A case of complex regional pain syndrome post herpes zoster; An underreported condition

Dear Editor,

CRPS (complex regional pain syndrome) was first reported in 1901 by Sudeck.[1] It is a chronic condition typically affecting the extremities (hand and foot) and is characterised by persistent regional pain, disproportionate to the initial event. Complex regional pain syndrome is distinct from other chronic pain conditions due to the presence of signs indicating prominent autonomic and inflammatory changes in the region of pain, and is diagnosed with the help of Budapest criteria.[2]

Herpes zoster (HZ) infection is characterised by vesiculobullous lesions in a dermatomal distribution, caused by the reactivation of the varicella zoster virus (VZV). The reactivation of latent varicella zoster virus in the dorsal root ganglia results in herpes zoster (shingles), a localised cutaneous eruption that is accompanied by neuropathic pain.

A 20-year-old male presented with pain along the right upper limb. The patient had a history of herpes zoster involving the C5 dermatome one month ago [Figure 1]. He presented to us with pain along the right upper limb. Considering post-herpetic neuralgia as a diagnosis, he was started on gabapentin, tramadol and non-steroidal anti-inflammatory drug (NSAID) therapy but 1 month later, he presented with an increase in pain and multiple discrete shiny vesicles on the right hand associated with excessive sweating [Figures 2 and 3]. There was no history of trauma, seizures or muscle weakness.

On physical examination, there was a deep burning pain (Numerical Rating Scale: 7/10), allodynia (tested by stroking cotton swab) and skin temperature changes with significant sweating leading to miliaria crystallina in the right hand and forearm, measured with a Vaporimeter (Delfin technologies Ltd.). The reading was 14 g/m^2 H in the left hand while it was 24 g/m^2 H in the right hand at the thenar eminence. The motor examination revealed no abnormalities in bilateral upper and lower limbs.

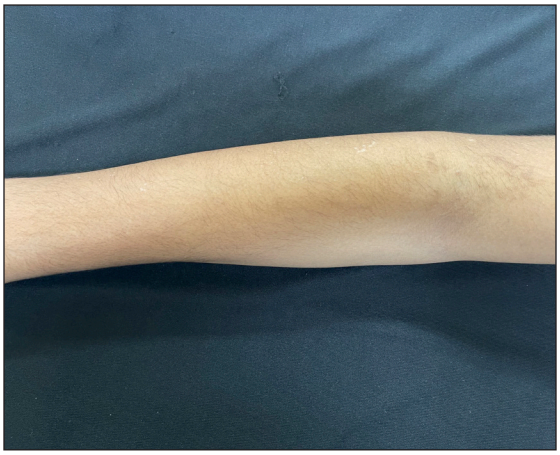

Figure 1: Scars of healed herpes zoster lesions over the right forearm involving the C5 dermatome

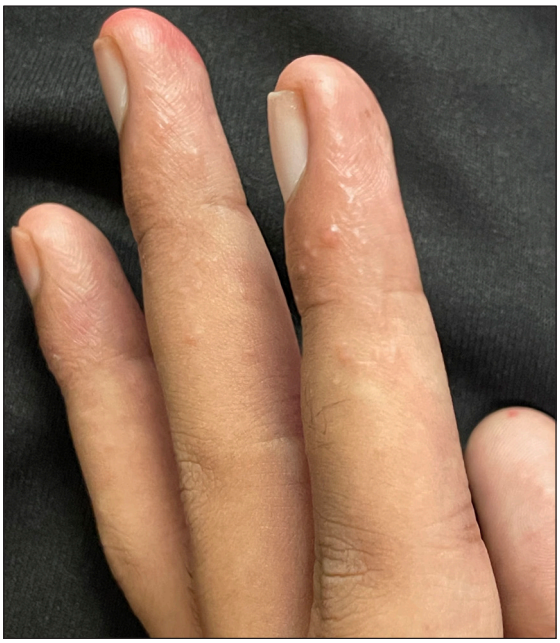

Figure 2: Multiple discrete shiny vesicles on the fingers of the right hand associated with excessive sweating

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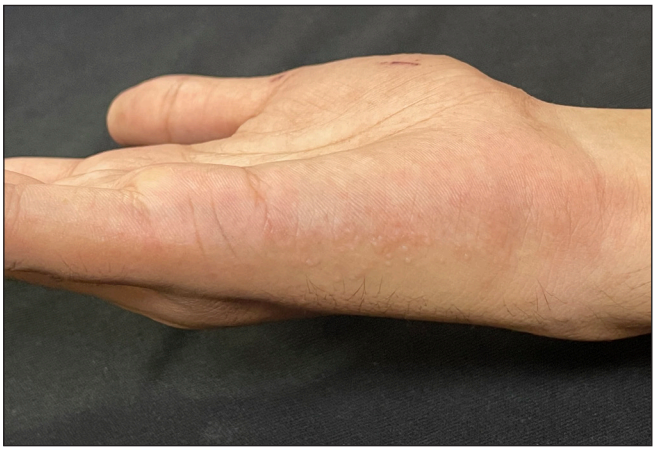

Figure 3: Multiple discrete shiny vesicles over the inner aspect of the right hand associated with excessive sweating

All haematological and biochemical investigations, including c-reactive protein and serum uric acid, were normal ruling out any inflammatory cause. A venous doppler and radiographs of both hands were found to be normal. Nerve conduction studies were normal.

The neurological examination of the patient included the presence or absence of complex regional pain syndrome-like symptoms as defined by the 2010 Budapest criteria along four categories: sensory, vasomotor, sudomotor/oedema and motor/trophic.[2]

The Budapest criteria were met [Table 1][2] and the diagnosis of complex regional pain syndrome post herpes zoster of the right upper limb was made in consultation with the neurologist. He was counselled about the condition and started on tab carbamazepine along with tab pregabalin 75 mg twice daily. He improved gradually over the next 1 month.

Complex regional pain syndrome of the hand with a limited extension does exist and it forms an important differential diagnosis, particularly in the case of localised and persistent pain post herpes infection. Approximate prevalence of localised complex regional pain syndrome affecting the hand is around 12%.

Several hypotheses can explain the mechanism of herpes zoster in causing complex regional pain syndrome. First mechanism can be attributed to the intense pain due to herpes zoster and this initial afferent nociceptive stimulus can sensitise multiple sympathetic neurons, resulting in sympathetic outflow. The second mechanism is secondary inflammation caused by herpes zoster infection induced cytopathic changes. The third mechanism is the development of abnormal synapses between efferent sympathetic nerves and afferent sensory nerves post herpes zoster infection.[3]

The significant accumulation of inflammation-related signs in our patient with only of 1 month duration, indicates an obvious inflammatory component during the early phase of

Table 1: Budapest clinical diagnostic criteria for complex regional pain syndrome (CRPS)

1. Continuing pain disproportionate to any inciting event
2. Must report at least one symptom in three of the four following categories:
   - Sensory: reports of hyperesthesia and/or allodynia.
   - Vasomotor: reports of temperature asymmetry and/or skin colour changes and/or skin colour asymmetry.
   - Sudomotor/oedema: reports of oedema and/or sweating changes and/or sweating asymmetry.
   - Motor/trophic: reports of decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (hair, nail, skin)
3. Must display at least one sign at the time of evaluation in two or more of the following categories:
   - Sensory: evidence of hyperalgesia (to pinprick) and/or allodynia (to light touch and/or deep somatic pressure and/or joint movement).
   - Vasomotor: evidence of temperature asymmetry and/or skin colour changes and/or asymmetry.
   - Sudomotor/oedema: evidence of oedema and/or sweating changes and/or sweating asymmetry.
   - Motor/trophic: evidence of decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (hair, nail, skin)
4. There is no other diagnosis that better explains the signs and symptoms

Table 2: Differences between Complex Regional Pain Syndrome (CRPS) and Post-Herpetic Neuralgia

	Complex regional pain syndrome	Post-herpetic neuralgia
Clinical	• Sensory symptoms like increased perception of stimuli • Vasomotor symptoms like changes in temperature or sweating in the affected limb in comparison to the normal limb • Sudomotor/oedema and motor/trophic (demonstration of weakness, decreased range of motion or trophic changes in hair, nails or skin) Complex regional pain syndrome causes include neuromas, ganglia and nerve entrapments	Persistent pain within a dermatome and affects the region which is innervated by the dermatome PHN causes include varicella zoster virus infection
Diagnostic	History Physical examination Budapest criteria	History Clinical examination
Management	**Pharmacologic therapy:** Corticosteroids, antidepressants, opioids, ketamine **Interventional treatment**: Sympathetic nerve block, spinal cord stimulation **Other treatments** include vocational, psychological and occupational therapy	**Pharmacologic therapy:** Acetaminophen, non-steroidal anti-inflammatory agents and opioids **Interventional treatment**: Sympathetic blocks, intrathecal injections or surgical interventions and zoster vaccine
Prognosis	Good	Good

the complex regional pain syndrome, which is consistent with the finding reported by Ott *et al*.[4]

Complex regional pain syndrome is to be suspected in patients of herpes zoster when there is no relief or an increase in symptoms, in spite of early pharmacological intervention and also when there is pain with trophic, sensory and motor changes [Table 2]. We report this case to draw attention to the existence of complex regional pain syndrome after herpes zoster infection, which is rarely reported in literature, as generally cases with similar presentations are misdiagnosed and managed as post-herpetic neuralgia.

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Preema Sinha, Juhi Sharma, Parul Kamboj, Prashantha GB

Department of Dermatology, Base Hospital, Lucknow, India.

Corresponding author:
Dr. Preema Sinha,
Department of Dermatology, Base Hospital, Lucknow, India.
sinhapreema@gmail.com

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Exploring the unusual: A case of vulvar crohn's disease with concomitant leukocytoclastic vasculitis

Dear Editor,

Crohn's disease is a chronic inflammatory disorder that affects the gastrointestinal tract and is often associated with extra-intestinal manifestations including cutaneous ones, which are estimated to be present in 18–44% patients.[1] These cutaneous manifestations could be due to contiguous spread from the gastrointestinal tract (sinuses and fistulae), or non-contiguous spread (metastatic) or reactive spread (erythema nodosum, pyoderma gangrenosum, vasculitis). Of these, the reactive manifestations are most common. Here, we report an interesting combination of vulvar Crohn's disease (a metastatic manifestation) with leukocytoclastic-vasculitis (a reactive manifestation) without overt evidence of gastrointestinal tract involvement.

A 24-year-old, unmarried woman presented for progressive asymptomatic swelling of the labia for the past four years and recurrent rashes over lower extremities for the past two years. The swelling initially started as a small (0.5 cm) papulo-nodule on left labia majora. This increased in size and new nodules developed to involve both labia majora and adjoining areas over the next six months. Two years later, she started developing recurrent palpable purpura on the lower extremities [Figure 1]. Though the genital lesions were not biopsied,

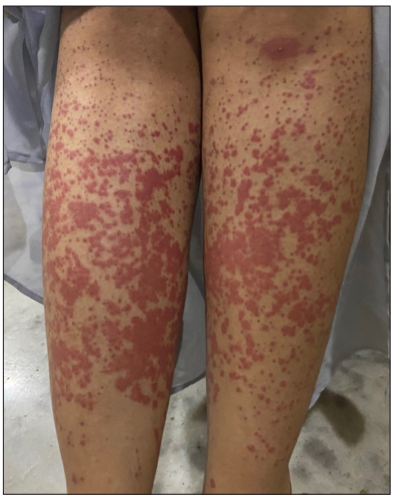

Figure 1: Palpable purpura over the lower extremities.

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