Systemic Sclerosis Following Physical Trauma

Sir,

The etiology of systemic sclerosis (SSc) is still unknown. Three components – the blood vessels, the immune system and fibroblasts – play important roles in the pathophysiology of the disease (1). Disturbances of the first two systems can cause dysregulation of fibroblasts through mediators such as cytokines and growth factors as well as disturbances in cell–cell and cell–matrix interactions with fibroblasts. As in various autoimmune diseases, the pathogenesis is partly based on a genetic background and modulated by environmental factors (2).

Here we report a patient who developed SSc shortly after physical trauma – not only in the injured skin area but also in other sites, and with internal organ involvement. It is obvious that the patient has a genetic predisposition to this development.

CASE REPORT

A 54-year-old woman was involved in a traffic accident in which her left wrist was broken and her neck was whiplash injured. Several plaster casts were applied to the left forearm successfully over the next 5 weeks and the neck was bandaged for stabilization for 3 weeks. Twelve weeks later she developed Raynaud's phenomenon, swelling and stiffness of the fingers and both hands, with the tendency to tighten and ascend both forearms, and affecting the face. There was no difference between the left and the right arm. Skin biopsy confirmed the diagnosis SSc. In addition, she developed dysphagia and oesophageal dysfunction (technetium scintigraphy) and lung fibrosis (high resolution computed tomography) with reduced pulmonary diffusion capacity. Antinuclear antibodies (ANA) were 1:1280 positive with an antinucleolar pattern; anticentromer antibodies were also positive (1:1000). HLA class II typing showed the following alleles: DR3, DR5, DR52, DQB2. The patient was treated with nifedipine, ilomedin (prostacyclin) infusions, prednisolone and cyclophosphamide, and this has resulted in some improvement of her skin condition and Raynaud's phenomenon. Three years after the beginning of SSc she is now in a relatively stabile phase of the disease.

DISCUSSION

Several environmental factors have been described that can induce SSc, e.g. vinyl chloride, chlorinated aromatic and aliphatic hydrocarbons, organic solvents, epoxy resins, drugs (bleomycin, pentazocine, methylsergide), anilides, L-tryptophan and silica. In the majority they induce a scleroderma-like syndrome, partly reversible after withdrawal of the exposure (2). Contrary to this, silica can precipitate a true SSc indistinguishable from idiopathic SSc (3). In addition, exacerbation or even development of anecdotal cases of SSc have been observed after X-ray treatment in a generalized form (4).

In 1996, five cases with SSc occurring shortly after episodes of physical trauma were described (5). In addition, physical exertion was reported as a possible precipitating factor in both adult (6) and paediatric (7) eosinophilic fasciitis. In 1996, Vancheeswaran et al. noticed a significant association between trauma and childhood SSc (8). Trauma has been associated with linear scleroderma (9), which often occurred in the same site as trauma within 6 months (10).

In our case, the SSc is not limited to the area of trauma and temporal immobilization, so the SSc cannot be explained as reflex sympathetic dystrophy. In the context of internal organ involvement, trauma may have played a precipitating role in a certain genetic background with an HLA class II pattern, suggesting an increased susceptibility to development of SSc (11). HLA DR52 and DR3 and 5, in particular, may be markers of a predisposition to increased risk of lung fibrosis (12).

We can only speculate on the pathogenetic mechanism operative in our case. Certain cytokines and growth factors which may be activated and released due to wound healing can induce fibroblasts to produce more collagen.

Our case, and the data of the literature, raises the question whether trauma can be associated with the onset of SSc. Although the trauma of our patient was neither life-threatening nor did it lead to peripheral nerve injury, mental anguish and psychological stress after the traffic accident may be associated with an increased release of corticotrophin-releasing factor and activation of the sympathetic nervous system.

The question whether the trauma actually caused the disease or was only coincidental has a major legal implication. The most probable hypothesis is that physical trauma and/or emotional stress may activate subclinical or aggravate pre-existing disease. The observation of ANA, anticemtromer antibodies, and the HLA pattern favours this hypothesis.

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