MSK Findings in Systemic Sclerosis - Multi Modality Approach

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Learning objectives

The aim of this poster is to summarize and display MSK findings in systemic sclerosis in soft tissues, joints and extraarticular bone as imaged by the appropriate imaging modalities, in order to be able to help the clinicians establish the diagnosis and get a complex information on the extent of the disease in each patient.
Background

Systemic sclerosis (scleroderma, SSc) is a generalized autoimmune connective tissue disorder, affecting the musculoskeletal system, multiple visceral organs (heart, lungs, GIT, kidneys) and vessels.

The common pathophysiologic change in all the organs is fibrosis and angiopathy. The main pathophysiologic mechanism of this entity is a chronic inflammatory reaction of collagen fibers, leading to their atrophy, to the reduction of elastic fibers of the connective tissues, reduction of hairs and skin glands, to pronounced fibroblast activity and thickening of endothelial basal membrane. All this is accompanied by elevated sedimentation rate and C-reactive protein, hypergammaglobulinaemia, specific antinuclear antibodies, thrombocytopenia and hemolytic anemia.

5 subtypes of SSc are recognized: Diffuse, localized, transitory between the two, sine scleroderma and malignant.

In the musculoskeletal system it can be encountered a) in the skin and subcutaneous tissue, b) in muscles, tendons and fascias, c) in peripheral bone prominences and d) in and around the joints - affecting the synovium, cartilage, bone and the periarticular soft tissues.

In the heart it produces direct sclerosis of the muscle in combination with cor pulmonale and arhythmias.

In the lungs it causes changes from fine reticulation to honeycombing, occasional pleural reaction and secondary alveolar infiltrates due to aspiration, yet the hallmark is histological - thickening of basement membrane of alveoli and small vessels, accounting for severe functional impairment and hemoptysis.

In the GIT it causes dysphagia, dilatation and dysmotility of the entire tract - esophagus, stomach and bowel, resulting in abdominal pain, constipation/diarrhea and malabsorption.

In the kidneys it causes constriction and occlusion of the afferent arterioles, leading to renal failure.

In the vessels it most often presents as Raynaud’s phenomenon - severe recurring vasospasm (occasionally occlusion) of peripheral arteries of the fingers and toes.

SSc may be part of the CREST syndrome, composed of calcifications, Raynaud’s phenomenon, esophagus involvement, scleroderma and teleangiectasia.
SSc tends to involve the following regions of musculoskeletal system:

**Involvement of skin and subcutaneous tissue**

In SSc both the skin and the subcutaneous fatty tissue are generally thickened due to edema and/or fibrotic infiltration - first in the hands and in the face, later in other parts of the body. This can be well imaged as widened, hypo/hyperechogenic skin and subcutaneous fat with loss of internal structure or on the contrary with thickened interlobular septae in US (Fig.1a-d) or as subcutaneous septae hypointense in T1-weighted images and hyperintense in T2-weighted images in MRI (Fig.1e). Fig. 1 on page 7

However, in the fingertips subcutaneous fatty tissue is typically thinned (Fig 2a, b), often in coexistence with Raynaud’s phenomenon. (The normal width of the soft tissues should be at least 20% of the width of the base of the distal phalanx.) (Fig. 2c) Fig. 2 on page 7 The pathognommonic feature in the subcutaneous tissue is the appearance of calcifications - in plain radiography as tiny solitary calcifications or amorphous cloud-like structures called "tumoral calcinosis". Calcifications can be encountered in up to 50% of cases in the dominant hand - in the fingertips (Fig.3a, b) together with soft tissue atrophy and acroosteolysis, or in other places of elevated pressure (Fig.3c). Fig. 3 on page 8 Periarticular calcifications can also be part of the joint involvement as will be discussed later. In US their acoustic shadow may or may not be present (according to their size and density) (Fig. 3d, e), in MRI they tend to be accompanied by a T1- and T2 - weighted hypointense scar.

**Involvement of muscles, tendons and fascias**

Involvement of these structures can be encountered in MRI in up to 70-96% cases, by far exceeding the clinical suspicion. It is caused by myositis and fasciitis (inflammatory infiltration) or myopathy and fascial thickening (fibrous changes). In these cases cardiomyopathy may be present.

The MRI images reveal usually bilateral symmetrical enhancing edema of the muscles, esp. perifascial muscle bundles, thickened edematous fascias and tendon entheses, all predominantly in the lower extremities (Fig.4). Fig. 4 on page 9 Plain radiography and CT reveal enthesophytes and calcifications in the tendons and US reveals tenosynovitis as fluid in the tendon sheath (Fig.5). Fig. 5 on page 10 However, tendon rupture is extremely rare.

**Involvement of extraarticular bone**
Two types of findings can be encountered in the extraarticular bone: typical acroosteolysis and a completely non-specific osteoporosis.

As already mentioned, peripheral soft tissue thinning and calcifications tend to occur together with acroosteolysis- most often in the fingertips, but also in other prominent bony structures, such as the styloids, carpal bones, lateral clavicles, ribs, mandible or spinal processes. The morphology of this osteolysis is usually pencil shaped - pointed osteolysis, less frequently band-like osteolysis separating the proximal and distal parts of the phalanx. The best imaging modality in these cases is the plain radiography (Figs. 6 and 7a). Fig. 6 on page 11 In MRI T1- and T2 - weighted images hypointense scar can be found in this region.

Rarely hyperostosis of the terminal phalanx has been reported (Fig. 7b).

Diffuse widespread non-specific osteoporosis can be found in up to 23% of cases (Fig. 7c). Fig. 7 on page 12

**Involvement of joints and periarticular soft tissues**

Joints can be affected both in their bony parts as well as in their soft tissue parts and periarticular zones.

40-100% of joints have been reported to reveal arthralgia and synovitis as demonstrated by US and MRI in the form of intraarticular synovial fluid content, often even in clinically unsuspected cases (Fig. 8). Fig. 8 on page 13

Inflammatory arthritis affects predominantly the joints of the hand (proximal and distal interphalangeal joints, metacarpophalangeal joints and carpal joints), including the carpometacarpal joint of the thumb, which is otherwise very rarely involved in other than degenerative disease. In cases of involvement of the metacarpophalangeal joints and carpal joints an overlap syndrome with rheumatoid arthritis should be considered. The morphology and severity is that of a wide range of changes - from a completely normal finding, through isolated joint space narrowing, up to changes resembling erosive osteoarthritis or psoriatic arthritis - i.e. joint space narrowing, erosions, subchondral osteosclerosis, enthesopfytes, osteophytes and periosteal reaction. Infrequently a purely lytic arthritis can be encountered. Calcifications, so typical for SSc, can be found also intraarticularly and in the joint capsule (Fig. 9). Fig. 9 on page 14 However, other joints can be involved as well - most often the feet, less often large joints and spinal joints (Fig. 10). Destructive arthritis can end up in bony ankylosis (Fig. 10b). Fig. 10 on page 15

Apart from synovitis and arthritis, all of the above mentioned joints can reveal periarticular soft tissue involvement, which includes again calcifications (in periarticular ligaments) (Fig. 11), Fig. 11 on page 16 flexion contractures and subluxation / dislocation (due to fibrous changes of the skin and subcutaneous tissue) (Fig. 12). Fig. 12 on page 17
Fig. 1: US - a: thickening of the skin (*), b, c and d: mild to pronounced thickening of the subcutaneous fat in the wrist with loss of structure and hyperechogenicity of the tissue or with coarse septations (#). T2-w FS MRI - e: coarse hyperintense interlobular septations.

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Fig. 2: Plain radiography - a, b: soft tissue atrophy in the fingertips, together with acroosteolysis and occasional calcifications. c: plain radiography based scheme of minimal soft tissue width in the fingertips.

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Fig. 3: Plain radiography - a, b, c: calcifications in the soft tissues of the fingertips and wrist respectively - discreet and tumoral calcinosis. US - d and e: calcifications in the subcutaneous fat.

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**Fig. 4:** MRI - myositis and fasciitis: b: symmetrical T2FS hyperintensity of the muscles of the pelvis and d: fascias of the thigh, a and c: without fatty degeneration.

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**Fig. 5:** US - a: tenosynovitis of the flexor tendon (fl), fluid (*) and thickened synovium (S) in the tendon sheath. Plain radiography - b, c, d: enthesophytes in the base of the patella, in the medial maleolus and calcificatons in the greater trochanter and spina iliaca anterior inferior respectively (#). CT - e, f, g: calcifications in the spine in the ligamenta flava, anterior and posterior longitudinal ligaments and in the annulus fibrosus of the intervertebral discs (#).

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Fig. 6: Plain radiography - a and b: discreet and severe pointed acroosteolysis of the terminal phalanx of the fingers with prominent soft tissue atrophy, c: band-like osteolysis of the basal phalanx of the toe (#). d: imminent osteolysis of the distal phalanx of the great toe adjacent to a large soft tissue ulcer (#).

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**Fig. 7:** Plain radiography - a: osteolysis of bilateral styloid processes. b: hyperostosis of the distal phalanx of the great toe. c: osteoporosis of the lumbar spine with multiple compression fractures.

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**Fig. 8:** US - synovitis a: of the MCP joint and b: of the knee - synovial fluid (*) and thickened synovium (S), patella (P), flexor tendon (FL).

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Fig. 9: Plain radiography - a, b, c: arthritis of the DIP joints revealing severe joint space loss, erosions both central and marginal, periosteal spurs and "mouse ears", axial subluxation, all accompanied by numerous chunky calcifications in the periarticular soft tissues and even beyond, d: purely lytic destruction of the PIP joint with medial dislocation, without osteoplastic changes, e: joint space narrowing and a small cloud-like calcification (#) in the CMC joint of the thumb, f: joint space narrowing and a tiny erosion in the 3.rd MCP joint, calcification at the otherwise normal 5.th MCP joint.

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Fig. 10: Plain radiography - a: articular soft tissue calcifications and enthesophytes in the IP joints of the great toes and PIP joints of the 2.nd and 3.rd toes, slight joint space narrowing of the IP joint of the right great toe, b: destructive arthritis of the 2.nd through 4.th MTP joint, the latter with ankylosis. Diffuse osteoporosis. c: large well defined central erosions of the lateral tibial plateau, slight joint space narrowing, osteosclerosis of the tibial eminence and numerous enthesophytes and calcifications at insertions sites. CT - d and e: erosions and calcifications at the intervertebral (#) and costovertebral (##) joints respectively.

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Fig. 11: Plain radiography and CT- tiny and scarce periarticular calcifications to massive tumoral calcinosis in the joints of the a: hand, b: costovertebral joint, c: shoulder, d: foot, e: knee, f: wrist.

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**Fig. 12:** Plain radiography - a: dislocation of the 5.th MTP joint, b: flexion deformity "claw fingers" of the hand, thumb amputated, sporadic calcifications, c: "en boutonnière" deformity of the PIP joint of the 5.th finger.

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Conclusion

Systemic sclerosis - if fully developed - is a severe, even life threatening disease. It is therefore mandatory for the radiologist to be well informed about its features. Due to its systemic character, the changes are widespread and extensive. We, as musculoskeletal radiologists, focus on the identification and report of changes in bones, joints and soft tissues. The musculoskeletal findings, which can be assigned to SSc, are the following:

1. Thinning / thickening of the skin and subcutaneous tissue, in fingers / toes / anywhere else, may cause flection contractures and subluxations.
2. Calcifications in fingertips, periarticular and in subcutaneous tissue esp. in places exposed to pressure.
3. Myositis, fasciitis, enthesitis of ligaments and tendons (with / without calcifications) and tenosynovitis.
4. Acroosteolysis of the distal phalanx and other bony prominences, rarely band-like osteolysis of a more proximal phalanx.
5. Osteoplastic erosive arthritis (resembling psoriatic arthritis or erosive osteoarthritis, leading up to ankylosis), with calcifications in the soft tissues of the joint and in periarticular tissues.
6. Rarely purely destructive arthritis
7. Diffuse osteoporosis

Some of these findings are pathognommonic, other are subject to a wide differential diagnosis.
References

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