Algodystrophy: complex regional pain syndrome and incomplete forms

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Summary

The algodystrophy, also known as complex regional pain syndrome (CRPS), is a painful disease characterized by erythema, edema, functional impairment, sensory and vasomotor disturbance. The diagnosis of CRPS is based solely on clinical signs and symptoms, and for exclusion compared to other forms of chronic pain. There is not a specific diagnostic procedure; careful clinical evaluation and additional test should lead to an accurate diagnosis. There are similar forms of chronic pain known as bone marrow edema syndrome, in which is absent the history of trauma or triggering events and the skin dystrophic changes and vasomotor alterations. These incomplete forms are self-limited, and surgical treatment is generally not needed. It is still controversial, if these forms represent a distinct self-limiting entity or an incomplete variant of CRPS.

In painful unexplained conditions such as frozen shoulder, post-operative stiff shoulder or painful knee prosthesis, the algodystrophy, especially in its incomplete forms, could represent the cause.

KEY WORDS: algodystrophy; bone marrow edema; complex regional pain syndrome; CRPS.

Introduction

Algodystrophy is a painful disease characterized by erythema, edema, functional impairment, sensory and vasomotor disturbance. The pathogenic mechanisms are not fully understood and some clinical aspects are still lacking of a whole pathogenetic comprehension, but significant progress in knowledge have been recently achieved (1). The internationally recognized term for this syndrome nowadays is Complex Regional Pain Syndrome (CRPS), divided in type I or II, regarding respectively the absence or presence of a neurological lesion (2).

At the same time, there are other pathological entities, such as bone marrow edema syndrome, transient osteoporosis of the hip and regional migratory osteoporosis, that show common features with CRPS. However, they do not have a clear pathogenetic position and their definitive classification remains uncertain.

History and terminology

In the 17th and 18th century literature, several references can be found and many Authors tried to describe this syndrome historically difficult to contextualize. In 1864, it was presented in detail for the first time by Mitchell et al. (3). They described symptoms as burning pain, swelling, skin color and temperature changes, the intense sensitivity to touch and joint stiffness following peripheral traumatic nerve injury due to distal extremity gunshot wounds in soldiers injured in the American civil war. They coined the term ‘causalgia’ to describe this pain syndrome, which means burning pain, used to describe a particular painful condition often associated to various sensory disturbances that sometimes followed major nerve injury. Early in the 1900 Sudeck described a pain syndrome with similar symptoms and many of the features of causalgia described by Mitchell developed in an extremity after bone fractures without peripheral nerve injury (4, 5).

In 1916 Leriche linked the sympathetic nervous system to causalgia, reporting pain relief after surgical sympathectomy (6). Evans in 1946 introduced the term reflex sympathetic dystrophy for this syndrome (7), today one of the most widely accepted term. Since that time, several researchers contributed to describe this pathological entity and many names have been proposed. In 1993, the International Association for the Study of Pain (IASP), after an evaluation of etiologic and pathophysiological aspects as well as clinical characteristics, developed the nomenclature, complex regional pain syndrome (CRPS), and proposed the IASP diagnostic criteria (8). The IASP also subdivided the CRPS into subtypes I and II, respectively without or with the presence of clinical signs of major peripheral nerve injury. However, there is no unanimity on the nosological definition; considering only the Italian scientific production, there are 13 different names for such disorders, 79 can be found in the English literature, and more than 100 in other languages. The new term “complex regional pain syndrome”, is currently the most commonly used (9).

CRPS

As previously shown, complex regional pain syndrome is known by various names, such as reflex neurovascular dys-
trophic phase are reversible, whereas the atrophic form, and even mild cognitive deficits (10). The acute and dys-

limb (15). In a subset of patients, CRPS becomes chronic to other body parts and even on the opposite or ipsilateral

sensory signs (hypoesthesia, hypoalgesia, and hypother-
mesthesia) can develop (14). Subsequently, the limb often

growth changes, muscle weakness. The affected area is

When established, is irreversible.

Because of the enormous clinical variability and the etiologi-
cal heterogeneity, the diagnosis is not easy. There are many non-standardized diagnostic criteria systems. New diagno-
tic criteria were codified by the International Association for the Study of Pain (IASP) task force on taxonomy at a con-
sensus workshop in 1994 (8). Subsequent validation re-
search found problems with lack of specificity and potential over-diagnosis using these criteria. Successively in the fall of
2003 in Budapest the diagnostic criteria were reviewed (16) (Table 1). These more specific diagnostic criteria were
adopted in 2012 as the new international standard for the di-
agnosis of CRPS by the IASP and these criteria have been
shown to reduce CRPS diagnostic rates by about 50% (10, 13).

The diagnostic criteria were designed to provide a standard-
ized, common methodology for making decisions and discern
if unidentified pain conditions represent CRPS or not. The application of inappropriate treatments due to misdiagnosis
can contribute to excessive medical costs, or worse, may de-
lay the appropriate treatment (17).

The distinction in CRPS-1 and CRPS-2 is made for the pres-
ence or absence of nerve lesion. A third diagnostic subtype
called CRPS-NOS was recommended and it would include
those patients who did not fully meet the new clinical criteria,
but whose signs and symptoms could not better be ex-
plained by another diagnosis. Those patients have fewer than
three symptoms or two sign categories, or do not show a
sign at the time of the examination but had exhibited this previously, and whose signs and symptoms were felt to be
best explained by CRPS (18).

The diagnosis of CRPS is based solely on clinical signs and
symptoms, and for exclusion compared to other forms of
chronic pain. The Budapest criteria excluded the use of
imaging exams to perform the diagnosis. There are not speci-

cific diagnostic procedures and their rule is to add informa-
tion to support and confirm the clinical diagnosis. Generally,
the conventional plain radiography is the first exam per-
formed and shows bone demineralization, but it is positive
only in chronic stages.

An early diagnosis and an interdisciplinary approach are fund-
damental factors for an optimal and successful treatment.
Incomplete forms

For the first time, in 1959, Curtiss and Kincaid (19) described a syndrome of transient demineralization of the hip in pregnant women. Since then, various terms were proposed in order to describe the common benign clinical and imaging conditions of bone marrow edema, such as regional migratory osteoporosis (RMO) and transient osteoporosis of the hip (TOH) (Figure 2). Subsequently, these clinical entities were included under the general term of “bone marrow edema syndrome” (BMES) (20). Now BMES represents a distinct entity with specific clinical and imaging features, but some Authors support that this syndrome should be a variant of algodystrophy or an abortive form of osteonecrosis (21).

Many pathogenetic hypotheses have been proposed but the etiology of these “incomplete forms” remains unknown. Many cases are misdiagnosed due to the lack of specific symptoms and the rarity of the disease. The main characteristic in common with CPRS is the chronic pain. Instead, characteristics missing in CPRS are: absent history of trauma or triggering events, the extremely rare involvement of the upper limbs, the absence of skin dystrophic changes and vasomotor alterations, the recurring and migrant nature and the complete restitution ad integrum (absent in cases of algodystrophy non promptly recognized and treated) (22).

Laboratory findings usually are not useful to the diagnosis. Standard radiographs could be normal in early phases. It can reveal bone demineralization at 3-6 weeks from the onset of the symptoms and this alteration may persist after the symptom resolution, also up to 2 years (23). In the absence of clinical signs (autonomic and dystrophic changes) and radiographic examination findings, it becomes important for a correct assessment, in contrast to CRPS, to perform further investigations such as RMI and scintigraphy. The three-phases bone scan may be positive after a few days from the onset of symptoms. Tc99m-MDP increased uptake representing focal increase in capillary permeability, hyperemia, and osteoblastic activity. MR imaging scans reveal low-signal intensity on T1-w images and high signal intensity on STIR or fat-suppressed T2-w images. These changes reflect the increased intra and extracellular fluid of the bone marrow resulting from new bone formation and repair processes (23). The differential diagnosis at MRI scans should be made with infection, osteonecrosis and post-traumatic marrow edema.

It is important to identify CPRS from other incomplete forms for the different evolution of the two conditions. In fact, these last are self-limited, and aggressive or surgical treatment is not needed, on the contrary CPRS, if not treated, may determine severe disability.

Figure 2 - Typical case of transient osteoporosis. A) Pelvis MRI with BME in the right hip. B) After 12 months complete remission of the BME. C) After 16 months new BME in the contralateral hip.
Conclusion

The CPRS represents a difficult disease to diagnose and treat. The diagnosis is made by excluding other diseases with chronic pain and the recent Budapest clinical criteria represent the most accepted diagnostic approach. It is characterized by slow-healing and easily chronicization that requires a multidisciplinary approach. The etiopathogenesis is not still completely known because of many factors contribute to create wide range of clinical forms.

In our opinion, the incomplete forms, characterized by pain and bone marrow edema, are a subtype of algodystrophy syndrome. They do not have clinical signs (such as skin alterations or vasomotor sign) and a trauma history. They are self-limited and the prognosis remains favorable. In the absence of autonomic and trophic involvement of the limb and with none standard radiographic alterations, it is necessary a further diagnostic study with bone scans or MRI. The magnetic resonance represents the gold standard for its easy accessibility and low invasiveness. The incomplete forms, generally, are observed and treated by orthopedic specialist.

Our treatment consists of the combination of: bisphosphonates, calcium supplements, magnetotherapy, hyperbaric oxygen therapy and partial weight bearing over the affected limb.

The algodystrophyc syndrome in one of its variants could justify also other clinical painful entities as frozen shoulder or post-operative stiff shoulder, in which an intense pain syndrome appears not justified. As well as in cases of painful "unexplained" knee prosthesis, in which intrinsic factors (instability, malalignment, loosening, component wear, metal hypersensitivity, infection) or extrinsic factors (adjacent region problems, vascular or neurologic disease) are excluded.

References