An Interesting Case of "Migratory" Complex Regional Pain Syndrome (CRPS)!

Dear Editor,

We had a 32-year-old man who came to us with complaints of pain, swelling, redness, and increased warmth affecting his ankles and wrists. He gave peculiar complaints of "shifting or migratory arthritis." He had an affection of right ankle, which lasted for 2 months with minimal response to symptomatic

treatment. However, not completely, most of the symptoms improved spontaneously over one month. This was followed by a similar affection of the left wrist and right wrist and presented to us with excruciating pain in the left ankle (8 months of illness). He had no constitutional symptoms, denied trauma in the past, and had no other comorbidities. His physical examination showed an erythematous and edematous left ankle and dry skin with decreased hair. He had painful restriction of the movements. There was no significant motor deficit or sensory loss. The rest of the general examination was normal. A working diagnosis of "migratory polyarthritis with autonomic features" was considered.

His routine laboratories were normal, and there was no evidence of inflammation (Erythrocyte Sedimentation rate (ESR) and C reactive Protein (CRP)). Other workup for inflammatory polyarthritis such as rheumatoid arthritis (RA) factor, antinuclear antibody (ANA) panel, uric acid, and anti-streptolysin antibody (ASO) titers were all unremarkable. Cardiac evaluation, nerve conduction studies, and Doppler studies of the lower limb were normal too. Since the patient had vasomotor symptoms, sensory symptoms, sudomotor symptoms, and trophic changes, the possibility of complex regional pain syndrome (CRPS) type 1 was considered. Since CRPS is a diagnosis of exclusion, all the other investigations were done to identify the underlying etiology. A triple-phase bone scan (TPBS) was done, which showed features of CRPS in the form of blood pooling and delayed metabolism in the affected limbs [Figure 1].

The patient was started on symptomatic therapy with gabapentin and low-dose steroids (though the evidence is not very convincing). He was referred to a psychiatrist to address the psychosocial aspects associated with the pain. The patient showed a decent improvement in his symptoms.

CRPS is a neuropathic pain disorder defined by the "presence of distinct clinical features, including allodynia, hyperalgesia, sudomotor and vasomotor abnormalities, and trophic changes," usually preceded by an inciting event.[1] The pain experienced is disproportionate to the degree of injury, and it outlasts the expected time for tissue healing.[1] The exact pathophysiology has not been described, and it is believed to be a multifactorial illness.[1] Peripheral and central sensitization, inflammation, altered sympathetic and catecholaminergic function, altered somatosensory representation in the brain, genetic factors, and psychophysiological interactions play a role in the pathophysiology of CRPS.[2] We speculate that altered somatosensory representation and altered expression of pain

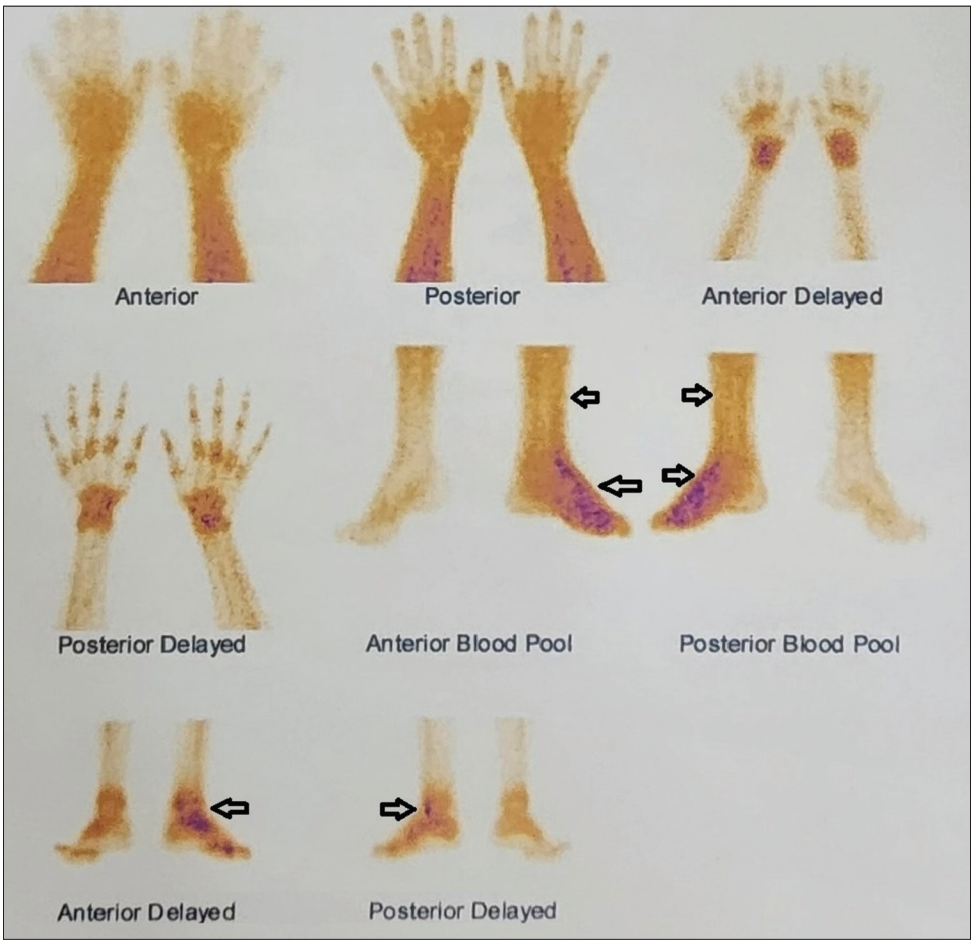

Figure 1: Black arrows showing the blood pooling and delayed metabolism in the affected limbs involving the dorsum of the left foot, the left ankle, and the lower part of the left leg consistent with CRPS

Table 1: Budapest Criteria[4]

A	They should report continuing pain disproportionate to the inciting event		
B	They should report at least one symptom in three of the four following categories:		
	1.	Sensory	Hyperalgesia and/or allodynia
	2.	Vasomotor	Temperature asymmetry and/or skin color changes and/or skin color asymmetry
	3.	Sudomotor/ edema	Edema and/or sweating changes and/or sweating asymmetry
	4.	Motor/ trophic	Decreased range of motion and/or motor dysfunction (weakness, tremor, and dystonia) and/or trophic changes (hair, skin, and nails)
C	They must display at least one sign at the time of evaluation in two or more of the following categories:		
	1.	Sensory	Evidence of hyperalgesia (to pinprick) and/ or allodynia (to light touch or deep somatic pressure)
	2.	Vasomotor	Evidence of temperature asymmetry and/or skin color changes and/or asymmetry
	3.	Sudomotor/ edema	Edema and/or sweating changes and/or sweating asymmetry
	4.	Motor/ trophic	Evidence of decreased range of motion and/ or motor dysfunction (weakness, tremor, and dystonia) and/or trophic changes (hair, skin, and nails)
D	There is no other diagnosis that better explains the signs and symptoms.		

receptors and altered "pain network" due to brain plasticity could be responsible for the migratory physiology of CRPS.[2] There are two subtypes of CRPS: type I, reflex sympathetic dystrophy (no nerve trauma), and type II, formerly known as causalgia (occurs in the setting of known nerve trauma). It follows a regional rather than dermatomal or nerve distribution.[1]

Patients with CRPS can have a varied presentation ranging from allodynia, hyperalgesia to endocrinopathies, and depression.[3] Budapest criteria [Table 1] has been used widely to recognize this entity.[4] CRPS can spread to involve more proximal portions of the affected limb; however, migratory CRPS is a rare entity, which has been seldom described in the literature.[3] A case report by Akkus S *et al*. described a similar clinical picture of migratory and recurrent CRPS 1 treated with sympathetic blocks and gabapentin.[3] Tony HC *et al*. described such similar presentation in pediatric patients.[5]

Various objective testing measures have been utilized to aid in the diagnosis of CRPS, such as thermography, TPBS, and the quantitative sudomotor axon reflex test.[4] Though the sensitivity of TPBS is 70–98% and specificity is 36–96%, when used in addition to the clinical criteria, it may serve as a vital aid in narrowing down the diagnosis.[6] On a TPBS, CRPS 1 often shows increased blood flow, blood pooling, and delayed metabolism in the affected limb.[6] Reliance only on a triple-phase bone scan for the diagnosis of CRPS is controversial.[6] The differential diagnosis includes small- or large-fiber sensorimotor neuropathy, cellulitis, erythromelalgia, vasculitis, vascular insufficiency, lymphedema, deep vein thrombosis, and Reynaud's phenomenon.[4]

Treatment of this illness is multimodal ranging from symptomatic treatment with anti-inflammatory medications, anticonvulsants, antidepressants, transdermal lidocaine, opioids, N methyl D aspartate (NMDA) antagonists, and bisphosphonates.[7] Elevated levels of catecholamines associated with depression can worsen CRPS by inducing central sensitization through adrenergic mechanisms; thus, behavioral therapy has an important additional role in CRPS.[8] Interventions such as sympathetic block, spinal cord stimulation, and dorsal ganglia stimulation have also been tried with varied results.[8,9]

Our patient in this study satisfied the Budapest clinical criteria for CRPS type 1, which was supported by the findings on the TPBS. This case throws light on the unusual presentation of a rather unusual clinical entity. Thus, migratory CRPS type 1 is a clinical condition, which requires a high index of suspicion and extensive evaluation. Early recognition, symptomatic therapy, and immunotherapy may have an important role if recognized early.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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