An Unusual Cutaneous Presentation of Reflex Sympathetic Dystrophy

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REFLEX sympathetic dystrophy (RSD) is a disabling, poorly understood syndrome with a wide variety of clinical presentations.1-9 It classically presents after trauma, even a trivial injury, and manifests as autonomic dysfunction in the injured distal extremity.3-5,7,10 RSD is diagnosed primarily by clinical symptoms and signs, including burning pain, allodynia, distal extremity edema, color changes (erythema, pallor, cyanosis, or mottling), vascular instability, temperature changes, hyperhidrosis, and cutaneous dystrophic or atrophic changes.1-7,9,10 Cutaneous changes of RSD may occur in any stage of the syndrome and mainly consist of skin atrophy, vascular instability, hyperhidrosis, or allogdynia.1,2,9

Several recent reports have documented previously unreported skin lesions as manifestations of RSD.1,2 These have included small numbers of patients with long-standing RSD who presented with recurrent ulcerating papules, bullous eruptions, inflammatory lesions, and reticulate hyperpigmentation.1,2 This suggests that skin disease in RSD is more diverse than previously appreciated.

We treated a young woman with a recurrent RSD of her left upper extremity, whose primary presentation consisted of burning pain of the left upper extremity associated with a large area of erythema, edema, hyperpigmentation, and bullous lesions on the proximal lateral aspect of her arm. Her presentation was unusual not only in the type of skin lesions but also in their location and the rapidity with which they resolved after sympathectomy.

Case Report

A 33-year-old, 80-kg woman presented to the pain clinic with a 5-yr history of left shoulder and upper, lateral, left arm pain. Her illness began when she sustained a rotator cuff tear and underwent a rotator cuff repair at another institution. One month after surgery, she injured her left hand, requiring placement of pins in the thumb and 6th finger. Postoperatively, she had persistent burning pain in the entire left upper extremity. Erythematous, bullous, upper, lateral left arm lesions developed. Biopsy of these lesions revealed a mild, diffuse vasculitis, with negative cultures. Triple-phase bone scan was consistent with an RSD.

Initial treatment of this presumed RSD consisted of oral analgesics and physical therapy. After 6 months with little improvement, several stellate ganglion blocks were administered, which temporarily relieved the pain. A continuous brachial plexus block was administered for 7 days, which produced complete and permanent pain relief, along with resolution of the upper, lateral, arm skin lesions.

Several years later, the patient underwent a second left rotator cuff repair under general anesthesia. Within 1 week of surgery, she experienced burning pain predominantly in the left upper, lateral arm. The skin lesions reappeared in the same distribution as previously noted. A series of 14 stellate ganglion blocks, administered every 2-4 days, was performed. Only 12-18 h of pain relief was obtained from each block, with no prolongation of relief after repeated blocks. The skin lesions were unchanged after the stellate ganglion blocks. Adjuvant medications, including propranolol, amlodipine, baclofen, amitriptyline, and various opioids were prescribed without significant improvement.

She was referred to the dermatology clinic at Indiana University, who also thought that she had an unusual presentation of RSD. They noted macules, ulcerating papules, hyperpigmentation, and bullous eruptions on the upper lateral left arm (fig. 1). Biopsy of these lesions documented a superficial and deep perivascular mononuclear infiltrate. Immunofluorescent studies, cultures, and antinuclear antibodies were negative (on several occasions). Because she previously responded to a continuous brachial plexus block, she was referred to our clinic for continuous sympathetic blockade.

The patient described a burning, tingling sensation that accompanied the large macular, papular ulcerations, hyperpigmentation, and bullous lesions evident on her proximal, lateral arm. Marked allodynia and hyperesthesia were noted in this area. Pitting edema and erythema were evident in this circumscribed area, which was...
Discussion

It has been accepted that the signs and symptoms of RSD must be present in an area at the periphery of an extremity.\textsuperscript{1,11} This report is in direct contrast to that observation. Our experience is similar to a few recent reports that have documented RSD localized to more proximal portions of an extremity.\textsuperscript{1,2,11} Veldman et al. described examples of “algodystrophy” restricted to localized burning pain and skin changes about the hip or knee.\textsuperscript{11} Webster reported two cases of presumed RSD skin lesions with pain confined to the trunk.\textsuperscript{1}

This case also presents unusual skin eruptions in RSD. Cutaneous changes are prominent in RSD and have been reported to consist primarily of edema, vascular instability, temperature changes, color changes, altered nail and hair growth, and late atrophic changes.\textsuperscript{1–5,6,8,9,11,12}

Our patient presented with ulcerating papules, macules, bullous lesions, and reticulate hyperpigmentation. Webster reported 16 cases of RSD with similar lesions.\textsuperscript{1,2} Only two of these cases exhibited bullous lesions, both on the leg. Our patient is unique to have bullous lesions on the arm as a manifestation of RSD.

The mechanism producing these skin eruptions remains obscure. Recent evidence suggests that patients with RSD have increased peripheral vascular α-adrenergic hypersensitivity.\textsuperscript{11} These lesions could represent areas of focal ischemia that respond to sympathetic blockade. The rapid resolution of these skin changes after sympathectomy suggests a readily reversible process.

RSD is a diagnosis of exclusion and remains a clinical diagnosis. Multiple attempts were made in our patient (and in Webster’s patients) to find another etiology for the skin changes. In all cases, viral, bacterial, and fungal cultures were negative. Collagen vascular diseases were excluded, as were other vascular diseases.

The International Association for the Study of Pain has defined four criteria that must be satisfied to diagnose RSD. These are (1) an initiating event or a cause of immobilization, (2) allodynia or hyperesthesia, (3) evidence of altered blood flow or edema, and (4) exclusion of all other conditions that would explain the symptoms.\textsuperscript{7,12} Criteria 2–4 must be satisfied. Our patient met all of these.

Concomitantly, however, we believe that we must be careful in extending the boundaries of the definition of RSD. This was discussed in the Sixth World Congress of Pain, in which the consensus was that the term RSD should be used in the descriptive sense and does not
imply a specific underlying mechanism. Others agree with this concept. Several years ago, Roberts and Campbell introduced the concepts of sympathetically maintained pain and sympathetically independent pain to define, respectively, those pains that are dependent or independent of the sympathetic nervous system. These pain states do not necessarily describe subsets of RSD. Our patient’s unique presentation was sympathetically maintained, as evidenced by the rapid resolution of her pain and skin lesions after sympathectomy. Even the bullous lesions resolved, which did not occur after sympathectomy in Webster’s two cases.

It should be noted that our patient had a recurrence of pain and skin lesions after a general anesthetic for a second rotator cuff repair. This is similar to two patients reported by Rocco, who had recurrence of sympathetically maintained pain after surgery under general anesthesia. One of Rocco’s patients did not have a recurrence of sympathetically maintained pain after epidural anesthesia. Therefore, we must be aware that the choice of anesthetic may influence the reappearance of sympathetically maintained pain in patients with a history of that type of pain.

In summary, we present a case of ulcerating papules, macules, bullous eruptions, and reticulate hyperpigmentation on the upper lateral arm consistent with the internationally accepted criteria of RSD. Our patient’s presentation included bullous eruptions on the upper extremity as a manifestation of RSD and is only the third reported case in the literature of bullous eruptions in RSD. These rare skin changes were sympathetically mediated and resolved with sympathectomy. From this experience and other reports, the skin lesions in RSD appear to be more diverse and dramatic than previously appreciated.

References

17. Rocco AG: Sympathetically maintained pain may be rekindled by surgery under general anesthesia (abstract). ANESTHESIOLOGY 79: A865, 1993