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Reflex sympathetic dystrophy syndrome (Complex Regional Pain Syndrome type 1) in a Nigerian woman – a case report

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Abstract
Reflex Sympathetic Dystrophy Syndrome also referred to as Complex regional pain syndrome type 1 (CRPS 1) is an uncommon regional musculoskeletal pain disorder which in spite of being rare is a well and long recognized rheumatologic condition. It is characterized by variable dysfunction of the musculoskeletal, skin, neurological, and vascular systems**.** This case presentation is to heighten our awareness about the existence of this rare disorder in our environment. The little attention received by this disorder is presumably because of paucity of knowledge about the disorder and also because it mimics various other musculoskeletal and rheumatologic conditions. Early diagnosis is key, as this will help the patient to receive prompt and adequate treatment, thus preventing complications. Multi-disciplinary approach is necessary for the management of the case.

Keywords: *Complex regional, mussculoskeletal pain, rheumatologic, condition*

Résumé
Le syndrome de dystrophie sympathique réflexe, également appelé syndrome de douleur régionale complexe de type 1 (SDRC 1), est un trouble de la douleur musculo-squelettique régionale rare qui, en dépit d'être rare, est une maladie rhumatologique bien connue depuis longtemps. Il se caractérise par un dysfonctionnement variable des systèmes musculosquelettique, cutané, neurologique et vasculaire**.** Cette présentation de cas vise à accroître notre prise de conscience de l'existence de cette maladie rare dans notre environnement. Le peu d'attention reçue par ce trouble est probablement dû au manque de connaissances sur le trouble et aussi parce qu'il imite diverses autres affections musculosquelettiques et rhumatologiques. Un diagnostic précoce est essentiel, car cela aidera le patient à recevoir un traitement rapide et adéquat, évitant ainsi les complications. Une approche multidisciplinaire est nécessaire pour la gestion du cas.

Mots clés: *Syndrome de douleur régionale, rhumatologique, dysfonction, musculosquelettique, prise en charge.*

Introduction

Reflex sympathetic dystrophy (RSD) is an uncommon regional musculoskeletal disorder usually of unknown aetiology. Also referred to as Complex Regional Pain Syndrome (CRPS) type 1 in which no nerve injury is implicated according to a classification arrived at during a special consensus workshop held in Orlando, Florida in 1993 [1]. Type 2 CRPS is termed causalgia, in which a nerve injury is an important component. The term causalgia was coined by Mitchell in 1864 and derives from the Greek word for burning pain.CRPS type 1 has also been variously termed shoulder - hand syndrome, Sudeck's atrophy and algodystrophy. Other names include Transient osteoporosis, Regional migratory osteoporosis and Post-trauma painful osteoporosis [2]. It primarily relates to abnormal functioning of the sensory, sympathetic, andmotor nerves and involves an exaggeration of normal physiological responses with changes at multiple levels in the central and peripheral nervous systems [3].

The clinical picture comes in various forms, but the main components are those of regional pain and tenderness which are disproportionate to any inciting event and commonly accompanied by vasomotor symptoms and signs which include swelling and colour changes, sudomotor (sweating), or motor abnormality such as stiffness, weakness, tremor, and dystonia [4]. There are anumber of clinical presentations of CRPS, with milder forms being fairly common and having a good prognosis, but with the more severe and less common CRPS often responding poorly to treatment and being more persistent [4]. Pain, emotional distress, and disability characterize this disorder. Although some diagnostic criteria have been proposed, they are still in the process of validation. These are complicated by the fact that not all features may be present at the same time and may vary in intensity.The condition tends to affect upper limbs more commonly than lower limbs**.**

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Usually one limb is affected, but it may go on to affect another limb. It is usually most evident distally affecting the handand wrist, or foot and ankle, but a whole limb can also be affected,such as in the case of "shoulder - hand syndrome" [5]. Some of the other clinical disorders that may be confused with this disease include; vasculitis and vascular insufficiencies, lymphedema, deep vein thrombosis and Raynaud's phenomenon [6].

Case history

A 58-year-old trader presented at the rheumatology out-patient clinic at the OlabisiOnabanjo University Teaching Hospital, Sagamu with recurrent history of burning pain and swelling of the right fore-arm of 3 years duration. She was involved in an accident in which she fell into a ditch and landed on the right arm, sustaining mild cuts and bruises. She had pain and mild local swelling which was managed appropriately then. About six months later, she developed pain which she described as peppery with burning sensation. The pain started insidiously after being free of the initial symptoms following treatment at a private hospital. There was associated tingling and burning sensation alternating with numbness over a visible scar that resulted from the injury. She also complained of sleeping difficulty and she got tired easily. She also sometimes lost interest in her usual daily activities and events happening around her worsened by feelings of frustration. She was ten years post-menopausal at presentation. Physical examination revealed an anxious looking, but otherwise healthy middle-aged woman. There was exaggerated response to mild touch with a light paper and to pain using a sterile office pin. There was associated guarding and vague tenderness over the right fore-arm. The affected right fore-arm initially appeared slightly bigger than the left fore-arm. The swelling became reduced and insignificant over the following three visits to the hospital. Patient was unusually frequent at the hospital, not waiting for her appointment dates because of the perceived severe pain and strange sensation she experiences over the affected arm. There was no active swelling seen at any joint, and musculoskeletal examinations were essentially normal. An assessment of chronic regional pain syndrome type 1 (reflex sympathetic dystrophy syndrome) was made to keep in view regional migratory osteoporosis.The Fasting blood sugar (FBS), Fasting Lipid Profile (FLP), and Full blood count (FBC) were all normal and Erythrocyte sedimentation rate (ESR) was 18mm/hour (Westergren). Plain radiograph of the right arm showed widespread osteopenia. Bone densitometry could not be done because of non-availability of facility.Patient was placed on a course of low-dose tricyclic antidepressant – amitriptyline; neuropathic and neuromodulatory agents - Pregabalin and

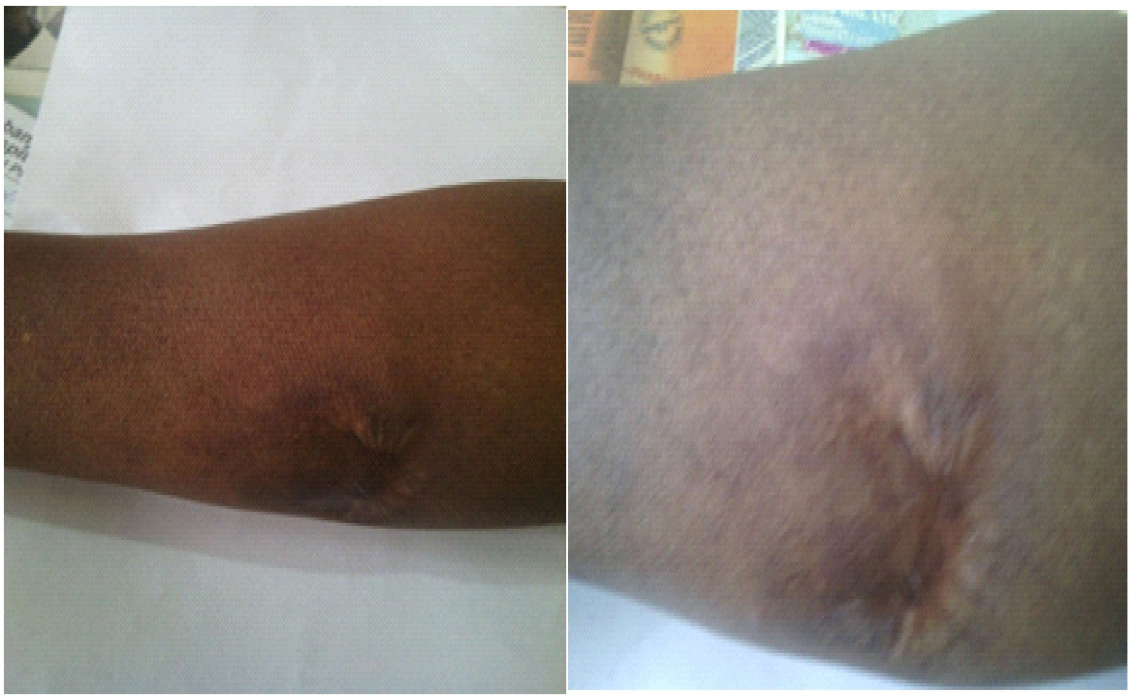

Fig. 1: *Showing the scar on the right arm of the patient*

Neurovite-forte, which were helpful. There was however recurrence of symptoms once those drugs were exhausted. She was reviewed by the Neuro-psychiatrist but was just re-assured and given an anxiolytic agent, clonazepam for a few weeks. She was also placed on oral bisphosphonate (Alendronate), Calcium and Vitamin D supplements. She was evaluated and commenced on physiotherapy. It is the authors' opinion that she might have benefitted from regional sympathetic nerve root or ganglion blocks, in view of the recurrent and disturbing symptoms. However, the anaesthetist suggested that all available medical options should first be explored. The patient was eventually lost to follow up.

affected limb. The vasomotor dysfunction (temperature and skin colour changes) and sudomotor abnormalities (swelling and sweating changes) were expectedly in an asymmetry fashion.

There was no evidence of nerve injury, joint pain or swelling, decreased range of motion in any joint or significant muscle weakness in the patient which could have suggested inflammatory arthritis or other autoimmune diseases. The normal ESR was also not in support of inflammatory arthritis. The widespread osteopenia seen on plain x-ray in our patient is as expected in RSD and this differentiates it from other common rheumatologic disorders that may present with joint or limb swellings which usually

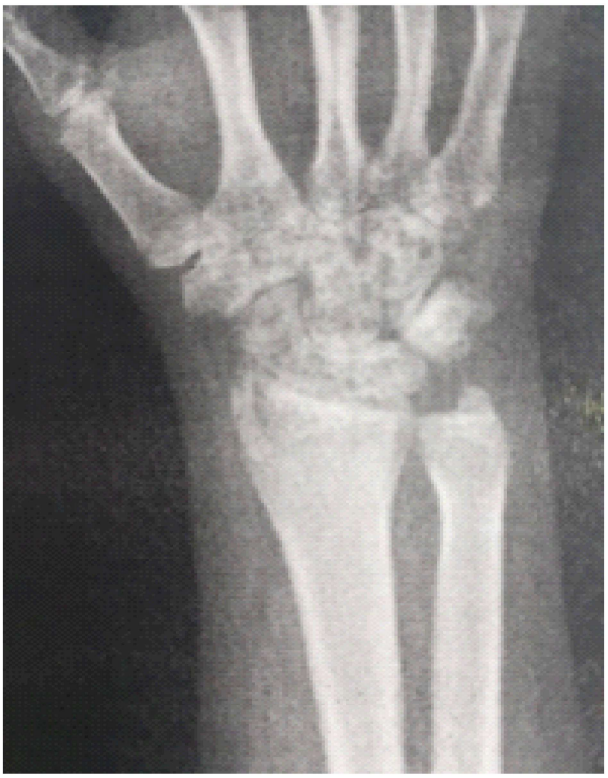

Fig. 2: *Plain x-ray of the right fore-arm showing widespread osteopenia*

Discussion

Type 1 Complex regional pain syndrome (CPRS 1) formerly known as Reflex Sympathetic Dystrophy (RSD) is a clinical syndrome of variable course and the diagnosis is clinical but requires a high index of suspicion.

In our patient, the Budapest clinical diagnostic criteria for CRPS was adopted [7,8]. The patient met all 4 criteria for diagnosis which include an initial noxious event which is injury in this case, continuous pain, which is disproportionate to the inciting event. Swelling and colour changes with excessive sweating were also observed on the

have in addition to osteopenia, findings such as bony erosions, joint space narrowing, deformities and osteophytes, which are characteristics of osteoarthritis, rheumatoid arthritis and other inflammatory arthritis. Other useful investigations include bone densitometry to assess the bone mineral density, bone scintigraphy and Magnetic Resonance Imaging (MRI) which are more of prognostic values than diagnostic. HLA (Human Leucocyte Antigen) typing has also been said to be useful in categorizing patients with RSD as it serves as an important susceptibility factor apart from the increased

sympathetic tendencies found in many other patients with RSD.

An algorithm of treatment has been proposed by Stanton - Hicks (2002) [9] which include medical, rehabilitation and psychological components. This algorithm has revealed the importance of multidisciplinary approach in the management of RSD. The management of patients with RSD is an individualized one.

Several drugs, either alone or in combination with sympathetic blockade, have been found to be effective in prolonging the duration of symptomatic relief. Some of these drugs reduce the activity of the sympathetic nervous system, whereas others are primarily anti-inflammatory. Two major approaches to the treatment of CRPS 1 however are sympathetic blockade and anti-inflammatory therapy. Our patient had been placed on various anti-inflammatory drugs from the referral centre. Sympathetic blockade has been found helpful in patients with RSD, however the patient did not have the benefit of use of sympathetic blockade as part of her management in spite of the prolonged, persistent and debilitative nature of her illness, because of loss to medical follow-up. Surgical sympathectomy is the last resort in patients with refractory RSD [10].

The patient presented with symptoms of depression and cognitive impairment which are not uncommon in patients with RSD. We instituted low dose tri-cyclic anti-depressant (amitriptyline) and low-dose anticonvulsants in other to alleviate the symptoms. Various anticonvulsants used in this patient include carbamazepine, gabapentin and pregabalin, all at low doses. Oral bisphosphonates and Calcitonin have also been found to play a role in the management of RSD because of the associated features of osteopenia or osteoporosis. Alendronate at a dose of 70mg per week was given to this patient. Other drugs that have been found to be useful in the treatment of RDS include Alpha blockers (prazosin), beta blockers (propranolol), and calcium channel blockers (nifedipine) [11].

It can therefore be concluded that early diagnosis and prompt treatment by the specialists will go a long way in preventing the attendant complications of this rare disease.

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