CRPS presenting on the Acute Medical Unit: an under-recognised cause of limb swelling

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Introduction

Complex regional pain syndrome (CRPS) is defined as a disorder of the extremities characterised by regional pain that is disproportionate in time or degree to the usual course of any known trauma or other lesion. It frequently begins following a fracture, soft tissue injury or surgery.

The patient is not restricted to a specific nerve territory or dermatome and usually has a distal predominance of abnormal sensory, motor, sudomotor, vasomotor and/or trophic findings. The syndrome shows variable progression over time.

The goals of management are to restore movement and strength of the affected limb. Bisphosphonates may be effective for reducing pain in patients with early CRPS who have abnormal uptake on bone scan, even though their positive effects in this condition are probably not related to their antiresorptive properties.

Case report

A 22 year old female presented to the Acute Medical Unit (AMU) with an insidious onset of a painful swollen left arm of 4 weeks’ duration. She had been experiencing stiffness in the left hand and wrist for the preceding 2 days. She was an active horse rider although denied recent trauma. Her past medical history was pertinent for juvenile idiopathic arthritis (JIA) although she had been asymptomatic and off disease modifying therapies since the aged of 14. She also suffered from anxiety and depression for which she was taking Sertraline.

On examination, the left arm revealed subtle swelling and purple discoloration from the proximal forearm down to the metacarpophalangeal joints. The arm felt warm to touch and was exquisitely tender throughout. There was no demonstrable synovitis or pitting oedema. Pulses were equal in the upper limbs.

The patient fulfilled 4 out of the 4 Budapest criteria for CRPS. She was seen promptly by the rheumatology team.

Diagnostics CRPS

Table 1. The Budapest Criteria: In order to make a clinical diagnosis of CRPS, the following four criteria must be met.

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>Causes</th>
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<tbody>
<tr>
<td>Hypersensitivity</td>
<td>Allodynia</td>
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<tr>
<td>Hyperalgesia</td>
<td>Temperature asymmetry: Changes in skin color; Skin color asymmetry</td>
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<tr>
<td>Oedema</td>
<td>Sweating changes: Sweating asymmetry</td>
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<tr>
<td>Changes in movement</td>
<td>Decreased range of motion (motor dysfunction); Motor changes (e.g., nystagmus, tremor, etc.)</td>
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The patient was treated promptly with intravenous pulses of Pamilonate (total course 240 mg) alongside Gabapentin 100mg TDS and Sertraline 50mg OD. She made an excellent recovery and returned to normal function within a few weeks.

Managing CRPS

A multidisciplinary approach is essential for the management of CRPS. Clinical experience suggests that treatment is more effective when begun early in the course of the disease, ideally as soon as the diagnosis is established and before radiographic changes appear. Physiotherapy and Occupational therapy are considered first-line for CRPS, though most of the relevant studies are limited by methodological problems.

Investigations

Initial inpatient investigations failed to identify a cause: bloods (including inflammatory markers, D-dimers and immunology), plain radiographs, ultrasound and MRI were all normal. Subsequently, an in-patient dynamic SPECT-CT was done that showed increased uptake at the left wrist (Figure 2).

Managing CRPS

Multiple treatment modalities are available to provide pain relief in patients with CRPS. The key to success is to use whatever works to reduce pain so that patients can tolerate physiotherapy. Drug classes that have demonstrated some scientific evidence to treat CRPS include: NSAIDs, anticonvulsants, antidepressants, bisphosphonates, topical lidocaine or capsaicin, nasal calcitonin or oral corticosteroids.

Conclusion

The Budapest criteria, based on consensus and expert opinion, offer a virtual menu of signs and symptoms and result in a syndrome that defies clear understanding of the syndrome. A better knowledge of CRPS, its aetiology and its mechanisms are urgently needed.

The main features of CRPS are pain, sensory disturbance, motor impairments, autonomic symptoms and trophic changes in the affected limb. The onset of CRPS generally occurs within four to six weeks of any inciting event. The initial symptoms usually include pain, erythaema and swelling of the affected limb. In most cases, the limb is warm initially.

CRPS in adults more commonly occurs in the upper limbs. Involvement of both upper and lower limbs in the same patient is unusual. However, the symptoms of CRPS may spread over time to involve adjacent areas of the affected limb or, occasionally, other ipsilateral or contralateral limbs.

Learning points

- The diagnosis of CRPS is largely clinical and one of exclusion although often mimics a deep vein thrombosis (DVT)
- The Budapest criteria can support the diagnosis
- CRPS does not depend upon the results of imaging studies although bone scintigraphy performed promptly after onset of symptoms can support the diagnosis if there is increased radiotracer uptake during the mineralisation (ie, third) phase.

Acknowledgements

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