LINEAR SCLERODERMA EN COUP DE SABRE

Relationship with Progressive Facial Hemiatrophy (PFH)

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ABSTRACT

We studied 58 cases of linear scleroderma of the face: 20 of them showed transition into progressive facial hemiatrophy (PFH). In several cases a distinction between these two conditions was not possible since the atrophic changes were arranged along the previous indurations, usually along the branches of trigeminal nerve. Neurological manifestations, most often epilepsy, were found in about 10% cases of PFH, and were similar in childhood cases of scleroderma en coup de sabre. The shrinking of deeper tissue and bone deformities were observed by early onset of the disease, not infrequently after trauma. In both scleroderma en coup de sabre and PFH, neurological complications are probably related to vascular involvement.

The double lines at the frontal area in 4 children were associated with epilepsy and usually pronounced deformities, whereas double lines in young adults did not produce evident wasting and shrinking.

Our study showed a close relationship of PFH with linear scleroderma of the face, and deep atrophies were comparable to the shrinking of subcutis and muscle in profound linear scleroderma of the limbs. There are however rare cases of PFH associated with atrophy of the brain and severe neurological manifestations. Their relationship with scleroderma is not known.

Progressive facial hemiatrophy (PFH) is a sporadic disease characterized by unilateral shrinking and deformity of the face, and which is associated at least in 10% of cases with central nervous system involvement [1, 2]. The etiology of this condition is unknown. A fre-

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quent coexistence with linear scleroderma of the face, morphea and linear scleroderma on various sites of the body suggests a close relationship of these conditions. [3–5].

In several cases overlapping symptoms of both conditions made a definite distinction impossible [6]. It is believed that while in PFH the primary disorder is atrophy of the subcutaneous tissue, muscle and bone without skin induration, in scleroderma sclerosis and atrophy affect primarily the skin and subcutaneous tissues, and only later pass to the deeper tissue. The atrophic changes tend to be band-like and arranged along the previous indurations, whereas in PFH they are often more confluent. However not infrequently the lesions recognized as PFH occupied limited areas resembling involutionary scleroderma. Neurological manifestations, epilepsy and occasional ocular and auditory complications, may be similar in both diseases [7–9], and in both may involve various levels of cerebrospinal and sympathetic nervous system [10–12]. PFH, both secondary to scleroderma and seemingly idiopathic, is localized mainly along branches of trigeminal nerve, with involvement of sympathetic fibers of this nerve and cervical sympathetic ganglia (1, 13). Cerebral changes both in scleroderma en coup de sabre and PFH are mainly related to vascular involvement (neurovasculitis) [2, 13].

We present 4 children with double lines of scleroderma en coup de sabre at the frontal area (Figure 1). In all of these children the disease started at the very early age, in most of them after trauma, believed to be responsible for neurologic injury [14]. Linear scleroderma of the face, by its onset in infancy, has a greater tendency to produce deep atrophy and bone deformities with not infrequent transformation into PFH. In all children

Figure 1. Double lines in a 6-year-old boy with no induration.