Progressive Facial Hemiatrophy with Localized Scleroderma

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A 46-year-old woman who presented with progressive facial hemiatrophy (PFH) following localized scleroderma is described. The patient had a markedly deformed and depressed plaque surrounded by erythema on the right cheek. She also showed linear scleroderma with hair loss on the occipital area and morpha lesions on the neck and shoulder.

Histological findings of the facial and other atrophic lesions were consistent with localized scleroderma. Therefore, the PFH and localized scleroderma may have had the same pathological background in this case.

(Key words: Progressive facial hemiatrophy, localized scleroderma, Morphea, Linear Scleroderma)

INTRODUCTION

Progressive facial hemiatrophy (PFH) is a rare disease which was first described by Parry in 1825 [7]. The facial atrophy progresses slowly, and skin, subcutaneous tissue, muscle and occasionally cartilage and bone are also affected. Localized scleroderma, especially linear scleroderma, which occurs on the scalp and face as \textit{en coup de sabre}, may lead to deformity of the affected site of the face [5]. The type of localized scleroderma is considered to be closely related to PFH with respect to the processes involved [3]. We describe here a case of PFH with multiple localized scleroderma lesions and suggest that both diseases belong to the same clinical entities based on the histopathological findings.

CASE REPORT

A 46-year-old woman was first seen at Tokai University Hospital on August 30, 1989, complaining of deformity on the right side of her face. Fifteen months previously, she noticed hair loss on the back of her scalp. Approximately one year later, in May 1989, a dimple-like depression developed on her right cheek and gradually enlarged.

On clinical examination, the depressed site was located between the right lower jaw and anterior of the right ear. The depressed plaque was surrounded by erythema and the right upper lip was thin (Fig. 1). Three other depressed plaques were also noticed on the left side of the neck, right shoulder and scalp. The lesion on the occipital area showed loss of hair, shininess and edematous hardening (Fig. 2).

Laboratory studies revealed normal values for the differential white blood cell counts, erythrocyte sedimentation rate, antinuclear antibody, rheumatoid factor, quantitative immunoglobulins and chemistry profile. Radiographic examination showed no bony abnormalities of the face and skull.

Three biopsy specimens were taken, from the lesions in the occipital area, and the center and erythematous margin of the facial depressed plaque. Histological examination revealed epidermal atrophy and mononuclear inflammatory infiltrates around the blood vessels and appendages in all three specimens. An increase of fine collagen fibers was also demonstrated around the appendages of the dermis (Fig. 3). These features were compatible with scleroderma.

DISCUSSION

The pathogenesis of PFH is unknown, but overlapping with localized scleroderma is obvious. Localized scleroderma is classified into...
Fig. 1 Depressed plaque on the right mandible.

Fig. 2 Shiny, slightly hard linear alopecia lesion on the occipital area.
six groups [6]. The segmental type of localized scleroderma occurs on one side of the face, resulting in hemiatrophy. The en coup de sabre form of scleroderma is associated with some degree of facial hemiatrophy [1], and occasionally, both diseases occur simultaneously [4]. Christianson et al. surveyed 235 patients with scleroderma and found facial hemiatrophy in 38 [2]. Tuffanelli et al. reported three cases of linear scleroderma associated with PFH and emphasized their close relation [8]. An analysis of 53 linear scleroderma patients revealed that five patients with lesions of en coup de sabre had facial hemiatrophy [5].

A review of the patient's clinical course showed that symptoms of localized scleroderma appeared first, followed by a slowly progressing PFH. Linear scleroderma may lead to deformities of an extremity or one side of the face [3]. In our case, the localized scleroderma did not show an en coup de sabre type distribution. It was seen on the occipital area and was not continuous with the PFH lesion. The morphea lesions were not distributed on the same side as the PFH, but symptoms such as the thin lips and depressed plaque on the right cheek should be regarded as the initial stage of PFH. This PFH lesion is considered to be independent from, and concurrent with, localized scleroderma.

However, both the center and periphery of the depressed plaque on the right cheek showed the same pathological findings as scleroderma. It is suggested that both the PFH and localized scleroderma had essentially the same pathogenesis in our case.

REFERENCES