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Journal

Dermatology Online Journal, 21(9)

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Publication Date

2015

DOI

10.5070/D3219028700

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Peer reviewed

Photo vignette

Immediate resolution of severe bullous chronic regional pain syndrome with onset of spinal paralysis

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Dermatology Online Journal 21 (9): 14

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Abstract

Complex regional pain syndrome (CRPS) is an incompletely understood disorder characterized by progressive regional pain and sensory changes, with fluctuating cutaneous edema and erythema. We describe a patient with a rarely reported severe bullous CRPS variant on the left lower extremity, which resolved immediately upon developing spinal paralysis.

Keywords: cRPS, Chronic Regional Pain Syndrome, RSD, Reflex Sympathetic Dystrophy, Causalgia

Case synopsis

A 25-year-old female with a history of a left above-knee-amputation (AKA), presented with two weeks of progressive pain, erythema, edema and bullae of her left thigh and stump. The pain worsened with light touch and heat. She also reported local tingling and acute temperature fluctuations. The patient denied any changes in her wound care regimen, new medications, or exposures to potential irritants. She was otherwise well. She denied any family history of neurologic conditions.

Four years prior, the patient had suffered a crush injury to the left leg, resulting in delayed onset of severe local pain, edema and erythema, diagnosed at the time as CRPS. Despite aggressive therapy including sympathetic blocks and various neuro-active medications, she underwent a palliative AKA two years later due to persistent severe pain and progressive immobility.

She did well until 20 months post-AKA, when she presented to our dermatology clinic with the sudden new left limb stump pain and erythema described in the opening paragraph. Her symptoms and morphology were identical to the prior CRPS presentation, except with tense bullae now overlying the erythema.

Physical examination revealed an edematous stump, with large bullae overlying well-demarcated pink, purple, and brown patches (Figure 1) that were markedly hyperesthetic to light touch. The distribution did not correspond to her prosthesis or areas of friction, and discontinuing the prosthesis had no impact. The local erythema and temperature on palpation fluctuated rapidly during some visits, suggesting acutely alternating vasodilation and vasoconstriction.



Figure 1. Chronic Regional Pain Syndrome. Tense bullae overlying erythematous patches on the left stump

Laboratory work-up included a normal CBC, chemistry panel, CRP, and ESR. Radiographs showed a possible erosive process in the femur remnant.

Biopsies from peri-bullous skin demonstrated epidermal necrosis with perivascular and interstitial neutrophils, lymphocytes, and eosinophils (Figure 2). These findings are non-specific and are not highly suggestive of any one diagnosis, but were noted to be consistent with traumatic or ischemic injury. In one of the two biopsies, foci of mild small vessel vasculitis were noted, and reported as likely a result of ischemia, rather than a primary vasculitis. Direct immunofluorescence exhibited no specific immunoreactivity. Tissue cultures and stains showed no bacterial, mycobacterial, or fungal organisms.

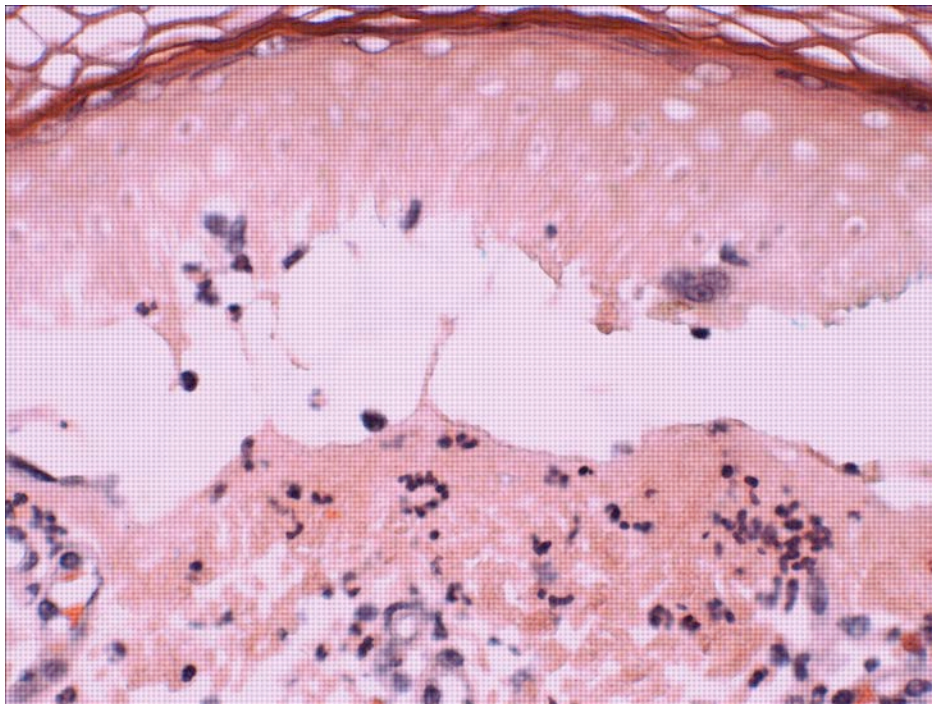


Figure 2. Chronic Regional Pain Syndrome. The biopsy shows epidermal pallor consistent with necrosis. A sparse mixed and non-specific inflammatory infiltrate composed of neutrophils, lymphocytes, and eosinophils was seen in the dermis (400x)

Given her pain, dysesthesia, temperature sensitivity, and fluctuating erythema, the patient was diagnosed with recurrent CRPS. Her condition improved modestly with gabapentin, opioids, and sympathetic blocks. Prednisone, pentoxifylline, and dapsone helped minimally. Local botulinum A toxin injections appeared to halt progression of one exacerbation.

Her symptoms finally remitted with an unfortunate development: she developed a spinal abscess around an intrathecal pain pump one week after placement, resulting in total lower extremity motor and sensory loss. Immediately with the onset of paraplegia, all of the patient's left stump cutaneous pain, erythema, vasomotor instability, and bullae fully resolved; at the time of this writing, 19 months after the spinal abscess, she remains paraplegic and completely free of all subjective and objective findings of CRPS. (Figure 3).

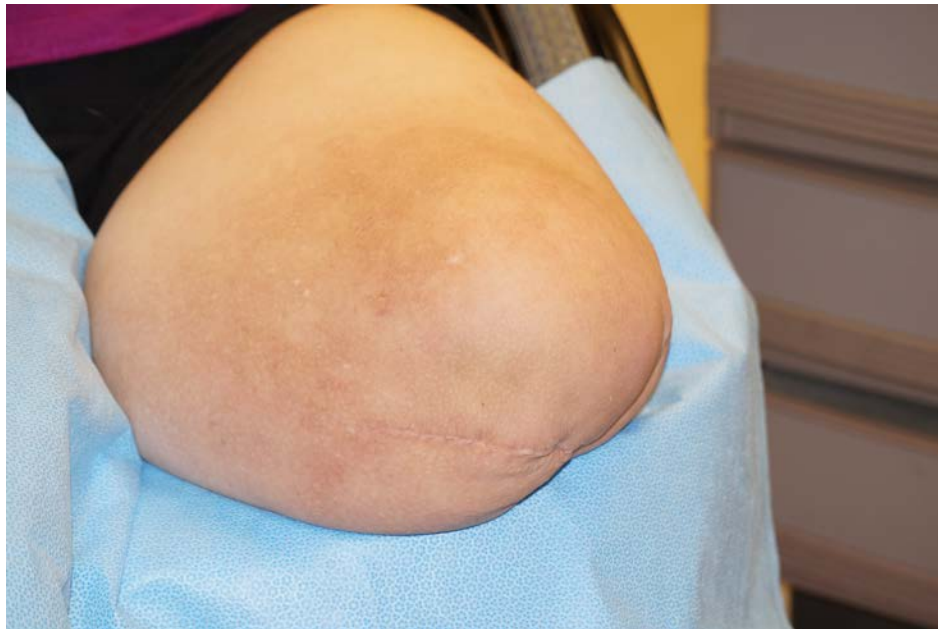


Figure 3. Normal appearing stump 19-months post-denervation showing mild residual post-inflammatory hyper-pigmentation

Discussion

CRPS, formerly termed reflex sympathetic dystrophy or causalgia, is an incompletely understood disorder characterized by progressive regional pain and dysesthesia, with a variety of potentially severe cutaneous changes.

The cause of CRPS is unknown, but the pathogenesis is thought to be related to post-traumatic changes to the sympathetic nervous system [1], which may explain why many CRPS symptoms are related to vasomotor instability. Manifestations usually begin at the site of an inciting injury and then spread [2, 3]. Limbs are the most common sites affected, and CRPS has been reported to recur after amputation of the affected limb, as in our patient [4].

Diagnosis is made clinically, although autonomic testing, bone scintigraphy, and patient response to treatment may help eliminate other etiologies. Autonomic testing may show altered sympathetic nervous system function such as an increased resting sweat output. Bone scintigraphy may show increased uptake. Patchy de-mineralization can be seen on radiographs. Abrupt relief following a sympathetic block is also suggestive of a diagnosis of CRPS. Histologic findings are non-specific, but ischemic changes as seen in our biopsies have been described [5], and are mechanistically consistent with the severe episodic vasoconstriction of CRPS.

Dermatologic manifestations of CRPS include fluctuating erythematous patches and plaques, edema, and in later stages, brawny thickened or atrophic skin [6]. Cutaneous allodynia and temperature sensitivity is common. Tense bullae, as seen in this patient, have been described very rarely, and attributed to severe edema and ischemia from transient and intermittent neurogenic vasoconstriction [1, 7].

The absence of remarkable serologies, cultures, imaging and vascular studies in our patient supported the diagnosis of CRPS by ruling out conditions with overlapping features, including cellulitis, perniosis, systemic or cutaneous lupus

erythematous, panniculitis, leukocytoclastic vasculitis, immunobullous disease, and edema bullae. Allergic contact dermatitis merits special consideration because of the geographic nature of her patches, and the eosinophils on histology. However, the biopsies showed no spongiosis or epidermal eosinophils, arguing strongly against allergic contact dermatitis, or any eczematous process. Furthermore, the lesions did not correspond to any known contactants including her prosthesis, and they did not resolve even months after removing the prosthesis. Contact dermatitis also could not explain the bulk of the symptoms associated with her local erythema. Finally, cutaneous lesions of CRPS have been previously reported to emerge in strikingly geometric patterns, as seen in this patient [2, 3].

Management of CRPS is challenging, and treatment modalities range from pharmacologic approaches (antidepressants, anticonvulsants, glucocorticoids, NSAIDs, bisphosphonates, IVIG) to invasive therapies such as sympathetic blocks.

Our patient's sudden paraplegia, while catastrophic, was also remarkable in that all bullae and other cutaneous manifestations of her CRPS resolved with spinal denervation, corroborating the neurogenic pathogenesis of her condition.

CRPS is a challenging condition to diagnosis and treat. Early treatment is critical, as it may improve prognosis. The prominent cutaneous manifestations of CRPS, including bullae in severe cases, can facilitate timely diagnosis in the context of a patient with localized sensory disturbances and vasomotor instability.

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