Soft tissue calcinosis in systemic sclerosis.

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Learning objectives

The purpose of the presentation is to:

1. Describe the distribution and patterns of soft tissue calcification associated with systemic sclerosis (SSc).
2. Emphasise important radiological findings that need conveying to the referring physicians and orthopaedic surgeons.

Background

• SSc is a systemic connective tissue disease characterised by sclerosis / fibrosis of multiple tissues and organs with specific involvement of the skin.
• SSc is mainly divided according to skin involvement into two types: 1. Diffuse SSc and 2. Limited SSc (CREST syndrome).
• It is an autoimmune condition characterised by excessive collagen production and deposition, vascular damage, and inflammation.
• Several cytokines have been implicated in the development of fibrosis. Transforming growth factor-beta 3 in particular has been suggested as having a major role in the pathogenesis of the calcinosis.
• Most commonly, calcium is deposited in injured tissues in the form of hydroxyapatite calcium crystals as dystrophic calcifications.
• The hands are involved in about 80% of cases, particularly the thumb and the radial and palmar aspects of fingers opposing the thumb with predilection for the dominant hand.

Imaging findings OR Procedure Details

• Calcinosis in SSc may take different forms. It can form punctate grouped areas of calcification or show more linear infiltrating pattern. Both patterns are shown in Fig. 1 on page 4. Other common configuration is of pseudotumours appearing as multilocular masses of calcification Fig. 2 on page 4.
• Preferential locations include the soft tissues overlying bony prominences such as the ulnar aspect of the forearm, the olecranon Fig. 3 on page 4, the acromion, and the ischial tuberosities.
• Tuft resorption is a characteristic imaging finding of systemic sclerosis. Resorption can also be observed in the distal radius and ulna, the carpal
bones, the distal end of clavicles, the superior aspect of third to sixth ribs and the periodontal membrane in the mandible.

- Plain radiograph and CT are excellent for assessment of soft tissue calcification. CT with multiplanar and 3D reconstruction is excellent to delineate the exact shape and pattern of calcification and has better anatomical assessment of soft tissues involved compared to plain radiographs. It is also useful to assess for any bony complications (like erosions). MRI is not as good as CT for exact assessment of the pattern of calcification however its strength is in its superior evaluation of soft tissues like ligaments, muscles and tendons which can be secondarily involved or injured. Ultrasound is less useful due to post acoustic shadowing associated with soft tissue calcification limiting the assessment, however it is useful for targeted or dynamic soft tissues assessment.
- Plain radiographs might be adequate for assessment, however other modalities can be helpful to assess for any complications and guide further management especially when surgical treatment is being contemplated.

- In addition to skin and subcutaneous soft tissue calcification, periarticular soft tissues are the second most common structures involved in calcification. Calcinosis may involve the articular capsule and/or ligaments Fig. 4 on page 5. Another common location is the paraspinal periarticular soft tissues, especially in the cervical spine Fig. 5 on page 6.
- Intra-articular calcinosis is less common. The most frequently affected joints are the elbow, the distal radio-ulnar and first carpometacarpal joints, the metacarpophalangeal and metatarsophalangeal joints, the knee, and the hip. Identifying intra-articular calcinosis is important as this can lead to rapid secondary joint destruction and may be treated by articular irrigation. An example of extensive intra articular calcification around the wrist joint is seen in Fig. 6 on page 7.
- Calcinosis in SSC has also a predilection for muscle fasciae or aponeuroses Fig. 1 on page 4 and may delineate the muscular bundles which can be difficult to distinguish from dermatomyositis or polymyositis.
- Complications of soft tissue calcinosis include mass effect and compression against adjacent vascular and neural structures Fig. 7 on page 8 and Joint injury/destruction in case of intra articular calcinosis as mentioned above. Symptomatic calcinosis (painful and/or ulcerating) is considered an indication for surgery Fig. 8 on page 10. In case of ulceration, a chalky discharge may be seen and the process is usually very painful. This also predisposes the area to infection and therefore its important to highlight these findings in the radiological report.
Images for this section:

Fig. 1: Right tibia and fibula radiograph showing linear fine calcifications within the calf muscles fasciae (red arrows) and grouped punctate areas of calcification within the soft tissues over the anterior tibial shaft (green arrows).

Fig. 2: Left: cervical spine plain radiograph, Middle: Axial CT and right fat suppressed T2WI showing extensive amorphous calcification involving C4 posterior elements (red arrows), with a large tumefative area of calcinosi around the right C4 and C5 transverse processes (green arrow). Note there is slight extension into the central canal.
Fig. 3: Left elbow radiograph demonstrating multilobulated areas of soft tissue calcification involving the pressure areas over the dorsal elbow and ulna (arrows).
Fig. 4: Right hand radiograph demonstrating multiple areas of dense calcification particularly within the distal soft tissues of the pulp, proximal interphalangeal joints of the thumb (red arrows) and ring finger (green arrows) involving the peri articular structures and the volar plate, and volar soft tissues within the carpus and middle finger metacarpal base. Note the Z deformity of the thumb and significant degenerative changes of the thumb carpometacarpal joint.
Fig. 5: Sagittal CT cervical spine demonstrating paraspinal periarticular soft tissues calcification with multiple well defined masses of calcification around the facet joints (arrows).
Fig. 6: Top row. Left: bilateral wrist radiograph, middle: axial CT of left wrist and right: 3D reconstruction image of left wrist. The plain radiograph demonstrates extensive soft tissue calcification around the wrist joints bilaterally more on the left side (red arrows). The left wrist CT examination with 3D reconstruction demonstrate the exact position of the calcific masses with multi focal intra articular large amorphous masses of calcinosis involving the radial aspect of the wrist joint, the carpal joints in particular the scaphotrapeziotrapezoid joint, distal radioulnar joint, carpometacarpal joints with a large area (pseudotumour) between the index and middle finger metacarpal bases (green arrow). Bottom row. MR examination (left T1WI, right fat suppressed T2WI) showing calcification with distribution as described on the CT. In addition it demonstrates intramedullary calcification within the thumb metacarpal (yellow arrow) and distal radius. There is bony destruction and disorganisation of the articular surfaces of the mid carpal articulation (orange arrows) particularly at the radial aspect of the carpus and cystic changes at the thumb metacarpal (blue arrow).
Fig. 7: Left wrist radiograph showing extensive, rounded soft tissue calcification (red arrows) along the radial border of the wrist and distal forearm. Patient presented with neurogenic symptoms secondary to compression of the superficial branch of the radial nerve. There is a linear component to the calcification (green arrows) representing calcification of the first extensor compartment tendon sheath. Further areas of calcification seen overlying the ulnar aspect of the carpus and between the distal aspects of the fourth and fifth metacarpals.

Fig. 8: Left: left knee radiograph, middle: left knee MR axial T1WI and right: sagittal PD fat suppressed image. The plain radiograph demonstrates calcification (red arrows) in the prepatellar soft tissues anterior to the extensor tendon attachments but difficult to comment exactly on tendon involvement. This was painful and MRI was requested to assess the involved structures prior to surgical removal. MR examination confirms calcification as areas of lobulated low signal anterior to the patellar surface. In addition the MR shows that the calcification does not involve the quadriceps tendon as evident by the thin fat plane (yellow arrow) separating the calcification (red arrow) from the quadriceps tendon attachment (green arrow). The patient is also at risk of ulceration of calcinosis through the skin.
Conclusion

- Certain patterns of calcification can be seen in several different tissues in systemic sclerosis.
- Awareness of imaging appearances and multimodality correlation are important in establishing the correct diagnosis and to guide local and general treatment. Early diagnosis and multidisciplinary team involvement are vital in the management of this multisystem disease.

References


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