RSDS/CRPS syndromes : an update

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1. Introduction

Complex regional pain syndrome (CRPS), historically referred to as reflex sympathetic dystrophy, Sudeck atrophy, or algodystrophy, is a complex pain disorder usually affecting an extremity after trauma, surgery, immobilization, or, less commonly, without an identifiable precipitating event.

CRPS is characterized by persistent regional pain disproportionate to the initial injury and associated with variable sensory, vasomotor, sudomotor, motor, and trophic abnormalities. The disorder most frequently involves the hand, wrist, foot, and ankle, but may also affect the knee, hip, shoulder, or other regions.

There are two schools that worked in silo over the years which have led to conflicting information in the literature. The imaging pathway studied imaging features observed on radiographs, then on CT and bone scan and finally on MRI. The clinical pathway developed clinical criteria based on symptoms and signs without taking into account imaging findings (Budapest criteria).

2. Clinical background relevant to imaging

CRPS type I and type II

Two major forms are recognized. CRPS type I occurs without an identifiable major peripheral nerve injury and corresponds broadly to the historical term *reflex sympathetic dystrophy*. CRPS type II occurs following a documented nerve injury and was previously termed *causalgia*.

Common precipitating events for CRPS include distal radius fracture, ankle or foot fracture, surgery or plaster, ligamentous or tendon injury, crush injury, stroke or neurological injury, heart or brain infarct. The severity of the original structural lesion may be surprisingly minor compared with the subsequent clinical syndrome. There is frequently a time gap between the triggering event and CRPS onset.

The Budapest criteria require continuing pain disproportionate to the inciting event together with combinations of symptoms and signs involving four domains, sensory abnormalities, vasomotor abnormalities, sudomotor/oedema abnormalities, motor/trophic abnormalities. As a consequence, CRPS predominates in the extremities and is barely recognized in the hip.

2. RSDS: Imaging findings

The imaging findings frequently encountered during the active phase of algodystrophy include regional osteoporosis on radiographs and computed tomography (CT), edema-like infiltration of the bone marrow and soft tissues on magnetic resonance imaging (MRI), and the absence of structural lesions such as erosions, chondrolysis, or periosteal reactions.

2.1. Regional Osteoporosis

In algodystrophy, increased bone radiolucency of variable severity, ranging from mild to marked, is observed (Figure 8). It appears after a delay following the onset of symptoms. This regional

osteoporosis is confined to the symptomatic joint and generally involves one of the two opposing epiphyseal ends.

2.2. Bone Marrow Edema

Bone marrow edema is an MRI finding defined by the presence of an area of moderately decreased marrow signal intensity on T1-weighted spin-echo sequences. Its margins are blurred and indistinct, with a broad transition zone between the abnormal area and the adjacent normal bone marrow. Signal intensity is generally homogeneous and moderately decreased on T1-weighted spin-echo images, in contrast to the abnormalities observed when normal bone marrow is replaced by a pathological process. The decrease in signal intensity can be confluent or patchy/reticular with occasional linear subcortical changes. The signal intensity is similar to that of normal hematopoietic marrow and higher than that of skeletal muscle. On fluid-sensitive fat-suppressed sequences, signal intensity is increased. On T2-weighted spin-echo sequences without fat suppression, signal intensity varies from normal to moderately increased.

In RSDS, edema-like marrow infiltration predominates in the subchondral epiphyseal region, usually on one side of the joint. This infiltration extends to the subchondral bone plate, with little or no intervening normal marrow between the edema-like abnormality and the articular surface. At the hip, bone marrow edema predominates in the anterosuperior region of the femoral head but may extend into the femoral neck and occasionally involve the acetabular side of the joint.

The term bone marrow edema has become established in routine practice because of its intuitive, albeit somewhat simplistic, meaning. The term edema-like bone marrow infiltration may also be used to emphasize the nonspecific nature of the MRI abnormality. Indeed, MRI-defined bone marrow edema corresponds only to a limited extent, or sometimes not at all, to actual edema on histopathological examination. In practice, this discrepancy between MRI findings and histopathological appearances explains the complete lack of specificity of edema-like marrow infiltration on MRI.

2.3. Articular and Periarticular Soft Tissues

A capsulosynovial reaction, characterized by synovial and/or capsular thickening, is frequently observed in RSDS. Soft-tissue involvement may be subtle, particularly at the hip. Periarticular soft-tissue swelling is an important clinical finding, especially in the warm form of algodystrophy affecting distal joints, such as those of the hand and foot.

Adhesive capsulitis of the shoulder, or frozen shoulder, has clinical manifestations, risk factors, and a natural history that are extremely similar, if not identical, to those observed in algodystrophy/CRPS. It is therefore currently included within the nosological spectrum of CRPS. In frozen shoulder, edema-like infiltration of the joint capsule and surrounding soft tissues is observed.

2.4. Absence of Structural Abnormalities

In RSDS, there are no structural joint abnormalities, since these conditions are associated with intense and disorganized stimulation of physiological reactive processes. The presence of any abnormality suggesting an aggressive synovial disorder (erosions or chondrolysis), a destructive or proliferative bone disorder (osteolysis or periosteal new bone formation), or cartilage damage (focal chondrolysis) should lead to exclusion of the diagnosis of algodystrophy/CRPS, unless these abnormalities were pre-existing or are related to the underlying cause of algodystrophy. It should nevertheless be emphasized that many cases of lower-limb algodystrophy exhibit microfracture-like

abnormalities. Whether these microfractures are the cause or the consequence of osteoporosis remains an open question, although both situations probably occur (49,56).

2.5. Evolution Over Time

In routine clinical practice, the diagnosis of RSDS can only be definitively established retrospectively, since progression toward almost complete recovery is an important diagnostic feature. On imaging, clinical recovery is associated with disappearance of the bone marrow and adjacent soft-tissue abnormalities. On radiographs, bone structure may return to normal. However, a so-called post-osteopenic dystrophic appearance may occasionally persist, characterized by thickening of the trabecular network in the hand and foot or relative bone sclerosis at the hip and knee.

On short-term follow-up MRI, new areas of edema-like marrow infiltration may appear while the initial abnormalities disappear. This migratory pattern may represent the most suggestive imaging feature of algodystrophy affecting the lower limb epiphyses. A wide variety of temporal patterns of edema-like marrow infiltration can be observed on MRI.

3. CRPS: Imaging findings

The diagnostic criteria for CRPS are based exclusively on the patient's history and clinical findings, without any contribution from medical imaging. A recent meta-analysis evaluating the diagnostic value of MRI in CRPS reported sensitivities ranging from 6% to 91% and specificities ranging from 50% to 100%. One might therefore conclude that MRI has no diagnostic role and that a normal MRI examination is to be expected in patients with CRPS.

In reality, the findings reported in the literature are more nuanced. Studies including patients with the warm form of CRPS consistently report a high prevalence of edema-like abnormalities involving the bones and periarticular soft tissues on MRI. Conversely, these bone marrow abnormalities appear to be absent in the cold form of CRPS. The authors of the meta-analysis nevertheless emphasize several methodological limitations: the poor quality of the available studies, the imprecise definition of MRI abnormalities, the almost exclusive inclusion of patients with hand or foot involvement, and considerable variability in the interval between symptom onset and MRI examination. Some authors have proposed reconsidering the diagnostic criteria for the warm form of CRPS in light of these MRI findings and incorporating MRI into the assessment of patients suspected of having warm CRPS of the lower limb.

In the upper limb, during the early stage, CRPS appears to be associated with edema-like infiltration of the periarticular soft tissues. Bone marrow edema-like abnormalities are subtle or even absent and are visible almost exclusively on fluid-sensitive fat-suppressed sequences or on fat-suppressed T1-weighted sequences following intravenous gadolinium administration. Adhesive capsulitis provides an excellent example. In upper-limb CRPS, bone microfractures are consistently absent. In the lower limb, during the early stage, CRPS appears to be associated with edema-like infiltration of the bone marrow and adjacent soft tissues, visible on both T1-weighted sequences and fluid-sensitive fat-suppressed sequences. Trabecular microfractures may also be observed, as discussed below. At the foot, bone loss and edema-like abnormalities are frequently dissociated, possibly because of the longer duration of the disease and its greater anatomical extent. During the chronic stage, in both the upper and lower limbs, MRI may be normal or may demonstrate isolated soft-tissue abnormalities.

Inconclusion, there is no pathognomonic radiographic, MRI, CT, ultrasound, or scintigraphic finding for RSDS/CRPS. Even the symptoms and clinical signs are not specific and can be encountered in numerous situations. This could be related with the fact that RSDS and CRPS are nondestructive disorders that are associated with reactional changes that are unusual in intensity and/or in duration.

4. Differential diagnoses

The principal differential diagnosis include disuse osteoporosis, subchondral insufficiency fracture without osteonecrosis, subchondral insufficiency fracture with osteonecrosis and osteoarthritis.

Other conditions include bone contusions and occult fractures, stress reaction and stress fracture, Osteonecrosis, Infection, Inflammatory arthritis

5. Diagnostic pitfalls

Several recurrent errors should be avoided.

Pitfall 1 — Excluding RSDS/CRPS because MRI is normal

MRI sensitivity is insufficient for this approach, particularly in chronic disease.

Pitfall 2 — Diagnosing RSDS/CRPS from bone marrow oedema alone

Bone marrow oedema is extremely nonspecific.

Pitfall 3 — Considering regional osteopenia pathognomonic

Disuse alone can produce very similar findings.

Pitfall 4 — Ignoring an occult fracture

Persistent post-traumatic pain with marrow oedema should prompt meticulous assessment for a fracture line.

Pitfall 5 — Calling extensive femoral head oedema “algodystrophy” without excluding osteonecrosis or subchondral insufficiency fracture

These diagnoses have substantially different management and prognosis.

Pitfall 6 — Expecting the same imaging pattern at every stage

CRPS is dynamic. Imaging appearances depend on disease duration, vascular phenotype, immobilization and treatment.