

Systemic disease involvement of the hand: manifestations on plain radiograph

This educational pictorial review provides an appreciation of the range of radiographic findings seen on a hand radiograph that contribute to a diagnosis of systemic disease, whether in clinical practice or the exam scenario.

In systemic disease, where there is arthralgia, there may occasionally be accompanying structural change to the hands. There may also be pathognomonic findings that enable the diagnosis to be reached after review of a single hand radiograph. This is most common in metabolic diseases, but is also seen in some infective, malignant, autoimmune and granulomatous conditions. This educational pictorial review looks at fourteen such conditions,

drawing out their characteristic radiological features and summarizing the pathological processes involved.

Rheumatoid arthritis

Rheumatoid arthritis is a chronic systemic connective tissue disease principally involving synovial joints and characterized by peri-articular erosions. The hands and wrists are a focal target area. Typical features are of a symmetrical, deforming polyarthropathy, principally of the proximal joints: the distal radio-ulnar joint (especially the ulnar styloid), radio-carpal, mid-carpal and the metacarpophalangeal joints (*Figure 1*). Most patients have radiological evidence of erosions by 3 years (Dixey et al, 2004). There is synovial swelling which is followed by joint space narrowing and peri-articular demineralization secondary to disuse. Erosions occur primarily at the periphery of the joint, where cortical bone is not protected by cartilage. Over time, joints may sublux as a result of flexion and extension contractures, leading to deviation of the fingers, typically towards the ulnar aspect. Bone fusion and gross deformity heralds the end stage of the disease.

Psoriatic arthritis

Psoriatic arthritis is a common seronegative spondyloarthritis which affects 20% of patients with psoriasis. It is characterized by signs of inflammatory arthritis,



Figure 1. Rheumatoid arthritis. a. Right hand radiograph shows proximal osteoarthritis and ulnar subluxation of the fifth metacarpophalangeal joint. Peri-articular osteoporosis, marginal bone erosions and subluxations are typical features. b. Left wrist radiograph demonstrates concentric joint space narrowing and fusion of the carpal bones of the left wrist (late signs). Also, changes in the ulna styloid and distal radioulnar joint (early signs). The patient also had a flexion deformity at the metacarpophalangeal joints.

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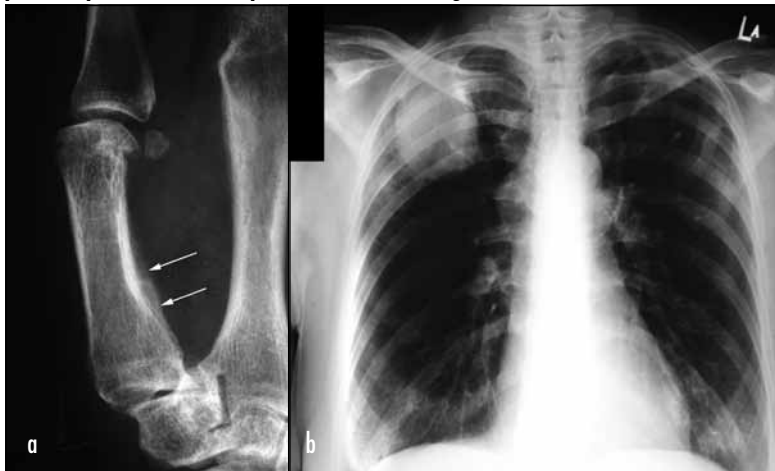
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Figure 2. Psoriatic arthritis. a. Right index finger radiograph shows distal interphalangeal joint erosion with characteristic ‘pencil in cup’ deformity (ill-defined erosion with adjacent periosteal new bone formation). b. Left middle finger radiograph of the same patient shows distal interphalangeal joint erosion with enthesitis and new bone formation at the distal phalanx (arrows). Soft tissue swelling of the digit (‘sausage digit’) is also noted.

enthesitis, periostitis and a predilection for the distal joints of the hands and feet. Erosions tend to occur at the margins of the distal interphalangeal joints causing a pattern of destruction such that the terminal phalanx may sit at an angle resembling a pencil resting in a cup (Figure 2). Bone proliferation is also a feature; periostitis may occur throughout the phalanges, bilateral and asymmetrical, and may be associated with soft tissue swelling giving rise to the typical ‘sausage digit’ appearance (Jacobson et al, 2008). Occasionally acro-osteolysis of the distal phalanx is also seen. Bone mineral density is preserved.

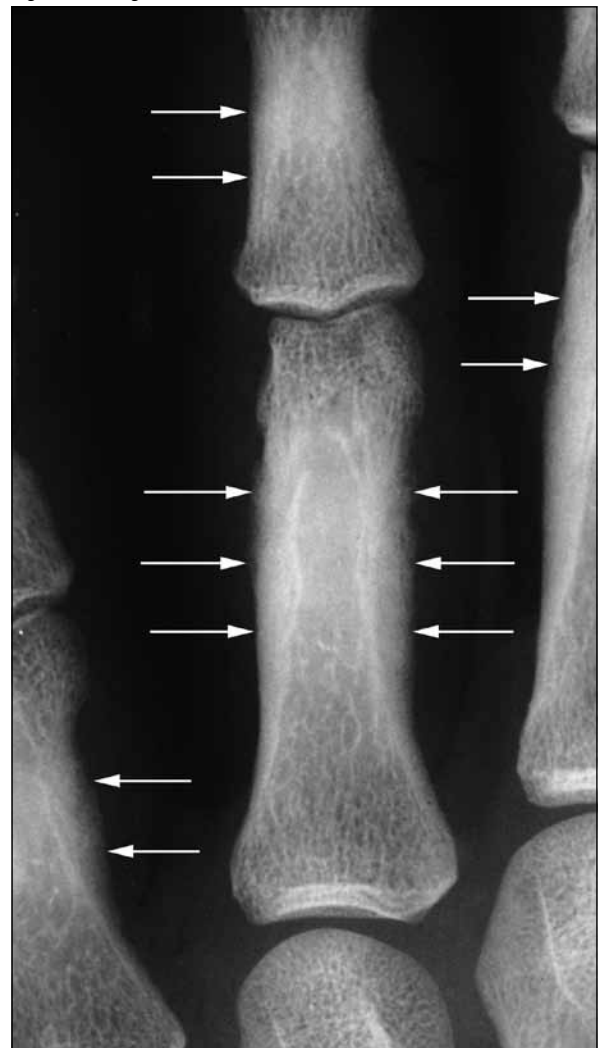
Figure 3. Hypertrophic pulmonary osteoarthropathy. a. Radiograph of the right thumb shows smooth laminar periosteal proliferation of the proximal metacarpal (arrows). The most common site of hand involvement is the ulnar aspect, peri-metaphyseal region of the proximal phalanx. b. Chest radiograph of the same patient shows a right upper lobe pulmonary mass lesion that proved to be a bronchogenic carcinoma.



Hypertrophic osteoarthropathy

The most common of the hypertrophic osteoarthropathies is hypertrophic pulmonary osteoarthropathy, a paraneoplastic syndrome secondary to carcinoma of the lung (Figure 3). However, hypertrophic osteoarthropathy can also develop alongside cardiac, hepatic or intestinal diseases. The most common site of skeletal involvement is in the radius and ulna (80%). In the hands, a bilateral, symmetrical polyarthropathy is seen with proximal phalangeal periostosis (60%) and soft tissue swellings (‘clubbing’). The idiopathic form, pachydermoperiostosis is a rare genetic disorder, characterized by the triad of skin thickening (pachydermia), finger clubbing and increased bone formation (periostosis) (Figure 4). There is little difference between the distribution of the periostosis in the hands of idiopathic and secondary osteoarthropathies (Pineda et al, 1987).

Figure 4. Pachydermoperiostosis. Radiograph of the ring finger of the left hand showing widespread irregular periosteal proliferation of the phalanges (arrows). There is cortical thickening without narrowing of the medulla. The patient had skin thickening and digital clubbing.



Scleroderma

Scleroderma (or progressive systemic sclerosis) is a multi-system connective tissue disorder of unknown aetiology. Musculoskeletal symptoms are very common with fatigue and myalgia accompanying arthralgia. In both limited and diffuse types, the majority of hand radiographs will show absorption of the distal phalangeal tuft (acro-osteolysis) which is thought to be the result of the high pressure imparted by the widespread sclerotic tissue. The result is thinning of the bone distally. Peri-articular osteopaenia and erosions are typical. Subcutaneous calcifications are also seen, particularly on the extensor surfaces of the hands (*Figure 5*). The combination of acro-osteolysis, articular erosions and subcutaneous calcinosis is strongly suggestive of the condition (Bassett et al, 1981).

Figure 5. Scleroderma. Radiograph of the radial aspect of the right hand shows acro-osteolysis of the phalangeal tufts (arrow A), and soft tissue calcifications (arrow B). There is peri-articular osteopaenia. Additionally, erosions are commonly seen at the interphalangeal and metacarpophalangeal joints.



Dermatomyositis

Dermatomyositis is an autoimmune inflammatory myopathy, with diffuse non-suppurative inflammation of striated muscle and skin, and a female predilection. It is characterized by gradual onset muscle weakness, elevated muscle enzymes and elevated myositis-specific antibodies (anti-Jo-1). Although predominant symptoms are those of muscle tenderness, weakness and skin changes, dermatomyositis may also cause a non-deforming arthritis with swelling of the distal joints of the hands. Radiographic features include cutaneous calcific deposits (40%) and, rarely, acro-osteolysis (more common in scleroderma). The calcific deposits are usually more 'linear, reticular and lacy', and more widespread than those seen in scleroderma (Blane et al, 1984). They are found in the soft tissues of extremities as well as overlying large joints, chest and abdominal wall (especially in younger patients) (*Figure 6*). Unlike scleroderma, osteopaenia is not a feature.

Thyroid acropathy

Acropathy ('thickening of the extremities') occurs as one of the extrathyroid manifestations of autoimmune thyroid disease. Most patients with acropathy have an initial diagnosis of Graves' disease although acropathy may also occur in patients with Hashimoto's thyroiditis and euthyroid Graves' disease. It is more likely to be seen in patients treated with radioiodine ablation for hyperthyroidism, and interestingly is much more common in smokers. It is almost always seen with ophthalmopathy. Solid periosteal new bone formation is seen along with finger clubbing and swelling of fingers and toes. It is usually bilateral, relatively symmetric and painless, and involves the diaphyseal tubular bones of the hands and feet. Radiographs

Figure 6. Dermatomyositis. a. Radiograph of the index finger of the right hand showing confluent digital soft tissue calcifications. No peri-articular erosions. b. Radiograph of the pelvis of the same patient showing extensive sheets of subcutaneous calcification.



typically demonstrate thick, spiculated ('feathery'), periosteal reactions (Rana et al, 2009) (*Figure 7*). Conversely the periostitis seen in hypertrophic osteoarthropathy is smoothly layered and usually tender. The long bones and peri-articular areas are less frequently involved than in other conditions, such as hypertrophic pulmonary osteoarthropathy and rheumatoid arthritis. Patients are often asymptomatic but may suffer with arthralgia of the small joints.

Hyperparathyroidism

Hyperparathyroidism is the uncontrolled production of parathyroid hormone which usually leads to an increase in serum alkaline phosphatase and calcium levels (as a result of accelerated bone turnover and increased calcium absorption) and a decrease in serum phosphate levels. Bone pain and tenderness may be the presenting features. Hand radiographs often show osteopaenia and bone demineralization. The most pathognomic finding, however, is subperiosteal bone resorption (Khan and Bilezikian, 2000). This is most marked along the radial aspect of the middle phalanges of the index and middle fingers (*Figure 8*). Early findings are of proximal metaphyseal cortical irregularity. Chronic features include spiculated cortical scalloping (may involve the entire digit), acro-osteolysis and in rare cases, lytic lesions

Figure 7. Thyroid acropachy. Radiograph of the right hand showing thick 'feathery' periosteal reaction of the index and middle fingers. The mid-portions of the diaphyses are typically affected.



caused by accumulations of osteoclastic giant cells known as 'brown tumours.'

Acromegaly

Acromegaly is the result of excessive growth hormone production, most commonly from a macroadenoma of the pituitary. It has a slow and insidious onset and is usually diagnosed in adults only when the external features become apparent. Radiographs may reveal osseous enlargement, presenting with 'spade-like' hands (*Figure 9*) and widening of the terminal phalangeal tufts, giving an 'arrow-head' appearance (Chapman and Nakielny, 2009). Other features in the hand may include prominent muscle attachments and widened joint spaces (as a result of cartilage hypertrophy). Generalized osteoporosis and cystic changes in the carpal bones are also described.

Marfan syndrome

Marfan syndrome is an inherited multi-systemic connective tissue disease caused by a fibrillin gene defect resulting in abnormal cross-linking of collagen fibres. Cardiac diseases such as aortic regurgitation, dissection or rupture and mitral insufficiency are substantial contributors to mortality. Musculoskeletal manifestations include scoliosis, posterior vertebral scalloping, pes planus, ligamentous instability, osteopaenia and muscular hypoplasia. The hands demonstrate arachnodactyly (*Figure 10*) (elongation of the phalanges) and protrusion of the thumb beyond the confines of the clenched

Figure 8. Hyperparathyroidism. a. Radiograph of the index finger of the right hand showing subperiosteal bone resorption of the radial aspect of the entire digit but particularly affecting the middle phalanx. b. Radiograph of the middle finger showing more developed features of hyperparathyroidism including acro-osteolysis (arrow A) and coarse spiculated cortical reaction (arrows B). This involves the entire digit but is more marked on the radial aspect.



fist (Steinberg sign). A metacarpal index (averaging the four ratios of length of the second to fifth metacarpals divided by their respective mid-diaphyseal width) greater than 8.8 for males or 8.4 for females is suggestive of arachnodactyly (Magid et al, 1990) and where recognized should lead to further investigation of the aortic root using echocardiography, computed tomography or magnetic resonance imaging.

Figure 9. Acromegaly. Radiograph of the left hand showing widening of the terminal phalangeal tufts with a 'arrow-head' appearance. The hand is enlarged and soft tissue swelling is noted.



Figure 10. Marfan syndrome. Radiograph of the hands showing elongation of the phalanges and metacarpals suggestive of arachnodactyly (metacarpal index over 8.4).



Haemochromatosis

Haemochromatosis is an iron overload disorder characterized by the pathological deposition of iron in tissues eventually resulting in dysfunction. Symptoms of either liver cirrhosis, cardiomyopathy, hypopituitarism, diabetes or arthritis declare themselves usually by the 3rd to 5th decades. The arthropathy, caused by iron deposition in synovium, is something of a hybrid between degenerative and inflammatory arthritides. It causes uniform symmetric joint space narrowing, particularly of the metacarpophalangeal joints, which is unusual for degenerative joint disease and does so without causing erosions. Hook-like osteophytes on the radial aspect of the metacarpal heads (especially the second) are characteristic. There may also be chondrocalcinosis (Figure 11), seen in the hands and within the triangular cartilage of the wrist, in up to 50% of cases (Dähnert, 2007).

Gout

Gout is a crystal deposition arthropathy caused by the deposition of monosodium urate crystals (tophi) in and around the joints. Presentation may be within the acute phase with soft tissue swelling and joint effusion, or within the chronic tophaceous phase (6–12 years after the initial attack) with eccentric asymmetrical nodular tophi within the synovium and subchondral bone (Figure 12). Bony erosions may be intra-articular, peri-articular or distinct from the joint with a rounded or 'punched out' appearance. The erosion margins are typically thin and sclerotic with a raised, overhanging, cortical lip, giving a 'rat bite' appearance. This characteristic late finding is caused by peri-articular tophaceous nodules and erosion. Severe erosive changes may lead to an arthritis mutilans (Chapman and Nakielny, 2009). Chondrocalcinosis is seen in just 5% of cases and is more commonly associated with pseudogout. Little or no osteopaenia is seen until late in the disease.

Figure 11. Haemochromatosis. Radiograph of the left wrist showing chondrocalcinosis of the triangular fibrocartilagenous complex (arrows).



Filariasis

Loa loa filariasis (Loiasis) is a cutaneous parasitic infection endemic to central and western Africa. Transmission from an infected host is via the bite of a mosquito. The larvae migrate into the puncture wound and reach the subcutaneous lymphatic vessels. The onset of lymphatic filariasis is slow and patients may have febrile or inflammatory episodes. These may represent either systemic acute adenolymphangitis, filarial fever or tropical pulmonary eosinophilia. Manifestations of infection include transient localized subcutaneous swellings (known as Calabar swellings) and painful migration of the adult worm across the subconjunctiva of the eye.

The dead Loiasis parasite calcifies and may be seen on plain film of the hand as hair-like whorls of calcification or beaded serpiginous calcification (Figure 13). This may be differentiated radiographically from other types of filariasis such as *Wuchereria bancrofti*, which appears tortuous and wavy (more commonly in breast parenchyma), and trichinosis which is caused by smaller, more numerous and not so serpiginous roundworms (more commonly found in the pectoral muscles) (Friedman and Kalisher, 2002). A tentative diagnosis on plain radiograph can be supported with a polymerase chain reaction or a blood smear.

Figure 12. Gout. Radiograph of the index and middle finger of the right hand showing periarticular soft tissue swellings (arrows A) and eccentric 'punched out' lytic bone erosions with thin sclerotic margins and overhanging edges (arrows B). There is preservation of the interphalangeal joint spaces and absence of periarticular demineralization (features that respectively help to differentiate gout from psoriatic or rheumatoid arthropathy).



Tuberculous dactylitis

Dactylitis is an inflammatory disorder of the digit (hand or foot) associated with seronegative arthropathies, such as psoriatic arthropathy and ankylosing spondylitis, in sickle cell disease as a result of vasoocclusive crisis with bone infarcts, and infectious conditions including tuberculosis and leprosy. Tuberculous dactylitis usually presents insidiously as a painless swelling in the hand or foot. Plain film findings are of a diaphyseal lytic lesion, either round and cystic with a variable amount of sclerosis and minimal periostitis or ill-defined with minimal or no surrounding sclerosis and thick lamellated periostitis.

Sickle cell dactylitis may present silently or with a painful bone crisis, and radiographs in young children similarly show diaphyseal lytic lesions (Figure 14) within the tubular bones of the hands or feet (Ejindu et al, 2007). Periostitis is uncommon.

Sarcoidosis

Sarcoidosis is a chronic granulomatous disease of unknown origin. It most commonly involves the lungs and lymphatics although any organ can be affected. In 15–25% of cases arthritis is a feature, more often as a

Figure 13. *Loa loa* filariasis. Radiograph of the ring and little finger of the right hand showing fine beaded subcutaneous calcifications in a tubular serpentine configuration, typical for *Loa loa* infection (arrows).



result of the effect of cytokines on joints rather than the direct deposition of granulomas. It is most commonly an oligoarthritis with the ankle joint most likely to be affected at presentation. If the small joints of the hand are affected it is usually the proximal interphalangeal joints as part of a symmetrical polyarthritis (Visser et al, 2002). The pattern of the arthritis follows one of two courses: the non-granulomatous type is more common and follows an acute transient course with a generally good prognosis. The granulomatous type (10–35%) occurs later in the disease and runs a more chronic, relapsing course with dactylitis a striking finding (Moore and Teirstein, 2003). Although the deposition of granulomas tends to be less painful than the synovial inflammation, they can cause lytic lesions leading to pathological fractures and alignment deformities (Figure 15).

Conclusions

A thorough appreciation of the radiographic features of systemic disease in the hand is necessary for the rheumatologist and radiologist alike. These illustrated examples provide a diagnostic framework when faced with a single radiograph. **BJHM**

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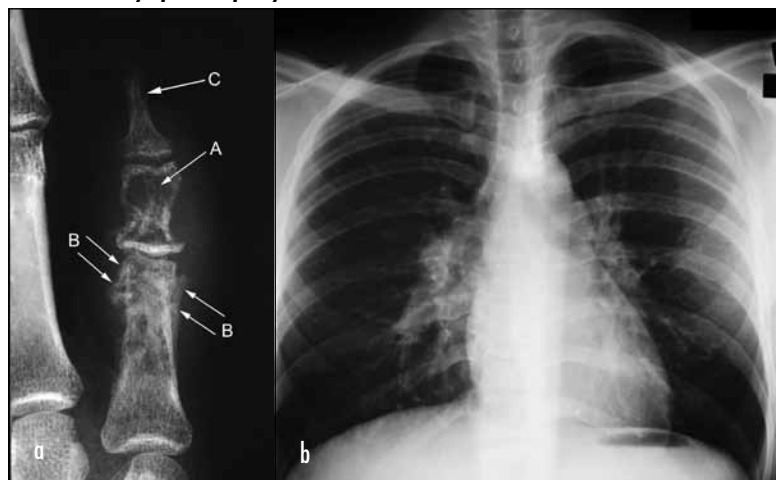
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Figure 14. Tuberculous dactylitis. a. Radiograph of the little finger of the right hand showing a rounded lucency in the proximal phalanx of a child with sickle cell anaemia. There is minimal periostitis. b. Radiograph of the right elbow of the same patient showing a well-defined lytic lesion in the ulna with no surrounding sclerosis and a less well-defined lucency in the distal humerus. There is overlying periostitis and a joint effusion. Sickle cell disease is a risk factor for tuberculosis infection, and this patient had numerous tuberculous abscesses in the liver and spleen.



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Figure 15. Sarcoidosis. a. Radiograph of the little finger of the right hand showing 'lace like' metaphyseal trabecular patterns and well-defined cyst like lesions (arrow A), phalangeal endosteal sclerosis and periosteal new bone formation (arrow B) and destruction of the terminal phalanx (arrow C). Close inspection reveals a pathological fracture through the base of the middle phalanx. b. Chest radiograph of the same patient shows bilateral lymphadenopathy.



KEY POINTS

- Plain films are commonly performed to investigate pain, swelling or deformity of the hand.
- The recognition and interrogation of radiographic signs such as erosions, subluxations, demineralization, soft tissue swellings and calcifications may lead towards the diagnosis of a systemic disease.
- When characteristic findings are present, the diagnosis of a systemic disease can confidently be made on a single radiograph.