Case Report

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A case of truncal complex regional pain syndrome: literature review

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Introduction and importance: Complex regional pain syndrome is a rare and chronic pain disorder characterized by an abnormal level of pain disproportionate to the initiating cause, often manifesting well after the triggering event.

Case presentation: The authors present a free past medical history 33-year-old female, employed as a nurse in an intensive care unit, presented with a 9-year history of diffuse back pain. Her symptoms initially emerged at the neck with muscle spasms and restricted neck mobility, eventually progressing along the spine. Notably, the pain became episodic and intensified over time, significantly impeding her daily activities. Analgesic (non-steroidal anti-inflammatory drugs) treatments proved ineffective, and a distinct feature emerged—a change in skin colour to dark purple spots on her back, accompanied by hyperhidrosis and extreme tenderness. Despite an exhaustive evaluation involving bloodwork, inflammatory markers, serological tests, and radiographic imaging, a definitive diagnosis remained elusive until she responded positively to Pregabilin.

Clinical discussion: Although typically associated with extremities, this case challenges the conventional understanding of complex regional pain syndrome by showcasing its manifestation in the truncal region. The patient's clinical history, examination findings, and diagnostic journey are detailed herein, shedding light on the complexity and diagnostic considerations associated with this condition.

Conclusion: The case underscores the importance of a comprehensive approach and prompts a reevaluation of the existing guidelines to encompass such atypical presentations.

Keywords: autonomic dysfunction, chronic pain, complex regional pain syndrome (CRPS), motor abnormalities, pain management, sensory abnormalities, skin discoloration

Introduction

Complex regional pain syndrome (CRPS) is a rare and chronic pain disorder characterized by an abnormal level of pain disproportionate to the initiating cause, often manifesting well after the triggering event[1]. This syndrome is frequently accompanied by autonomic and motor symptoms. CRPS can be categorized into two distinct subtypes: CRPS type 1, which lacks a clear nerve injury, and CRPS type 2, where a known nerve injury (or its branch) is present. A notable hallmark is the presence of autonomic dysfunction and persistent localized inflammation that defies a dermatomal pattern. The clinical presentation of CRPS typically involves hyperalgesia, allodynia, skin alterations, temperature fluctuations, and swelling. Remarkably, females with upper limb injuries seem to be more susceptible to this condition[2,3].

HIGHLIGHTS

- The case challenges conventional notions by demonstrating complex regional pain syndrome (CRPS) in the trunk, deviating from the typical extremity-focused manifestation.
- Despite exhaustive tests, diagnosis remained elusive until the patient responded positively to Pregabalin, underscoring the intricate diagnostic challenges posed by atypical CRPS cases.
- The case prompts a reevaluation of existing guidelines, advocating for broader consideration of atypical CRPS presentations and highlighting the need for comprehensive approaches to diagnosis and treatment.

Case presentation

History

A free past medical history 33-year-old female, employed as a nurse in an intensive care unit, presented with a nine-year history of diffuse back pain without history of trauma. She did not complain of fever, chills, or other neurological symptoms. Her symptoms initially emerged at the neck with muscle spasms and restricted neck mobility, eventually progressing along the spine. Notably, the pain became episodic and intensified over time, significantly impeding her daily activities. Analgesic treatments proved ineffective, and a distinct feature emerged—a change in

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skin colour to dark purple spots on her back, accompanied by hyperhidrosis and extreme tenderness. The patient did not report family history of similar disease Figure 1.

Clinical examination

Physical examination revealed no tender points upon palpation of the back, accompanied by pronounced hypersensitivity (increased sensitivity to stimuli, such as touch or temperature), dysgeusia (distortion in the sense of taste, leading to abnormal or unpleasant taste perceptions), allodynia (the perception of pain in response to non-painful stimuli), and hyperalgesia (exaggerated response to painful stimuli). Despite extensive evaluations, which included MRI and computed tomography (CT) scans, bloodwork, and assessments for autoimmune conditions [such as antinuclear antibodies (ANA), extractable nuclear antigens (ENA), rheumatoid factor (RF), and anti-cyclic citrullinated peptide antibodies (ACPA)—all of which returned negative results], as well as screenings for endocrine disorders, the underlying cause of the symptoms remained elusive. In addition to these evaluations, psychological and neurological assessments were performed, revealing a comorbid generalized anxiety disorder.

Diagnosis and outcome

Guided by the diagnostic criteria established by the international association of the study (IASP) of pain criteria the patient was ultimately diagnosed with truncal CRPS. This case serves as a unique illustration of CRPS manifesting in a non-conventional anatomical region, prompting a reevaluation of prevailing assumptions about the condition. After ruling out, neurological, autoimmune diseases, this patient was started on a trial of NSAIDs, Amitryptalin and responded very well to Pregabalin.

Discussion and review

There are few reported case reports of CRPS[4–8]. The origins of CRPS can be traced back to historical records, with the first documented description dating back to the seventeenth century. The renowned French surgeon, Ambroise Paré, reported a peculiar condition affecting King Charles IX, where the monarch experienced persistent pain and contractures in his arm following bloodletting treatment. This early observation laid the foundation for the understanding of CRPS[9]. The incidence and prevalence of CRPS are not precisely known worldwide. However, some studies have provided estimates of its occurrence. The reported incidence rate varies between 5.46 and 26.2 cases per 100 000 individuals per year[10]. Regarding prevalence, retrospective studies have indicated a range from 0.03 to 37% in individuals who have experienced trauma[11].

The severity of symptoms does not necessarily correlate with the severity of the original injury. Psychological factors like stress can worsen symptomatology[12]. CRPS is associated with various diseases and conditions, such as stroke, mastectomy, pregnancy, and the use of certain drugs. Besides trauma and diabetes, other predisposing factors for CRPS include immobilization, which may lead to increased sensitivity to stimuli and changes at the spinal level[10]. Psychogenic or hysterical factors, especially associated with depressive symptoms, may also contribute to CRPS through interactions with catecholamine release, although this remains a hypothesis[13]. COVID-19 was discriminated in causation of CRPS[14]. Antibiotics like cephalosporin was reported to improve symptoms of CRPS[15].

The exact underlying cause of the condition remains uncertain, and various pathogenetic theories have been proposed, including autonomic dysfunction, neurogenic inflammation, and alterations in central nervous system neuroplasticity[16]. However, these concepts are still debated and no definitive conclusion has been reached. Presently, it is believed that the origin of this issue is likely to be multifactorial based on existing evidence. The clinical manifestations of CRPS can manifest anywhere from hours to

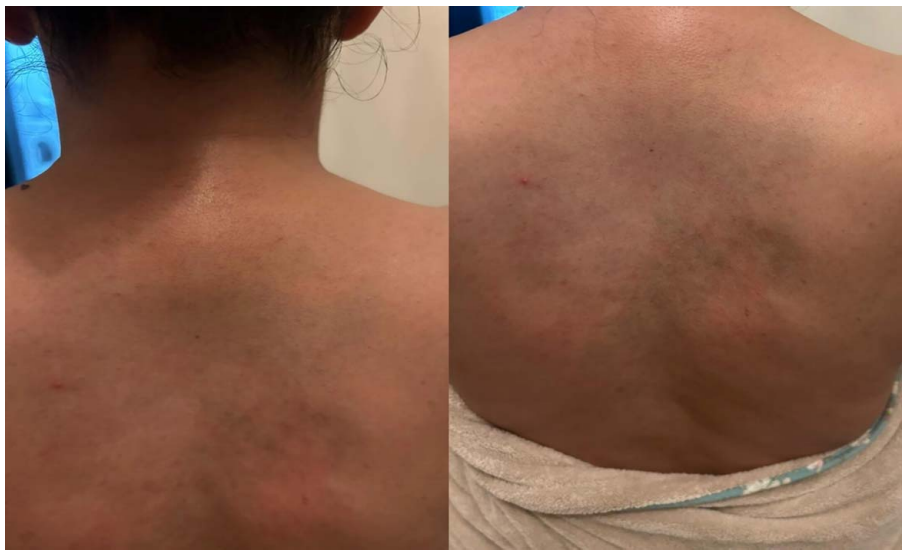

Figure 1. The patient presented with skin discoloration on her back, appearing as dark spots with a purple colour. The affected skin was very painful to touch, and she also experienced excessive sweating. Additionally, she had difficulty moving her spine in all directions due to the pain and other associated symptoms.

months after the initial noxious event. Typically, they present as a triad of autonomic, sensory, and motor abnormalities[10]. CRPS lacks pathognomonic signs or symptoms, and there is no definitive diagnostic test available. Diagnosis is primarily reliant on a comprehensive medical history, which includes assessing the severity and duration of symptoms, the type and severity of the initial injury, and a thorough physical examination of the affected limb[17].

Early intervention is crucial for achieving full recovery and preventing further damage in CRPS. Addressing CRPS requires a comprehensive, multidisciplinary pain management approach that also focuses on restoring the functionality of the affected limb. The treatment regimen often includes non-steroidal anti-inflammatory drugs (NSAIDs), corticosteroids, cyclooxygenase (COX) 2 inhibitors, and antioxidants. These medications aim to alleviate pain and address the inflammatory processes associated with CRPS. However, it's important to note that the inflammation in CRPS may primarily stem from neurogenic sources (triggered by inflammatory mediators from nerve endings), and as of now, there is no specific drug that has been extensively studied for targeting this particular type of inflammation[18]. Both minor opioids (e.g. tramadol) and major opioids (such as morphine) play a crucial role in managing moderate to severe pain that proves challenging to control, and they have shown effectiveness in treating neuropathic pain. In addition to opioids, adjuvant medications like antiepileptics (e.g. gabapentin) and tricyclic antidepressants (such as amitriptyline) and pregabalin have been utilized in the treatment of CRPS. These adjunct therapies complement the main treatment approach and contribute to the comprehensive management of the condition[11]. Sympathetic nerve block is a viable treatment option for patients who do not respond well to pharmacological treatments, particularly when administered early in the disease's progression. It has shown to improve short-term pain relief and joint mobility, and its effectiveness is more pronounced when conducted in the initial stages of the condition. Though there is limited data on long-term efficacy, some controlled studies have not demonstrated consistent results. Nevertheless, nerve blockage can offer a pain-free period that enhances limb mobility, enabling intensive physiotherapy, especially when utilizing continuous techniques like local anaesthetic infusion via an auxiliary or lumbar epidural catheter[19]. Early rehabilitation plays a crucial role in preventing muscle atrophy and contractures, which, in severe cases, may become irreversible. However, patient participation in rehabilitation can be challenging due to the presence of severe pain and associated psychological disorders. Physical therapy is an important component of rehabilitation and can effectively reduce pain and improve limb mobility, but the intensity of treatment varies based on the severity of the syndrome.

Conclusion

This case challenges the conventional understanding of CRPS by showcasing an atypical presentation in the truncal region. The diagnostic journey underscores the complexity and diagnostic considerations inherent to such cases. As our understanding of CRPS continues to evolve, incorporating diverse clinical presentations into existing guidelines is essential to ensure comprehensive and accurate management strategies for patients.

Ethical approval

Ethical approval for this study was not required as it is a case report.

Consent

Written informed consent was obtained from the patient for publication and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Author contribution

All authors contributed equally in writing and reviewing the paper.

Conflicts of interest disclosure

There are no conflicts of interest.

Guarantor

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Data availability statement

Data is available upon request by emailing the corresponding author.

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