“Complex Regional Pain Syndrome” after Trauma from High-heeled Shoe

Sir,

Complex regional pain syndrome, type I (CRPS I) (reflex sympathetic dystrophy) is a syndrome which can develop even after a minor injury or operation. The reported incidence varies from 2–32% after Colles fractures and 1–5% after various other injuries (1). It is defined as follows (criteria 2–4 must be satisfied): 1) the presence of an initiating noxious event, or a cause of immobilization; 2) continuing pain, allodynia, or hyperalgesia to any inciting event; 3) evidence at some time of edema, changes in skin blood flow, or abnormal sudomotor activity in the region of pain; 4) the diagnosis is excluded by the existence of conditions that would otherwise account for the degree of pain and dysfunction. We here report 2 patients who developed CRPS I after receiving a trauma from a high-heeled shoe during dancing (no shoe protection), which was further complicated by delayed wound healing.

CASE REPORTS

Case 1

A 53-year-old woman’s right foot was stepped on (between the 4th and 5th metatarsal) by a person wearing high-heeled shoes. A 1 by 1 cm and approximately 1 cm deep wound appeared, as described by the patient. Because of edema and pain the patient went to her doctor 3 weeks later. The X-rays did not show any fracture and there was no sign of infection. Five weeks after the trauma we examined the patient. We found the right foot edematous and red, and warmer when compared to the left. A minor dry, necrotic wound was observed, from which no bacteria could be cultured. There was no apparent relief of symptoms with the use of non-steroid antiinflammatory drugs, penicillin, or local disinfecting agents. The wound finally healed after 4 months, and after that the pain slowly regressed.

The patient had been treated for hypertension for 5 years with a calcium entry blocker and a thiazide; otherwise she was healthy. The following tests were normal: toe and ankle blood pressures, hemoglobin, leukocytes and differential counts, platelet count, sedimentation rate, liver enzymes, thyroid stimulating hormone, fasting-glucose, cholesterol and triglycerides.

Case 2

A 50-year-old woman was stepped on (between the 3rd and 4th metatarsal on her right foot) by a person wearing high-heeled shoes. A 1.5 by 1 cm and approximately 1 cm deep wound appeared, as described by the patient. Approximately 2 months later the patient consulted her doctor because of increasing pain, redness and edema in the area. The symptoms increased during the day. X-rays did not show any fractures. A 1 by 2 cm ulcer was observed, which was revised twice. Since infection was suspected, the patient was treated with penicillin, but with no convincing effect.

Because of continued edema and pain in the whole foot, in particular ankle stiffness, we saw the patient 6 months after the trauma. A small, dry, necrotic ulcer was observed. The patient was treated with non-steroid antiinflammatory drugs without effect. Suspecting CRPS I, we treated the patient with prednisolone: 30 mg/day for 1 week, 20 mg/day for 1 week and finally 10 mg/day for 1 week. The pain ceased almost immediately, and after 1–2 weeks the ulcer had healed completely.

The patient was healthy, except for moderately decreased toe and ankle blood pressures: the first toes (right: 50 mmHg, left: 45 mmHg (norm >60 mmHg)), the ankles (right: 90 mmHg, left: 85 mmHg (norm >110 mmHg)). Arm blood pressure: 110/60. The following blood tests were normal: hemoglobin, leukocytes and differential counts, platelets, sedimentation rate, liver enzymes, thyroid stimulating hormone, fasting-glucose, cholesterol and triglycerides.

DISCUSSION

Our patients fulfilled the criteria of CRPS I.

The pathophysiological mechanisms are still under debate, but an exaggerated inflammatory response and /or altered activity of the sympathetic nervous system are possible mechanisms (1). Sudeck (3) first suggested an excessive inflammatory response. In the acute phase, all classical signs and symptoms of inflammation – rubor, calor, dolor, tumor and functio laesa are present. This hypothesis is further supported by the finding of increased vascular permeability of macromolecules, which is an important characteristic of inflammation (4). Furthermore, a therapeutic effect of corticosteroids has been shown (5), and today this is widely used in the clinic.

A lot of symptoms have been described and attributed to the syndrome (1), including abnormalities in skin blood flow and atrophy of the skin and nails. Delayed wound healing has not been described as far as we know and could possibly be due to both exaggerated sympathetic response and inflammation. Increased temperature of the muscles and increased blood flow, as measured by plethysmography, have been found. However, nutritive blood flow might be decreased as oxygen consumption is reduced in the limbs affected by CRPS I, and treatment with oral vasodilators might reduce or abolish pain (1).

The wound healing was delayed in both patients, 4 and 6 months, respectively, which could be explained by decreased nutritive flow following the CRPS I.

In patient number two we found moderately decreased toe and ankle pressures, but this might only explain a minor part of the symptoms, as treatment with prednisolone both cured the pain and increased the rate of healing. This indicated that an inflammatory process was going on.

Patient number one was treated with a calcium entry blocker for her hypertension, which might increase peripheral blood flow. However, it did not seem to prevent development of CRPS I but might have decreased the symptoms. Anyway, her symptoms regressed after some months without any further treatment. The spontaneous course of CRPS usually takes months or years.

CONCLUSION

We suggest that delayed wound healing may accompany CRPS I as a result of trophic changes and inflammatory mechanisms. Treatment with prednisolone has an effect on pain and may increase the rate of wound healing.

REFERENCES


Acta Derm Venereol (Stockh) 77
The Simultaneous Occurrence of Hailey-Hailey Disease, Graves' Disease and Multiple Sclerosis in the Same Patient

Sir,

Familial benign chronic pemphigus, (Hailey-Hailey disease) is a chronic vesiculo-bullous disease, characterized by its distribution along the sides of the neck, axillae and groin areas. This condition may have a hereditary auto-immune basis (1).

Multiple sclerosis is a disorder of the brain, optic nerves and spinal cord, characterized by areas of demyelination. This condition is thought to be of hereditary auto-immune basis, associated with HLA-DR2 antigen (2).

Graves' disease is an auto-immune disorder of the thyroid, characterized by the presence of circulating antibodies directed against different fractions of the thyroid gland. This disorder is also of a hereditary origin in the vast majority of cases and also has a 10:1 predilection for young women between 20-40 years (3). We report a case of a 35-year-old woman with the coexistence of Hailey-Hailey disease, Graves' disease and multiple sclerosis.

CASE REPORT

We have been following a 35-year-old white female, who for the last 15 years has had Hailey-Hailey disease. Her occasional exacerbations were controlled with topical 2% erythromycin solutions and an occasional intramuscular injection of 40 mg triamcinolone acetonide for her more severe outbreaks. More than one dozen biopsies were performed over the last 15 years to make sure that this was not some other disease process in evolution, but the biopsies always came back as consistent with Hailey-Hailey disease. Two years ago the patient stated that she was experiencing weight loss, palpitations and extreme anxiety attacks. A laboratory thyroid profile (T3, T4 and TSH) as well as levels of antimicrosomal and antithyroglobulin antibodies were indicative of Graves' disease, at which point she was referred to an endocrinologist for further evaluation and treatment. Her thyroid condition was successfully treated with radioactive iodine (I131). One year later, after she had developed limb weakness, diplopia, and retrobulbar neuritis, her family physician referred her to a neurologist for further work-up. At this point it was decided that she was developing multiple sclerosis.

She is currently under the care of her neurologist and her multiple sclerosis is causing a deteriorating condition which is progressing rapidly. Her outbreaks of Hailey-Hailey have become almost generalized and basically unresponsive to all forms of treatment at this point in time. It is interesting to note that the patient's mother also has Graves' disease and Hailey-Hailey disease. Both the patient and her mother express HLA-DR2 antigen. Whole phenotyping of the patient and her mother was not performed. This is a most curious situation, in which a previously healthy young woman now has a barrage of three autoimmune diseases, which appear to be refractory to all forms of therapy at this point. Tests for HIV, lupus and other immunological conditions have all been negative up to now.

We wish to alert our dermatologic colleagues who have occasion to treat Hailey-Hailey disease, to be on the lookout for other potential immunological abnormalities. We believe this to be the first case report of an association between Hailey-Hailey disease, Graves' disease and multiple sclerosis in the same patient.

REFERENCES


Accepted January 24, 1997.

E. K. Edwards, Jr., M.D., FACP and E. K. Edwards, Sr., M.D., Ridge-Edwards Dermatology Center and University of Miami, School of Medicine Dermatology and Cutaneous Surgery, 1800 N. Federal Hwy., Pompano Beach, Fla. 33062, USA.