A 45-year-old female dentist fell on a slippery floor at work and immediately felt pain in her right wrist. Her physician diagnosed a sprained wrist without bony fracture or rupture of the ligaments. The physician wrapped a bandage around the wrist, and anti-inflammatory analgesics were prescribed to alleviate the pain. Rest, ice application, and elevation of the hand higher than the elbow were recommended, and after 3 or 4 days, everything seemed fine. However, in the following week, she felt a burning pain in her hand and forearm that became steadily worse. She also noted swelling over the dorsum of her right hand. In subsequent months, the dentist was having difficulties in performing her job owing to muscle weakness and severe pain with movement of her arm. She also noticed sweating in the region of the painful area, as well as increased growth of her fingernails and of the hair on the dorsum of her right hand.

Three months after her injury, the dentist visited a university pain clinic. On physical examination, her orthopedist observed erythematous, edematous, and shiny skin on the swollen distal part of her forearm and the dorsal side of her hand. Passive and active range of motion of her wrist was restricted. Physical systemic examination and blood test results revealed no abnormality. On neurologic examination, hyperalgesia and allodynia were noted in her right forearm, wrist, and hand. Results of nerve conduction studies were normal. Grip strength was 7 kilograms and 24 kg in the right and left wrists, respectively. Radiographs showed decalcification in her right carpal bones. Thermography revealed a temperature 1.0°C higher than on the unaffected side.

EXPLANATION

Given this symptom history and these clinical findings, this dentist is likely to have complex regional pain syndrome (CRPS), which is a chronic painful disorder that is primarily a disease of the limbs. It has been called causalgia, reflex sympathetic dystrophy, Sudeck atrophy, shoulder-hand syndrome, algodystrophy, peripheral tropheneurosis, sympathetically maintained pain, and posttraumatic pain syndrome. In 1993, the International Association for the Study of Pain agreed on the descriptive term CRPS to avoid the question of etiology or mechanism. CRPS type I replaced the term reflex sympathetic dystrophy, and CRPS type II replaced the term causalgia. The previously used term sympathetically maintained pain now is used only for defining a symptom and might be a feature of several painful diseases; it may be found in association with CRPS as well, but it is not an essential requirement for diagnosing CRPS.

CRPS types I and II typically arise as a result of minor to severe trauma that leads to sensory, autonomic, motor, and trophic changes. Trauma severity and CRPS symptoms are not proportionate. CRPS type I is distinguished from CRPS type II by the absence of obvious nerve lesions, and CRPS type II is classified under neuropathic pain. Researchers have reported that CRPS may occur in both sexes but predominantly in women, and it is most common in people aged 55 to 75 years. The incidence of CRPS type I is higher than that of CRPS type II.

The internationally accepted Budapest criteria are helpful for clinical diagnosis of CRPS. The box shows the 2013 version of the clinical criteria; there are also research diagnostic criteria presented in the same article. There is no specific diagnostic test for CRPS, but some tests, including thermography, radiographs, and sympathetic blocks, may be useful. Ruling out serious neurologic disease may require imaging of the brain and spinal cord.

Although researchers have proposed various theories to explain the pathophysiology of CRPS, it is not well understood as yet. Neurogenic inflammation, autonomic dysfunction, and neuroplastic changes are possible mechanisms to account for CRPS development. Evidence suggests that both peripheral and central mechanisms play a role in the syndrome’s pathogenesis. A genetic predisposition and psychological dysfunction may play a role as well.

Clinical symptoms change throughout the course of CRPS in accordance with its pathophysiology. Onset of the clinical symptoms is not always seen immediately after initiating events, and it may delay for months. In the early stages, the affected area is warm, painful, red, swollen, and sweaty. The pain is spontaneous or stimuli evoked, and it can be continuous, paroxysmal, or episodic. The pain may be described as aching, burning, pricking, or shooting in character, and it feels localized deep in somatic tissue.
Increased hair and nail growth are usually present within a few weeks after the initiating event, and 50% of cases have these trophic changes. Muscle weakness occurs during the acute stage as indicative of motor abnormality, which is seen in approximately 50% of patients with CRPS. Initially, the symptoms are confined to the injured tissue but not necessarily within the same nerve root. Alldynia and hyperalgesia with thermal and mechanical stimuli are frequently present in CRPS. Conversely, sensory loss to touch, heat, and pressure may develop. Thus, CRPS may be characterized by both positive and negative sensory disturbances. These sensory deficits are seen in almost all patients with CRPS, with glove- or stocking-like distal distribution.

In the chronic stage, hair and nail growth decreases, and atrophy of the skin with ulcerations may be present. Tremor, dystonia, exaggerated tendon reflexes, and myoclonic jerks are the other features of this stage. The skin color appears rather pale or livid with chronicity, and the skin temperature becomes colder than that on the unaffected side. This finding is contrary to findings during the acute stage, during which the skin temperature of the affected side is approximately 1°C warmer than the temperature of the unaffected side.

Over time, symptoms may begin to spread to other parts of the body contralaterally, ipsilaterally, or diagonally. Patients with CRPS may report that their involved limb feels foreign to them (cognitive neglect) or that they need to pay mental and visual attention to move their limb (motor neglect).

The management of CRPS requires an interdisciplinary approach because the occurrence and the maintenance of CRPS are not explained by only 1 pathologic pathway. A stepwise time-contingent functional restoration algorithm, involving occupational therapy, recreational therapy, physiotherapy, and vocational rehabilitation, is the main strategy for treating CRPS. The clinical team should consider using psychotherapy, as well as pharmacologic and interventional therapies, if needed, at any step.

CONCLUSIONS

Trauma to the upper extremities can compromise a dentist’s work ability considerably. Even minor trauma can lead to CRPS in vulnerable people. This condition is poorly understood, although it appears that both peripheral and central mechanisms play a role in its pathogenesis. Early diagnosis is crucial for preventing CRPS from becoming chronic; however, the signs and symptoms of CRPS mimic those of some other common diseases, which may delay diagnosis. Dental practitioners encounter many patients with orofacial pain in their daily practice, but they also should be aware of clinical features of painful conditions in other parts of the body. As this case history shows, a dentist also can be affected personally by a clinically significant pain disorder. Therefore, dental practitioners should recognize and refer any patient suspected of having CRPS (including themselves) immediately to a neurologist, rheumatologist, or pain specialist for confirming the diagnosis of CRPS and for initiation of appropriate treatment.

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