



Case report

Truncal complex regional pain syndrome, myth or reality: Case report

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ABSTRACT

Introduction: Complex regional pain syndrome (CRPS) is an under diagnosed, chronic pain condition commonly described occurring in the extremities. Its occurrence in the trunk is rarely reported and is thought by some to be non-existent.

Case presentation: We report an unusual case of truncal CRPS post appendectomy which presented with debilitating pain and review relevant literature.

Discussion and conclusion: We recommend that subsequent descriptions and diagnostic criteria should include the trunk as a site of occurrence of CRPS and not just the extremities. This will help reduce under diagnosis of this important condition.

1. Introduction

Complex regional pain syndrome is a rare chronic pain syndrome characterised by pain disproportionate to the triggering factor and may present much later than the trigger [1]. It is usually accompanied by autonomic and motor symptoms. CRPS is divided into two distinct sub types by the absence (CRPS type 1) or presence (CRPS type 2) of a known injury to a major nerve (or branch thereof). It is characterised by the presence of autonomic dysfunction, persistent regional inflammatory changes without following any dermatomal distribution. CRPS usually presents with hyperalgesia, allodynia, skin changes, temperature alterations and oedema. Females with upper extremity injuries are more predisposed [2,3].

True incidence of CRPS is unknown but data suggests that it may be under reported [4]. It typically affects extremities [5]. In fact its occurrence on the trunk is so rare that most authors refer to CRPS as originating only from the extremities [1,6,7]. And others openly doubt its occurrence in trunk and face despite case reports [8]. This can be misleading and lead to under diagnosis. Occurrence of CRPS following appendicitis is even rarer [9]. Here we report an unusual case of truncal CRPS following appendectomy we managed at our academic institute. The report is in accordance to the SCARE guidelines 2020 [10]. We

review relevant literature to show why it can also occur in the trunk.

2. Case history

A 26-year-old female of African descent, previously well, presented to outpatients' department with hyperalgesia of a McBurney's point appendectomy scar 3 weeks after a seemingly uncomplicated appendectomy. The initial symptoms that had led to a diagnosis of appendicitis had completely resolved, what remained was disproportionate wound pain despite there being no signs of wound infection or inflammation. During periods of partial pain relief, the pain was localised to a trigger point on an area where a tiny lump could be felt underneath the skin along the healed appendectomy scar (Fig. 1). The point was hypersensitive with dysgeusia, allodynia and hyperalgesia. Merely touching it would trigger nausea, sometimes vomiting, and severe pain.

The pain would not respond to oral standard pain killers, including opiates. However, the pain responded to trigger point injections with methylprednisolone and lignocaine, giving a relief lasting 2–3 weeks. This led to an entertainment of the diagnosis of a neuroma. Given the need for frequent injections, a surgical exploration of the wound for a neuroma was done by a senior surgeon. This did not yield any results. The pain gradually evolved. The severity of 100/100 on the visual

Abbreviations: CRPS, complex regional pain syndrome; MRI, magnetic resonance imaging; CT, computer tomography; IASP, International Association of the Study of Pain.

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Fig. 1. McBurney's point appendectomy scar.



Fig. 3. Brittle nails with brownish discoloration.

analogues score ensued. It was associated with allodynia, hypersensitivities, sweating, skin colour changes on the affected side of the body (anterior abdominal wall going down the right thigh) (Fig. 2), nail colour changes to a brownish discoloration were also noted on all limbs (Fig. 3). The nail brownish discoloration would worsen with the worsening pain. The abdominal wall on the affected side was warmer to touch compared to the normal side.

Extensive evaluations including abdominopelvic magnetic resonance imaging (MRI) scans, computed tomography (CT) scans and extensive bloodwork including a full endocrine work-up, screen for auto-immune conditions and full blood count did not yield any possible explanation to the pain or the other symptoms. The evaluation included psychological, psychiatric and neurological assessments. A diagnosis of complex regional pain syndrome (CRPS) was made according to the international association of the study (IASP) of pain criteria. The pain had become debilitating and she became bedridden and would not be able to go about her daily routine. This was despite analgesia, physical therapy and psychotherapy. She was then commenced on pregabalin 75 mg thrice daily and gradually titrated to 150 mg three times a day to get adequate relief. The pain subsided to about a 20/100 on the visual analogue scale and she became functional, albeit with limits due to pain. A year down the line she continues to have episodes of worsening pain on and off, which improves on above treatment.

3. Discussion

CRPS remains a very difficult condition to treat despite medical



Fig. 2. Skin discoloration.

advancement [11,12]. The diagnosis of CRPS is purely on clinical grounds as there are no specific diagnostic tests to confirm it [13]. Our patient demonstrated all the signs and symptoms in all categories of the Orlando diagnostic criteria for CRPS, the Budapest diagnostic criteria of IASP for CRPS and the Japanese CRPS 1 diagnostic criteria for clinical purposes [3,7,8]. Orlando diagnostic criteria comprises of the following: 1. The presence of an initiating noxious event, or a cause of immobilization (not absolutely necessary for diagnosis. Absent in 5–10%) 2. Continuing pain allodynia, or hyperalgesia in which the pain is disproportionate to any known inciting event. 3. Evidence of sometime of oedema, changes in skin blood flow, or abnormal sudomotor activity in the region of the pain 4. This diagnosis is excluded by the existence of other conditions that would otherwise account for the degree of pain or dysfunction.

The Budapest diagnostic criteria were designed to be an improvement to the Orlando one, by incorporating objective findings. It comprises of the following: pain processing (allodynia, hyperalgesia), vasomotor dysfunction (skin colour and/or temperature changes, oedema or sudomotor dysfunction and motor/trophic signs and symptoms [14]. The Japanese CRPS 1 diagnostic criteria is almost identical to the above mentioned [3]. For the diagnosis one does not need to have every symptom in all the categories but our patient had, hence the diagnosis was without doubt regardless of it being truncal.

Even though development of truncal CRPS is extremely rare it is not impossible as there are a few documented cases [9,15]. Our case is a testimony to that. There has been a documented case of a patient who developed CRPS, weeks after laparoscopic surgery for appendicitis [9]. This is similar to our patient. However, our patient developed CRPS after an open appendectomy. Chronic abdominal wall pain is not uncommon following surgery and can occur in up to 30%, and may sometimes evolve into CRPS [9,15] as in our patient.

It is noted that up to 80% of CRPS patients may be severely disabled. The difficulty in finding a rational and effective treatment regimen lies in the lack of a clear understanding of the foundational pathophysiological processes causing this aptly named complex condition [16].

Understanding of CRPS has increased over the decade. Recently both CRPS-1 and-2 have been postulated and proven to be due to a nerve injury [17]. After nerve injury, CRPS development is multifactorial. The common factors being neurogenic inflammation, autonomic dysfunction and changes in central nervous system neuroplasticity. CRPS affects both peripheral and central nervous system thus it can occur not just in limbs but also in the trunk. The pain observed in the patients is reported to be due to local afferent fibres releasing histamine and bradykinin, which causes development of allodynia, hyperalgesia and central sensitization. Decreased catecholamine flow causes vasodilation and endothelial injury tends to cause increased nitric oxide which then cause

circulatory impairment. Peripheral sensitization increases inflammatory cytokines. There is acute tissue damage mediated by IL1, IL6, TNF-alpha, CD4, macrophages, neutrophils and neurogenic mediated pro-inflammatory cytokines and neuropeptides which are released directly by nociceptors [2,14–18]. This pathophysiology is not limited to extremities.

4. Conclusion

There is nothing in the pathophysiology of CRPS that precludes it from occurring in the trunk. We therefore suggest to the scientific community that description of CRPS and any further diagnostic criteria should not be limited to extremities only, as this can further lead to under diagnosis of an already under diagnosed condition.

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Ethical approval

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Consent

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

Author contribution

LJ and TMJ contributed in study concept and design. LJ, TMJ and SM data collection. ALL authors data analysis, interpretation and editing and reviewing the paper. LJ wrote the first draft.

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N/a.

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Luxwell Jokonya.

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Declaration of competing interest

Authors declare no conflict of interest.

References

- [1] R. Weissmann, Y. Uziel, Pediatric complex regional pain syndrome: a review, *Pediatr. Rheumatol.* 14 (1) (2016) 1–10, <https://doi.org/10.1186/s12969-016-0090-8>.
- [2] H. Shim, J. Rose, S. Halle, P. Shekane, Complex regional pain syndrome: a narrative review for the practising clinician, *Br. J. Anaesth.* 123 (2) (2019) e424–e433, <https://doi.org/10.1016/j.bja.2019.03.030>.
- [3] Y. Takahashi, T. Tominaga, K. Okawa, K. Tanaka, Recovery from acute pediatric complex regional pain syndrome type i after ankle sprain by early pharmacological and physical therapies in primary care: a case report, *J. Pain Res.* 11 (2018) 2859–2866, <https://doi.org/10.2147/JPR.S164708>.
- [4] S. Castillo-guzmán, T.A. Nava-obregón, D. Palacios-ríos, J.Á. Estrada-cortinas, Complex regional pain syndrome (CRPS), a review, *Med Univ.* 17 (67) (2015) 114–121, <https://doi.org/10.1016/j.rmu.2015.03.003>.
- [5] Misidou C, Papagoras C. Complex Regional Pain Syndrome: An update. *Mediterr J Rheumatol.* 2019;30(1):16–25. doi:10.31138/mjr.30.1.16.
- [6] D.E. Logan, S.E. Williams, V.P. Carullo, R.L. Claar, S. Bruehl, C.B. Berde, Children and adolescents with complex regional pain syndrome: more psychologically distressed than other children in pain? *Pain Res. Manag.* 18 (2) (2013) 87–93, <https://doi.org/10.1155/2013/964352>.
- [7] E.L. Goh, S. Chidambaram, D. Ma, Complex regional pain syndrome: a recent update, *Burn Trauma.* 5 (2017) 1–11, <https://doi.org/10.1186/s41038-016-0066-4>.
- [8] F. Birklein, V. Dimova, Complex regional pain syndrome—up-to-date, *Pain Rep.* 2 (6) (2017) 1–8, <https://doi.org/10.1097/PR9.0000000000000624>.
- [9] Zhou L, Chou H, Holder E. Abdominal wall Type-I complex regional pain syndrome treated effectively with peripheral nerve field stimulation: a case report. *J Surg Case Reports.* 2017;2017(1):rjw222. doi:https://doi.org/10.1093/jscr/rjw222.
- [10] R.A. Agha, T. Franchi, C. Sohrabi, et al., The SCARE 2020 guideline: updating consensus surgical CAse REport (SCARE) guidelines, *Int. J. Surg.* 84 (2020) 226–230, <https://doi.org/10.1016/j.ijssu.2020.10.034>.
- [11] S. Grieve, R.S. Perez, F. Birklein, et al., Core outcome measures for complex regional Pain syndrome clinical trials (COMPACT): development of a core measurement set, *Pain.* 158 (6) (2017) 1083–1090, <https://doi.org/10.1097/j.pain.0000000000000866>. Recommendations.
- [12] J. Eldufani, N. Elahmer, G. Blaise, A medical mystery of complex regional pain syndrome, *Heliyon.* 6 (2) (2020), e03329, <https://doi.org/10.1016/j.heliyon.2020.e03329>.
- [13] K.D. Bharwani, W.A. Dik, M. Dirckx, F.J.P.M. Huygen, Highlighting the role of biomarkers of inflammation in the diagnosis and Management of Complex Regional Pain Syndrome, *Mol. Diagnosis Ther.* 23 (5) (2019) 615–626, <https://doi.org/10.1007/s40291-019-00417-x>.
- [14] V Pergolizzi J, LeQuang JA, Nalamachu S, Taylor R, W Bigelsen R. The Budapest criteria for complex regional pain syndrome: The diagnostic challenge. *Anesthesiol Clin Sci Res.* 2018;02(01). doi:10.35841/anesthesiology.2.1.1-10.
- [15] R. Munisamy, H. Uppalu, R. Raghavendra, N.P.S. Venkata, S. Harshita, S. V. Janarthanam, Type-I complex regional pain syndrome of umbilical port site: an unforeseen complication of laparoscopic surgery, *J. Minim. Access Surg.* 8 (2) (2012) 50–53, <https://doi.org/10.4103/0972-9941.95535>.
- [16] J. David Clark, V.L. Tawfik, M. Tajerian, W.S. Kingery, Autoinflammatory and autoimmune contributions to complex regional pain syndrome, *Mol. Pain* 14 (2018), <https://doi.org/10.1177/1744806918799127>.
- [17] J. Marinus, G.L. Moseley, F. Birklein, et al., Clinical features and pathophysiology of complex regional pain Syndrome – current state of the art, *Lancet Neurol.* 10 (7) (2017) 637–648, [https://doi.org/10.1016/S1474-4422\(17\)70106-5](https://doi.org/10.1016/S1474-4422(17)70106-5). Clinical.
- [18] T. Bar-Shalita, A. Livshitz, Y. Levin-Meltz, D. Rand, L. Deutsch, J.J. Vatine, Sensory modulation dysfunction is associated with complex regional pain syndrome, *PLoS One* 13 (8) (2018) 1–13, <https://doi.org/10.1371/journal.pone.0201354>.