osteolytic defect due to a pressure effect rather than direct tumor invasion.[11] Over time this can appear as a large lytic defect over the dorsal phalanx mimicking a metastatic deposit or squamous cell carcinoma [Figure 3].

USS usually shows a heterogeneous lesion with posterior acoustic enhancement. T1 and T2 signal intensity on MRI

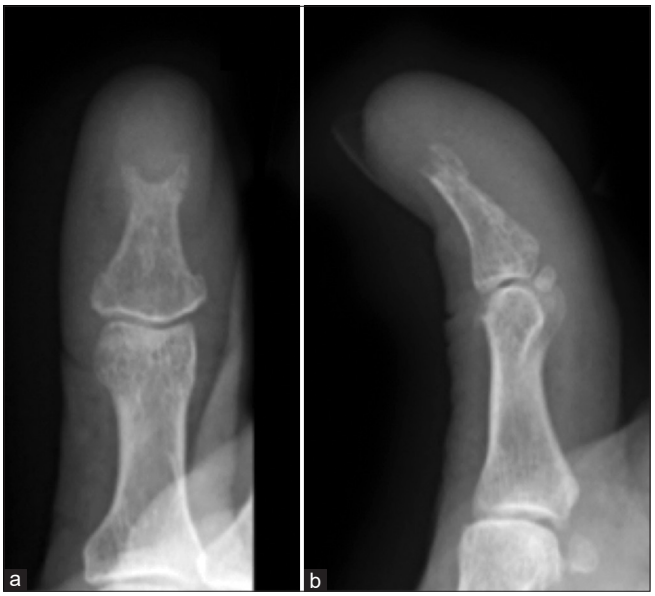

Figure 3: Keratoacanthoma. Radiographs AP (a) and lateral (b) showing crescent-shaped area of erosion and lysis affecting the terminal tuft of the distal phalanx of the thumb. Note the destructive nature of this lesion, mimicking squamous cell carcinoma.

is non-specific, but studies have reported thin peripheral enhancement post gadolinium contrast implying a surrounding inflammatory reaction.[11] Given the rather similar imaging features with squamous cell carcinoma, a biopsy is usually done. Keratoacanthoma is normally treated conservatively, with amputation usually required in squamous cell carcinoma.

Epidermal inclusion cyst

Epidermal bone cysts are not a true neoplasm but are a well-defined cystic cavity with keratin and a peripheral zone of squamous epithelium.[12] The origin is most likely trauma-related, where epithelial tissue can implant itself into soft tissue or bone at the time of trauma. They occur exclusively in the distal phalanges of the fingers, typically at the tuft. Lesions are normally asymptomatic until they present with a fracture or infection.

A lucent, well-circumscribed lesion is seen with a surrounding thin sclerotic cortical rim on plain radiographs [Figure 4]. There is no calcification and lesions may be expansile. However, not all epidermal inclusion cysts involve bone, and some are confined to the subcutaneous tissues. The key to differentiating this from other lesions in a similar location is that the nail bed is not involved and there is a history of previous trauma.[12]

Glomus tumor

Glomus tumor represents a soft tissue hamartoma of the glomus body. This is an arteriovenous anastomosis with

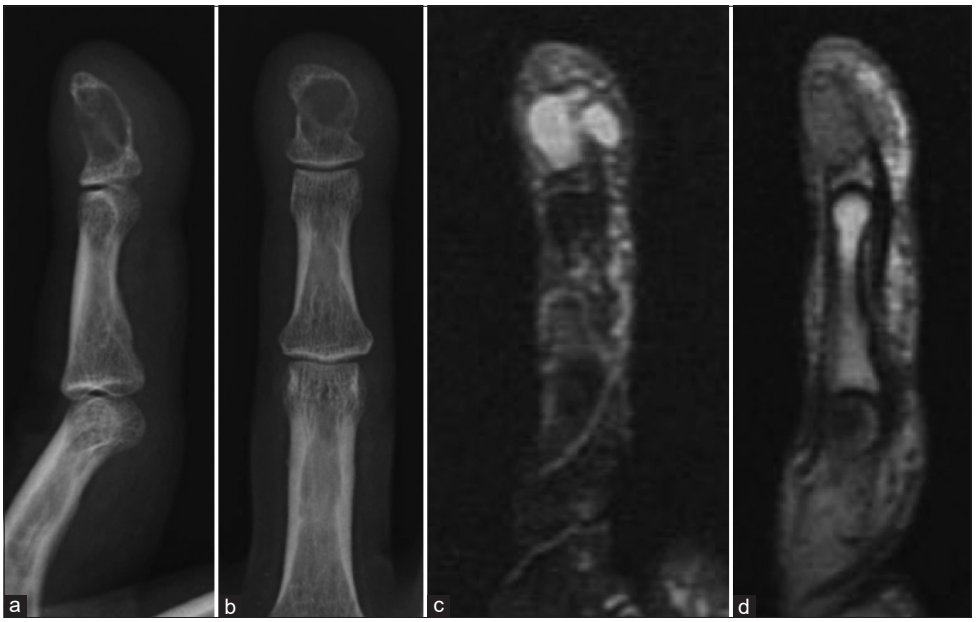

Figure 4: Epidermal inclusion cyst. Radiographs AP (a), lateral (b), sagittal T1(c), and STIR (d) showing a cystic lesion in the distal phalanx.

an intervening capillary bed underlying the fingertips that regulate local temperature and circulation.[13] The glomus body can hypertrophy appearing as a pale pink or purple mass under the nail bed. Glomus tumors are typically solitary, but multiple lesions have been associated with type 1 neurofibromatosis.[14]

As the tumor is extrinsic to bone, there may not be any radiographic findings as bony involvement is less common than pure soft tissue involvement. Bony abnormalities are typically pressure erosions, appearing as shallow, well-corticated osseous defects involving the distal phalangeal tufts adjacent to the tumor.[15] Completely intraosseous glomus tumors are generally located in the terminal phalanx and encased by cortical bone appearing as smooth bony expansions with a sclerotic rim [Figure 5]. MRI typically shows a high T2/STIR signal central dot surrounded by an area of lower signal, with enhancement post gadolinium injection. Although the signal characteristics are nonspecific, contrast enhancement can help differentiate glomus tumors from mucoid cysts and epidermoid inclusion cysts as the latter two are non-enhancing.[14] MRI has been shown to be particularly more beneficial over ultrasound in detecting small lesions, especially those as little as 2 mm.[14]

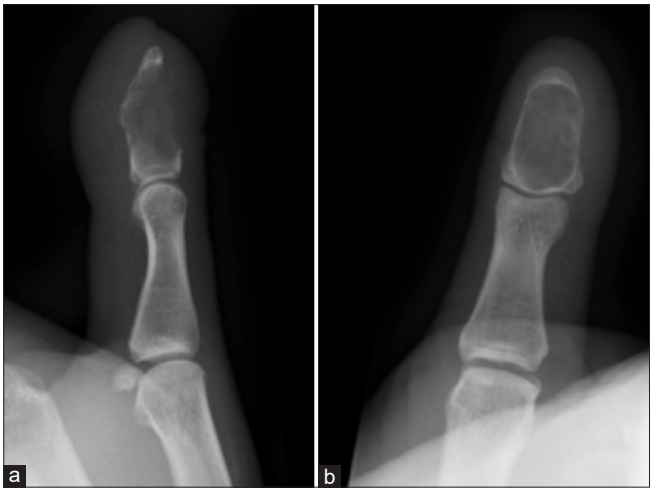

Figure 5: Radiographs lateral (a) and AP (b) showing well-defined lytic lesion in the distal phalanx typical of an intraosseous glomus tumor.

Schwannoma

Schwannomas of the hand are very rare but normally appear on the volar surface with only isolated case reports of a dorsal location.[16] Most present as a soft tissue mass but a few intraosseous cases and a subungual location have been reported in literature.[16] They present as a mass with or without paraesthesia along the flexor surface of the distal phalanx.

There are no specific features to suggest schwannoma on a plain X-ray. The most common findings are a suggestion of a lump in the subcutaneous tissues, which may cause scalloping or erosion of the underlying bony cortex [Figure 6]. On MRI, schwannomas tend to show low to intermediate T1 and high T2 signal with homogenous enhancement.[17] Larger lesions can display the "split fat sign," where there is a rind of fatty tissue around both poles of the lesion. Other signs include the "target sign," which is an area of central low T2 signal surrounded by high T2 signal.[17] However, most of

these findings are nonspecific, and there is no single reliable imaging feature to differentiate this from a neurofibroma.

Another differential for schwannoma confined to the distal phalanx is a digital mucous/mucoid cyst. These small fluid-filled sacs are essentially ganglion, which typically occurs at the distal interphalangeal joint near the nail bed. They follow fluid signal intensity on all MRI sequences with no appreciable enhancement; differentiating this from a glomus tumor. Mucous cysts are also very sharply defined, with a few reported cases showing internal septae.[18] The most common associated feature of mucous cysts are degenerative changes at the distal interphalangeal joint.[18]

Metastases

Metastases in hand are rare as most involve the axial skeleton due to the increased amount of hematopoietic activity. If seen in the hand, metastases show a predilection for the distal phalanx, due to the increased arterial flow in this location.[19] Bronchial carcinoma is the most common primary tumor involved (40%), followed by breast (16%), renal (6%), and prostate (3%).[20] Chondrosarcoma is the most common non-metastatic primary bone malignancy of the distal phalanx presenting as a lytic lesion with popcorn calcification, endosteal scalloping, and cortical thinning.[20]

Clinically, distal phalanx metastasis presents as a swollen and painful digit, often making it difficult to differentiate from infection. Typically, the history of a known primary tumor will make the diagnosis straightforward.

Radiographic appearances of peripheral metastases are usually of an osteolytic, destructive lesion [Figure 7]. A periosteal reaction is unusual and joint involvement is rare. In the absence of a known malignancy, the lesion may resemble osteomyelitis, septic arthritis or acute monoarticular rheumatoid arthritis. Unlike in infection, metastatic bony destruction of the digit is fairly local with relatively well preserved surrounding bone mineral density.[15] There can be an associated soft tissue mass. Characteristically, metastases do not cross the joint surface and so the articular cortex at the phalanx base is normally preserved, giving a "blown out," cortical shell appearance.[19]

NON-NEOPLASTIC LESIONS

Infection

Osteomyelitis

Osteomyelitis is often the result of direct inoculation from penetrating injuries or contiguous spread to the bone through soft tissues. Primary osteomyelitis of the distal phalanx is rare, with *Staphylococcus* being the most commonly involved organism.[5]

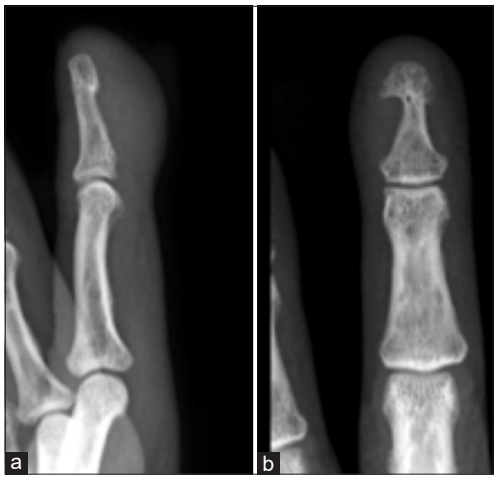

Figure 6: Schwannoma. Radiographs lateral (a) and AP (b) show a swelling at the volar surface of the distal phalanx, causing underlying bony scalloping.

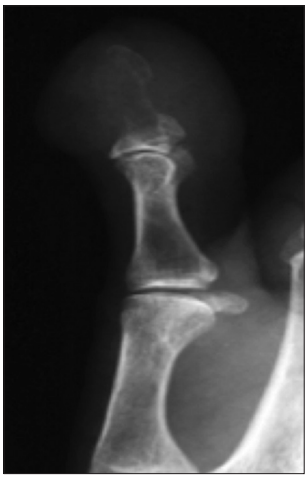

Figure 7: Metastasis from bronchial carcinoma. Lateral radiograph shows a lytic, destructive lesion in the distal phalanx.

Radiographs have low sensitivity and specificity for detecting acute osteomyelitis. Early radiographic changes include blurring of the soft tissue planes, followed by actual soft tissue swelling [Figure 8].[21] As time progresses there are established changes which include focal osteopenia, periosteal reaction, joint space narrowing, and bone destruction. In more chronic cases one may see a bony sequestrum (dead bone) within an involucrum (cavity) with subsequent cloaca (opening in an involucrum allowing drainage into soft tissues) formation.[21]

MRI is the most sensitive for early diagnosis, bone marrow edema as early as 1 day after the onset of infection can be detected.[22] Increased uptake can be delayed even up to a few days from the onset of infection in nuclear medicine studies.

Tuberculous osteomyelitis

Tuberculous dactylitis describes the painless involvement of the short tubular bones of the hands in tuberculous infection. Mycobacterium infection tends to have a predilection for the synovium and tendon sheaths compared to bone.[23] Early radiographic features are nonspecific and are mainly soft tissue swelling with or without periostitis. The classical pattern of bony mycobacterium involvement is the so-called Phemister triad of juxta-articular osteopenia, peripheral erosions, and gradual narrowing of the joint space.[24]

Although not always seen in the phalanx, there can be underlying bone destruction leading to a cyst-like central cavity and ballooning of the remaining bone, giving a wind-filled sail appearance termed "spina ventosa."[25] Other bony changes include coarsening of the trabecular pattern and acro-osteolysis of the distal phalanx [Figure 9]. However, many of these features are non-specific. The presence of sequestration and lack of osteopenia tend to favor pyogenic over the tuberculous infection.

Leprosy

Leprosy is a chronic infectious disease caused by *Mycobacterium leprae*. It affects the superficial skin causing peripheral nerve sensory loss with characteristic cutaneous plaques. Bony loss is very slow; hence, radiographic changes are often only seen in longstanding cases. Ten–30 years may elapse before there is a substantial shortening of the digital soft tissue and underlying bones.[26] Fusiform swelling of the overlying digital soft tissue is an early finding, as to is enlargement of the nutrient foramen. Early bony involvement is more commonly seen when there is superadded infection leading to ulceration.[27]

Leprosy usually affects the cancellous bony metaphysis of the proximal and/or middle phalanges with less frequent involvement of the metacarpal bones.[28] Direct *Mycobacterium leprae* infection results in trabecular destruction giving a "honeycomb and cystic" appearance on radiographs.[28] The terminal phalanges may be involved in two ways; directly by leprotic osteomyelitis and secondarily by neurotropic changes as a consequence of neural involvement by the infective granuloma. Neurotropic changes result in terminal tuft resorption giving a characteristic tapered "licked candy stick" appearance [Figure 10].[27] Chronic cases with the established disease over many decades radiographically tend to resemble a severely deforming polyarthritis.

AUTOIMMUNE DISEASES

Scleroderma

Scleroderma is a connective tissue disorder, which can cause skin tightness and decreased elasticity in the hands. Due to the impairment of local blood supply, there is often distal digital ischemia and severe sensory neuropathy.

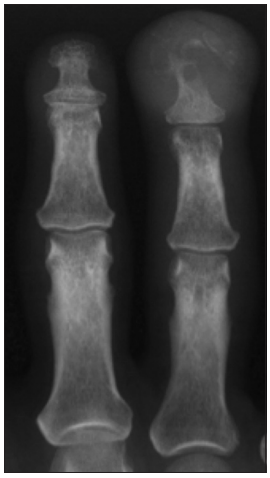

Figure 8: Osteomyelitis. AP radiograph demonstrates osteomyelitis of the distal phalanx secondary to a soft tissue infection with significant soft tissue swelling.

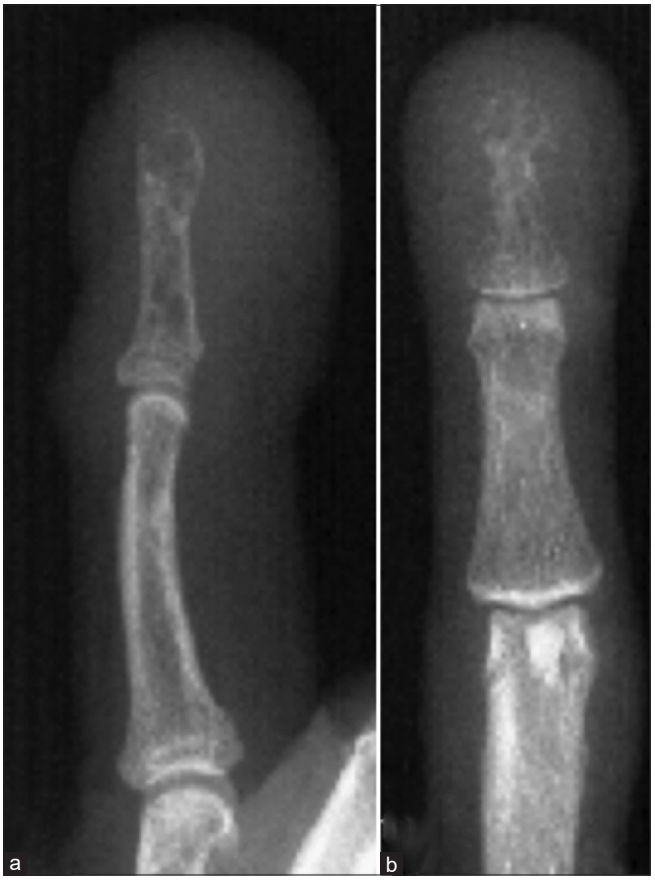

Figure 9: Tuberculous dactylitis. Lateral (a) and AP (b) radiograph demonstrates soft tissue swelling, osteopenia and small erosions of the distal phalanx.

Typically, radiographs show acro-osteolysis affecting the terminal tuft or the shaft of the distal phalanx (transverse or band acro-osteolysis) [Figure 11].[29] Bone resorption tends to start at the tuft and if advanced gives rise to the "penciling" appearance typical of acro-osteolysis where much of the distal phalanx may be destroyed with the fingers appearing tapered.[30]

Another characteristic radiographic feature of scleroderma is calcinosis [Figure 11]. This can either appear as amorphous calcium hydroxyapatite crystals in the subcutaneous tissues of the hand or digits. The presence of calcinosis tends to be associated with erosions and digital ulcers.[31] The combination of calcinosis and acro-osteolysis is thought to be pathognomonic for scleroderma. Observational studies interestingly report that more severe calcification tends to occur in the dominant hand, at sites of chronic stress such as the radial sides of fingers and soft tissue adjacent to bony protuberances implying a role of trauma in its pathogenesis.[31]

METABOLIC

Renal osteodystrophy

The radiographic features of renal osteodystrophy reflect hyperparathyroidism and deficiency of 1,25-dihydroxyvitamin D leading to osteomalacia, osteoporosis, soft tissue, and vascular calcifications.[32]

The most commonly reported radiographic finding apart from generalized osteopenia is subchondral bone resorption in more than one interphalangeal joint.[32] This is shown as areas of cortical and cartilaginous collapse due to osteoclastic resorption. Subperiosteal bone resorption is often seen in hyperparathyroidism associated with chronic renal disease and is present along the radial aspect of the phalanges, especially the middle phalanx of the index and middle fingers.[32]

Bone resorption in the phalangeal tufts results in loss of the cortical "white line" progressing to acro-osteolysis as discussed in previous examples [Figure 12].[33] In advanced disease, there can be a periosteal reaction due to parathormone osteoblastic stimulation. This is typically seen as linear new bone paralleling the cortical surface with a radiolucent zone between the two.[33]

Brown tumors are also radiographic features seen in chronic renal disease and appear as osteolytic lesions with an eccentric or cortical location occurring in the long bones of the hands.[33]

Acro osteosclerosis

Acro osteosclerosis refers to focal areas of sclerosis within the distal phalanx. This has a wide differential including

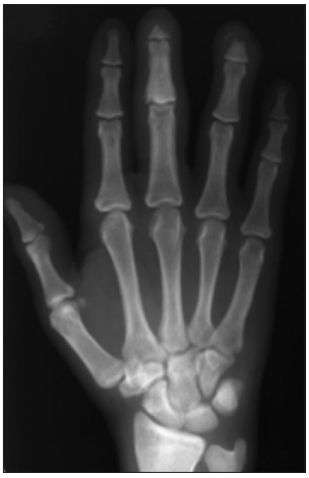

Figure 10: Leprosy. AP radiograph demonstrates bony sclerosis with resorption of the terminal tufts of the middle and ring fingers.

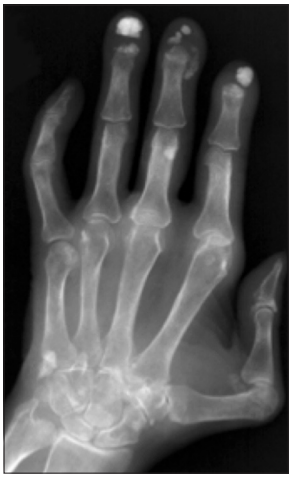

Figure 11: Scleroderma. AP radiograph of the hand shows acro-osteolysis and calcinosis in the distal phalanges.

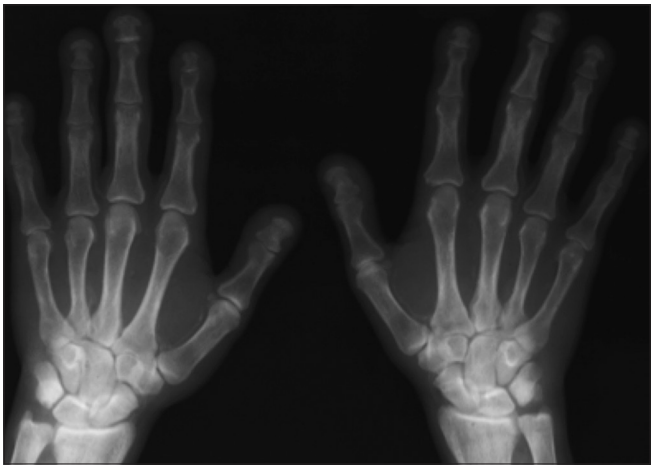

Figure 12: Acro-osteolysis. AP radiograph of both hands demonstrates acro-osteolysis of the terminal tufts in a patient with chronic kidney disease.

sarcoidosis, rheumatoid arthritis, Hodgkin's disease, and various hematological disorders.[34] It can also represent a long-term re-ossification process following acro-osteolysis. Diffuse osteosclerosis is seen typically in scleroderma, systemic lupus erythematous, and collagen vascular disorders.

In aging, the normal conical shape of the terminal tuft becomes more rounded and spades like appearing denser on radiographs. Several studies have shown that focal distal phalangeal osteosclerosis within the elderly is a common asymptomatic finding, especially in the mid and distal portion of the phalanx.[35] The severity of sclerosis has been formally categorized from Grade 0–4 by Halim and authors.[35] Grade 1 osteosclerosis is defined as a thickening of both bony cortices by more than 1 mm. The most severe form, Grade 4, consists of dense, compact bone affecting the entire terminal phalanx, including the epiphysis.

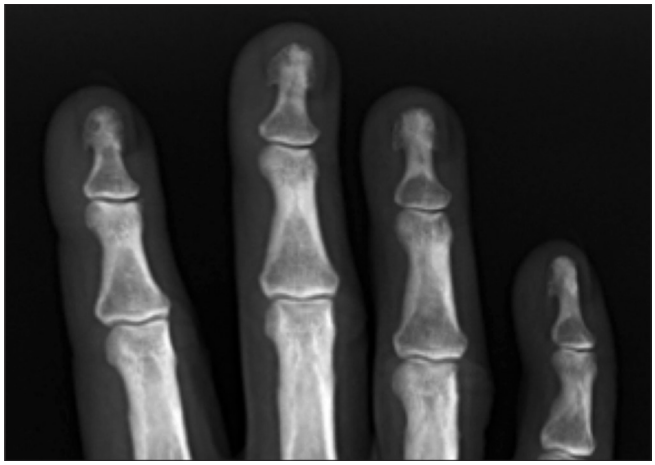

Figure 13: Acro-osteosclerosis. AP radiograph showing focal sclerosis across all four terminal tufts of the distal phalanx. Note how the terminal tuft itself has a well-defined margin despite the spade-shaped lobulations/irregularities associated with aging.

The main indicator of pathology is sclerosis affecting all or most of the terminal phalanges [Figure 13]. On plain radiographs, it is important to distinguish the normal aging appearance of a dense terminal tuft from a focal area of osteosclerosis. The terminal tuft, despite any irregularities, has a well-defined margin, which can be traced around and is in full continuation with the cortex unlike osteosclerosis. Osteoid osteoma should be considered in the differential for this. The presence of nidus on CT and florid osseous edema on MR enables to diagnose this entity [Figure 14].

Sarcoidosis

Sarcoidosis is a granulomatous disease affecting multiple organ systems. Nail and distal phalanx involvement is only usually seen in chronic disease states. Fingernail involvement presents radiographically as areas of thickening and irregularity.[36] If advanced, there may well be resorption of the underlying distal phalangeal tufts, with some even showing secondary acro-osteosclerosis.[35]

The most frequent intraosseous abnormality is a "lacy," lytic like appearance akin to small bone cysts [Figure 15].[36] These

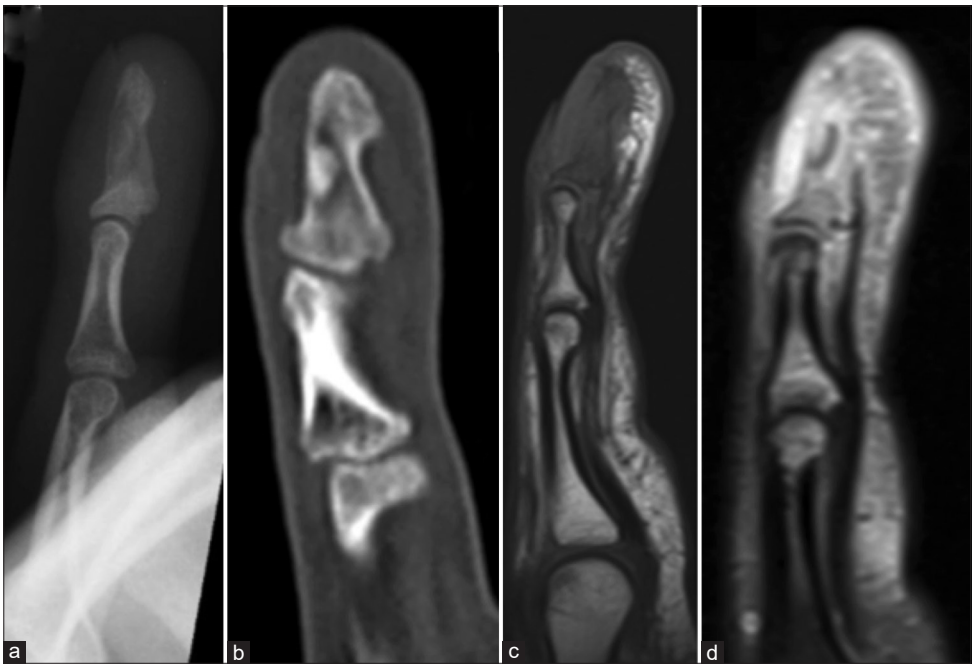

Figure 14: Osteoid osteoma. Lateral radiograph (a), sagittal CT (b), T1 (c), and STIR (d) showing osteoid osteoma of the distal phalanx with marked soft tissue and osseous edema.

cystic areas are typically reported as having a "honeycomb," trabeculation pattern sparing the articular joint surface.

The main differential for phalangeal sarcoidosis is gout. If the mentioned characteristic features are not present, then MRI may be helpful. Tophaceous gout is typically hypo or intermediate signal intensity on T2-weighted sequences; sarcoid nodules are usually of high T2 signal intensity.[37]

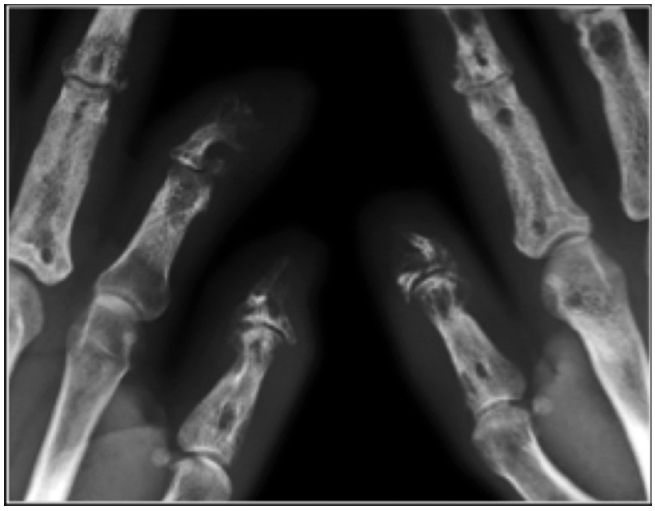

Figure 15: Sarcoidosis. AP radiograph demonstrates multiple punched-out lytic lesions throughout the phalanges sparing the articular surfaces. Note that some of the lesions have the typical honeycomb trabeculation pattern. There is also marked osteolysis affecting the terminal tufts with secondary remodeling.

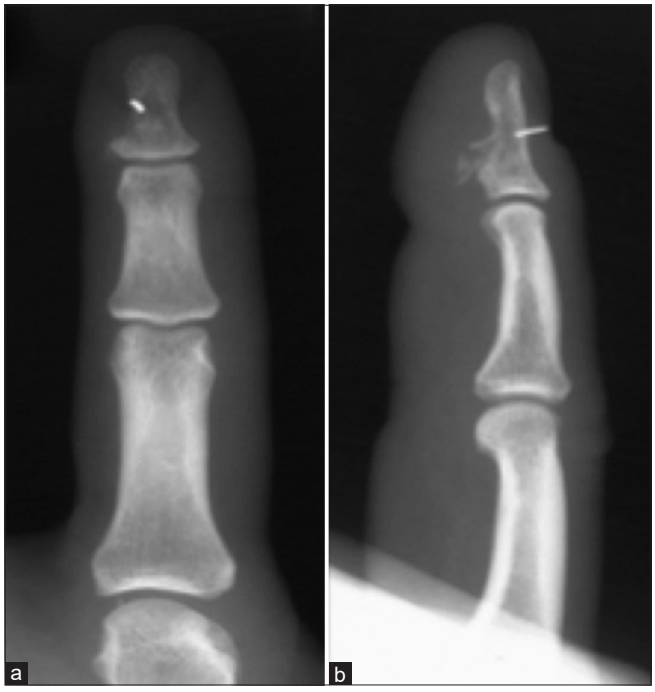

Figure 16: Foreign body. Radiographs AP (a) and lateral (b) demonstrates a radio-opaque foreign body in the projection of the soft tissue and distal phalanx.

FOREIGN BODY

Penetrating injuries to the hand are a common occurrence and may result in embedment of foreign bodies [Figure 16].[38] A thorough history makes the diagnosis straightforward. Radiographs with multiple views are advisable. Non-radiopaque foreign bodies may be detected by ultrasound aiding the surgeon in surgical removal.[39]

CONCLUSION

Lesions in the distal phalanx include neoplastic, autoimmune, inflammatory, and traumatic causes among a few. A surgical sieve can aid in providing a differential diagnosis. Although most lesions of the distal phalanx are benign, it is important to exclude a possibly malignant lesion or an underlying systemic condition.

Knowledge of the characteristic imaging features of some specific lesions can significantly narrow the differential diagnosis and allow appropriate referral for management. We have presented a myriad of common and rare cases and have outlined some differentiating features between them on plain radiographs. However, in some, only a "best diagnosis" can be given and a multimodality imaging approach possibly involving definitive biopsy needs to be carried out.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

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