Sympathectomy for Causalgia: Experience with Military Injuries

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Background: Causalgia is a rare disease in civilian practice, and most reports regarding causalgia in the literature are from major extended wars. To increase awareness of this syndrome, our wartime experience with this disease is presented.

Methods: The charts of patients with causalgia referred to two university hospitals for treatment from 1985 to 1989 were retrospectively studied. Characteristics of the cause, clinical manifestations, and the response to therapy were evaluated.

Results: Among 1,564 patients with peripheral nerve injuries, there were 54 cases (3.4%) of causalgia. All of the patients, except five, were injured in battle by high-velocity bullets or missiles. Upper extremities were involved in 28 patients (52%) and lower extremities in 26 patients (48%). The most common presenting symptoms were as follows: burning pain, 100%; wet extremity, 100%; cold extremity, 93%; sensitivity to cold, 89%; paresthesia, 78%; and color changes in the extremities, 55%.

In 48 patients (89%), pain was relieved by sympathetic block (3 patients had permanent cure). Six patients had no response to the blocks (11%). Of 45 patients who had temporary relief, all underwent sympathectomy. All of these patients had complete relief of symptoms in the immediate postoperative period and for follow-up from 1 to 6 years.

Conclusion: Causalgia is essentially a war casualty disease. The condition is associated with burning pain, hyperesthesia, and symptoms of sympathetic overactivity. Sympathectomy is effective and the treatment of choice, particularly for patients who respond temporarily to sympathetic blocks.

Key Words: Causalgia, Sympathetically maintained pain, Sympathetic block, Sympathectomy


Causalgia is an uncommon condition and is believed to have been first recognized by Denmark in 1812. The word “causalgia” was first coined by Mitchell in 1867 and describes a condition that is characterized by an extremely painful syndrome that develops after a penetrating wound causing a partial injury to a large peripheral nerve. Associated with the pain are autonomic dysfunction and hyperesthesia.

The word “causalgia” comes from the Greek and means “heat” (κυστός) and “pain” (αλγος), i.e., burning pain. Causalgia, or major causalgia, should be differentiated from minor causalgia and other posttraumatic pain syndromes, such as reflex sympathetic dystrophy, Sudeck atrophy, sympathalgia, and posttraumatic edema. These syndromes are presumed to result from a reflex disturbance initiated by the injury or inflammation in which effective therapy necessitates measures not ordinarily required to manage the inciting cause. In the literature, patients with the same clinical manifestations are variously labeled by the above terminology.

Although some patients with different manifestations of sympathetically induced pain have been treated medically with various results, the only effective treatment for major causalgia is thought to be complete interruption of all sympathetic pathways to the affected limb, usually achieved by surgical sympathectomy. Patients with other posttraumatic pain syndromes are significantly less responsive to this mode of therapy. This emphasizes the importance of requiring both a complete descriptive definition of the clinical manifestations of causalgia and the responses to therapeutic interventions to anticipate the need for surgical ablation.

Causalgia is rarely seen except during wartime and is consequently unfamiliar to most physicians whose training and experience are limited to civilian practice. As a result, the diagnosis is frequently delayed or missed, causing protracted suffering and increased disability. To our knowledge, the last major report, consisting of a group of 20 patients with causalgia, was more than a decade ago. To increase awareness of this syndrome and to identify patients most likely to benefit from surgical sympathectomy, a review of its salient features and surgical outcomes in our experience with 54 patients treated at our institute is presented.

MATERIALS AND METHODS

The records of patients referred for treatment of causalgia to the departments of general and vascular surgery at Shohada and Khatam Medical Centers affiliated to Shahid Beheshti University of Medical Sciences, Tehran, Iran, between 1985 to 1989 were reviewed retrospectively. Both centers are teaching hospitals, one of which is a Level I trauma center. These centers are considered tertiary referral...
centers for trauma and vascular surgery, and most of the patients in this series were referred from other medical centers throughout the country. Data collected included age, sex, mechanism of injury, nerve and limb involved, associated injuries, and time from injury to onset of pain. The clinical manifestations included type and distribution of pain, sweating, limb warmness, color changes, and sensitivity to cold. Treatments involved the response to sympathetic blocks and sympathectomy.

As originally recommended by White et al., the diagnosis of causalgia was based on the criteria listed in Table 1. Sympathetic block was performed in every patient to further confirm the diagnosis.

**Sympathetic Block**

The stellate ganglion was anesthetized with 15 mL of lidocaine to produce an upper extremity sympathetic block. The block was considered successful if Horner syndrome and a warm dry extremity were produced. Alternatively, for the lower extremity, 20 mL of lidocaine was injected around the first lumbar ganglion. A warm, dry extremity after the injection was considered a successful block. A negative response was defined as no pain relief despite an otherwise successful block. However, if pain was relieved with the block, the diagnosis of causalgia was confirmed. Patients, in whom the response was temporary, were scheduled for surgical sympathectomy.

**Sympathectomy (Technique)**

For thoracic sympathectomy, either a limited standard lateral thoracotomy or an axillary thoracotomy (performed in most patients) was used according to the surgeon’s preference. Resection included half of the stellate ganglion and continued distally down to at least the fourth thoracic ganglion. A chest tube was then inserted to assure complete expansion of the lung postoperatively.

Two approaches were also used for lumbar sympathectomy. Most patients underwent a posterior muscle-splitting incision. However, in some patients, a nephrectomy approach was chosen on the basis of surgeon preference. The first to third lumbar ganglia were excised in every patient. If bilateral lumbar sympathectomy was required (as in one patient), the first lumbar ganglion was left untouched to prevent ejaculatory dysfunction.

**RESULTS**

Among 1,565 patients operated on for peripheral nerve injuries, there were 54 cases of causalgia (3.4%). Only one female patient was present in the entire series. Ages ranged from 18 to 59 years (mean, 21.6) (Fig. 1). Fifty patients (79%) were injured in battle: 35 by missile fragment, 14 by bullet, and 1 injured by chemical gas weapons. Three of our patients had iatrogenic peripheral nerve injuries: two after first-rib resections for thoracic outlet syndrome and one (female) after anterior fusion of lumbar spine plus laminectomy for nerve decompression. One patient was stabbed by a knife, an injury that was not war related.

Except for the one patient with a chemical injury, at least one major nerve injury, usually partial, was present in all other patients. In 28 patients (52%), the upper extremity was involved; the lower extremity was involved in the other 26 patients (48%). The prominent nerve injured was the median nerve in the upper extremity and the sciatic nerve or its terminal major branches in the lower extremity. Associated injuries included abdominal injuries in 8 patients, thoracic injuries in 6 patients, bone fractures in 18 patients, major lacerations in 25 patients, soft tissue injuries in 44 patients, vascular injuries in 14 patients, and chemical gas burn with respiratory insult in 1 patient.

The time from injury until onset of pain is shown in Figure 2. The majority of patients (89%) experienced pain within 24 hours. The most common presenting symptom was burning pain, which was present in all of our patients. Burning pain usually did not correlate with the skin distribution of the injured nerve. Involvement of the entire extremity distal to the site of injury was the usual presentation. In no case did pain extend to the trunk. There were symptoms of excessive sympathetic activity in most of the patients (Fig. 3).

Six patients (11%) did not respond to sympathetic blocks. They were treated medically. Three of these eventu-

<table>
<thead>
<tr>
<th>Table 1 Diagnostic Criteria&lt;sup&gt;a&lt;/sup&gt;</th>
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<tr>
<td>History of penetrating trauma specially high velocity (bullets/missiles)</td>
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<tr>
<td>Major peripheral nerve injury</td>
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<tr>
<td>Burning pain</td>
</tr>
<tr>
<td>Sympathetic symptoms, e.g., wet extremity, sensitivity to cold</td>
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<td>Pain aggravation by physical and/or emotional stimuli</td>
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<sup>a</sup> No reliable paraclinical test available to date.

Fig. 1. Graph showing the age distribution of patients.

Aggressive physiotherapy was always started as soon as tolerable for the patient. Patients were either followed in clinic, or by correspondence for 1 to 6 years.
ally underwent sympathectomy because of unrelenting severe pain; one patient had complete relief of pain, one patient improved, and one patient remained unchanged. Forty-eight of the patients (89%) responded to sympathetic block. Of these, 3 patients (6%) had permanent cure after three sessions and needed no further therapy, and 45 patients (83%) who had temporary relief underwent sympathectomy. All of these patients had complete relief of symptoms in the immediate postoperative period, which lasted until the time of this study. Therefore, a total of 49 patients (91%) were cured by either temporary or permanent sympathetic interruption. One additional patient (2%) benefited partially from sympathectomy.

Two patients had operative complications (4%). One patient developed a gram-negative empyema that required treatment with a chest tube and a hospital stay of 1 week. Another patient had a lumbar incision infection requiring drainage. There were no deaths. Hospital stay was from 1 to 7 days.

DISCUSSION

The great majority of reported cases of causalgia are from war casualties. The very first case of causalgia was described by Denmark1 from a battle-related injury. Mitchell et al.2,14 first described and named the syndrome during the American Civil War, and the subsequent bulk of cases reported in the literature were published during major wars (Table 2). In our series, 49 patients (91%) had high-velocity injuries caused by war-related trauma.

Causalgia is usually rare in the civilian environment, with iatrogenic injury to the brachial plexus being a major cause.15 The second most common cause of causalgia in our

Table 2 Clinical Presentation and Response to Treatment of Causalgia a

<table>
<thead>
<tr>
<th>Series</th>
<th>Time of Conflict</th>
<th>Patient (n)</th>
<th>Partial Nerve (%):</th>
<th>Burning Pain (%):</th>
<th>Paresthesia (%):</th>
<th>Sympathetic Activity b</th>
<th>Response</th>
<th>Block</th>
<th>Sympathectomy</th>
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<tbody>
<tr>
<td>Mitchell et al. 14</td>
<td>CW</td>
<td>7</td>
<td>3/3</td>
<td>7</td>
<td>5/5</td>
<td>↓ &gt; ↑</td>
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<tr>
<td>Carter 21</td>
<td>WWII</td>
<td>24</td>
<td>n/r</td>
<td>10/10</td>
<td>22/22</td>
<td>↓</td>
<td>N/A</td>
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<tr>
<td>Albritten and Maltby 5</td>
<td>WWII</td>
<td>67</td>
<td>65/67</td>
<td>67</td>
<td>n/r</td>
<td>n/r</td>
<td>30/30</td>
<td>30/30</td>
<td></td>
</tr>
<tr>
<td>Mayfield 7</td>
<td>WWII</td>
<td>105</td>
<td>105</td>
<td>105</td>
<td>75/75</td>
<td>↓ &gt; ↑</td>
<td>105/105</td>
<td>98/98</td>
<td></td>
</tr>
<tr>
<td>Rasmussen and Freedman 6</td>
<td>WWII</td>
<td>100</td>
<td>n/r</td>
<td>55</td>
<td>n/r</td>
<td>↓ &gt; ↑</td>
<td>89/91</td>
<td>35/35</td>
<td></td>
</tr>
<tr>
<td>Freeman 8</td>
<td>WWII</td>
<td>114</td>
<td>Often</td>
<td>7/42</td>
<td>114</td>
<td>no change</td>
<td>114/114</td>
<td>17/17</td>
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<td>Shumacker et al. 9,24</td>
<td>WWII</td>
<td>90</td>
<td>89/90</td>
<td>90</td>
<td>90</td>
<td>↓ &gt; ↑</td>
<td>82/83</td>
<td>56/57</td>
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</tr>
<tr>
<td>Barnes 10</td>
<td>KC</td>
<td>48</td>
<td>48</td>
<td>48</td>
<td>n/r</td>
<td>n/r</td>
<td>27/27</td>
<td>24/24</td>
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<tr>
<td>Baker and Winegarner 11</td>
<td>VC</td>
<td>28</td>
<td>28</td>
<td>n/r</td>
<td>n/r</td>
<td>↓</td>
<td>32/7</td>
<td>28/32</td>
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<tr>
<td>Bucketer et al. 12</td>
<td>VC</td>
<td>24</td>
<td>n/r</td>
<td>n/r</td>
<td>n/r</td>
<td>↑</td>
<td>8/8</td>
<td>16/16</td>
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<tr>
<td>Ghostine et al. 22</td>
<td>LCW</td>
<td>40</td>
<td>40</td>
<td>40</td>
<td>n/r</td>
<td>↑</td>
<td>15/15</td>
<td>20/20</td>
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<tr>
<td>Jebra and Saade 13</td>
<td>LCW</td>
<td>20</td>
<td>20</td>
<td>18</td>
<td>n/r</td>
<td>↓</td>
<td>48/54</td>
<td>50/51</td>
<td></td>
</tr>
<tr>
<td>Present study</td>
<td>IIW</td>
<td>54</td>
<td>53/54</td>
<td>54</td>
<td>42</td>
<td>↑</td>
<td>518/527</td>
<td>374/380</td>
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Total 721 451/455 (99%) 501/538 (93%) 436/448 (97%) 518/527 374/380

N/A, not performed at that time; n/r, not reported in the paper; CW, American Civil War; WWI, World War I; WWII, World War II; KC, Korean Conflict; VC, Vietnam Conflict; LCW, Lebanon Civil War; IIW, Iran-Iraq War.

a Reports were chosen to represent the majority of patients presented in the English literature in different war periods. Some papers do not report all data for all patients; therefore, the data are shown in that fraction of patients reported.

b Based on the symptoms, e.g., sweating, warmness, color and sensitivity to cold in the involved extremity.
series (three patients [6%]) was the occurrence of complications after surgery associated with major nerves. Two patients undergoing first-rib resection for thoracic outlet syndrome developed the symptoms of causalgia immediately after surgery. This was probably attributable to excessive traction on the arm for better exposure through the axillary thoracotomy, causing partial distraction injury of the brachial plexus. Both of these patients responded to sympathetic block and subsequent sympathectomy that was performed in the first postoperative month. A third patient (the only female in the series) was referred from the neurosurgery department for causalgic pain after an anterior fusion of the lumbar spine. There are other isolated cases of causalgia reported after discectomy\textsuperscript{16} or laminectomy.\textsuperscript{17} However, to our knowledge, this is the first report of causalgia after spinal fusion.

One patient in our series presented with typical manifestations of causalgia after exposure to a chemical attack. The patient had no physical injuries except the skin lesions associated with chemical contamination. He developed symptoms within 24 hours after exposure, and his diagnosis was confirmed by sympathetic block. His causalgia responded to sympathectomy; however, his other symptoms from the chemical exposure required further therapy. In one study, exposure to capsaicin reproduced the symptoms of causalgia.\textsuperscript{18} However, to our knowledge, this is the first report of causalgia as a result of chemical warfare.

All of our patients, except the patient with chemical burns, had peripheral nerve deficits consistent with a major nerve injury. Results of clinical examination demonstrated the injury to be partial in all cases, and this was confirmed during exploration for the treatment of the nerve injury. Of all past reports (Table 2), more than 99\% of cases had partial major nerve injuries. This distinguishes causalgia from other sympathetically maintained pain syndromes, such as reflex sympathetic dystrophy, Sudeck atrophy, sympathalgia, and posttraumatic edema, in which the inciting cause is not penetrating partial nerve injury.\textsuperscript{19}

In our series, the ratio of upper (52\%) to lower (48\%) extremity involvement was similar to that in other reports.\textsuperscript{20} In the literature, involvement of the median nerve either alone or combined with other peripheral nerves of the upper extremity and the sciatic nerve or its major terminal branches in the lower extremity is the usual presentation.\textsuperscript{6,9–11,13,14,21,22} The same distribution was observed in our patients.

Mitchell et al.\textsuperscript{14} emphasized that burning pain was a constant feature. Our observations and those of other investigators support this.\textsuperscript{4,5,10,13,15,22–24} However, in the only series of patients reported from World War II, a substantial proportion of the patients had no burning pain.\textsuperscript{6,8} Moreover, it is stated that these exceptional cases had a clinical picture quite similar to cases of minor causalgia, as originally reported by Homans.\textsuperscript{25} It must be emphasized that burning pain is a constant and specific feature of causalgia, and there is significant reason to question whether patients reported as having no burning pain have true causalgia.

A characteristic feature of the pain of causalgia is aggravation by certain physical and emotional stimuli.\textsuperscript{4–8,10,13–15} Very mild stimuli, such as the sound of traffic, noise of other patients, or even water falling from a water tap may aggravate the symptoms. Typically, patients keep the involved extremity in a moist towel, guarded, immobile, and supported, if possible. If not familiar with this syndrome, physicians frequently consider these patients to have an underlying psychotic disorder.\textsuperscript{26} In our experience, these patients that excessively focus on their extremity and are detached from their environment, return to baseline, normal behavior after successful sympathetic block or sympathectomy.

The pain started either immediately or within 6 hours in 63\% of the patients. Only six patients (11\%) reported their onset of pain more than 24 hours after injury. Similarly, Richards in a collected series of 310 cases found that 2 of 3 of the patients had pain begin either immediately or very shortly after injury.\textsuperscript{20} Other associated distracting injuries, e.g., shock, surgical interventions, and anesthesia for other emergent injuries, may explain why some patients identify a later onset. The time of onset is a major differentiating factor of causalgia from other sympathetically maintained pain syndromes, which typically develop burning pain weeks after injury. In addition, all of our patients reported their pain as distal in the extremity. Frequently, pain would involve the extremity up to the level where the nerve injury occurred; in no patients did the pain extend to the trunk. If pain affects the extremity proximal to the site of nerve injury, or if the pain follows the distribution of a specific injured nerve, one must question the diagnosis of causalgia.\textsuperscript{2,13,14,24}

The subcommittee on taxonomy of the International Association for the Study of Pain (IASP) further defines a typical presentation of cool, reddish, and clammy skin.\textsuperscript{19} This proved to be true in our series in patients whose symptoms included cold extremity (93\%), sensitivity to cold (89\%), increased sweating (100\%), and red skin (55\%). Thus, causalgia illustrates the classic clinical manifestations of increased sympathetic tone. To diagnose causalgia, the galvanic skin response and plethysmography have been proposed to document sympathetic overactivity.\textsuperscript{19} However, their efficacy and practicality have yet to be confirmed. Currently, chemical sympathetic blockade is the sole reliable method for confirming the clinical diagnosis of causalgia (Table 2). In our patients who were clinically diagnosed with causalgia, all but six (89\%) had cessation of pain at the induction of sympathetic anesthesia. The results were both immediate and spectacular. The patient, in agonizing pain, guarding the injured extremity and emotionally detached from the environment, has immediate pain relief, and sympathetic overactivity disappears as soon as the injection occurs. However, the pain usually recurs when anesthesia ends. None of the patients had only partial relief of their symptoms after a block. The sympathetic block not only establishes the diagnosis, but also predicts the patient’s response to surgical sympathectomy.
In three of the patients who responded to sympathetic block, the relief was permanent and no other intervention was required. Some authors recommend repeated injections in the hope of cure, and many of our patients had more than one block before referral. However, we recommend early sympathectomy to prevent permanent disabilities and enhance active physical therapy in patients who demonstrate only a temporary response to sympathetic block, while optimally treating associated problems such as nerve injuries.

Multiple alternative approaches to treating this condition have proved unsuccessful. These include neurolysis, repeated resection of neuroma, proximal resection or chemical block of the affected nerve trunk, periarterial sympathectomy, and division of the posterior spinal roots. Definitive treatment of the inciting nerve injury has been reported to be totally ineffective for relief of severe causalgia, and, if nerve repair is indicated, should be undertaken only after the burning pain has been relieved by sympathectomy. While sympathectomy was first suggested by Leriche in 1917, Spurling has been credited for establishing sympathectomy as a curative measure for causalgia in 1930. Once causalgia is confirmed, immediate sympathectomy should be performed, inasmuch as spontaneous disappearance of causalgia pain occurs rarely. The operation for causalgia in both the upper and lower extremities is simple and safe, and the postoperative period of disability is short. Our patients spent 1 to 7 days in the hospital (the majority of patients were discharged within 2 days). Sympathectomy for the upper extremity has also been performed thoracoscopically, and this may be considered the method of choice. There have been no long-term complications in any of our patients, and two operative complications were managed eventually. Therefore, one should operate early, before the patient becomes dependent on narcotics or develops irreversible atrophic musculoskeletal changes and loses motion in affected joints.

Failure to achieve pain relief after resection is attributable to incomplete resection. When sympathetic denervation is complete, the causalgia pain disappears. To achieve complete sympathetic denervation, we resect half of the stellate ganglion down to not less than the fourth thoracic ganglion for upper extremity lesions and the first to third lumbar ganglion for lower extremity involvement. In patients with bilateral lumbar sympathectomy, sparing of the first lumbar ganglion preserves ejaculation function while achieving relief. In general, the area of sympathetic denervation must reach well above the level of nerve injury. Because surgery was performed early in the course of their disease, no correlation can be drawn between duration of disease and outcome of treatment.

Finally, the exact pathophysiology of causalgia remains uncertain. Many interesting theories have come from knowledgeable students of the subject. One attractive explanation is that causalgia is caused by fiber crossover at the site of the nerve injury between efferent sympathetic and afferent sensory fibers. The alternative hypothesis of peripheral receptor up-regulation and subsequent pathologic response to circulating catecholamines in the affected limb, although interesting, requires further investigation in patients with causalgia.

CONCLUSIONS
Surgeons in general, and particularly those dealing with penetrating injuries, need to be conscious of the possibility of causalgia developing in their patients. Causalgia occurs only with incomplete major peripheral nerve injuries. The disorder is characterized by burning pain, hyperesthesia, and manifestations of sympathetic overactivity localized to the injured extremity. The burning pain starts soon after the injury and is unusually aggravated by emotional and/or physical stimuli. Chemical block of the appropriate sympathetic chain is a critical diagnostic procedure. Sympathectomy should be performed soon after the diagnosis is established to prevent the trauma of prolonged pain and crippling joint dysfunction. Sympathectomy is a safe procedure, and the dramatic resolution of the causalgia pain is noticed immediately after denervation. Recovery of function is frequently rapid after relief of pain. Thoracoscopic sympathectomy may become the treatment of choice for causalgia of the upper extremities. The pathologic mechanism is still obscure and an interesting subject for further investigation.

REFERENCES
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