Keloidlike Morphea

Sir,

Although localized scleroderma usually presents as well-circumscribed flat or depressed sclerotic lesions, the development of nodular or keloidlike lesions has also been reported.

We describe a patient with localized scleroderma, who had flat elevated keloidlike lesions. These developed symmetrically on the neck, chest and back over 6 years.

CASE REPORT

A 61-year-old woman, without contributory personal or family history of keloid formation or rheumatoid diseases, came to our hospital in December 1994 for sclerotic skin lesions that had developed over 6 years. She had no history of injury on the neck, chest, back or face. Physical examination revealed irregularly pigmented sclerotic plaques that were nearly symmetrical on the neck, chest, back and face. The sclerotic skin on the neck limited the movement of the neck. Furthermore, the lesions on the neck, chest and back were elevated from the surrounding normal skin and appeared similar to keloid lesions (Fig. 1). She did not show any symptoms suggesting systemic scleroderma, such as Raynaud's phenomenon, sclerodactyly, or nail fold bleeding.

Laboratory data showed no abnormalities on complete blood count. Biochemical tests revealed moderate liver dysfunction (GOT activity (111 U/ml: normal 4–11 U/ml) and GPT activity (111 U/ml: normal 4–11 U/ml)) due to fatty liver and slightly elevated aldolase activity (9.1 U/ml: normal 2.1–5.7 U/ml). Antinuclear antibody, anticentromere antibody, anti-topoisomerase I, anti-DNA, anti-Sm, anti-nRNP, anti-SS-A, anti-SS-B and rheumatoid factor were all negative. A biopsy specimen from a keloidlike lesion on the chest showed thinned epidermis with basal pigmentation, and a thickened dermis composed of overlying homogenized collagen fibers. The hyalinized coarse collagen fibers that are often observed in keloids were absent. Perivascular mononuclear infiltration was sparse. Elastica van Gieson stain revealed a decreased number of elastic fibers.

Because of the progressive limitation of neck movement by the skin sclerosis, oral corticosteroid therapy (prednisolone 30 mg/day) was started and resulted in gradual improvement of the limitation of neck movement and skin sclerosis. At present, after 50 weeks, the patient is taking 12.5 mg/day of prednisolone. Skin sclerosis is remarkably improved, and the keloidlike elevation is also flattened to some extent.

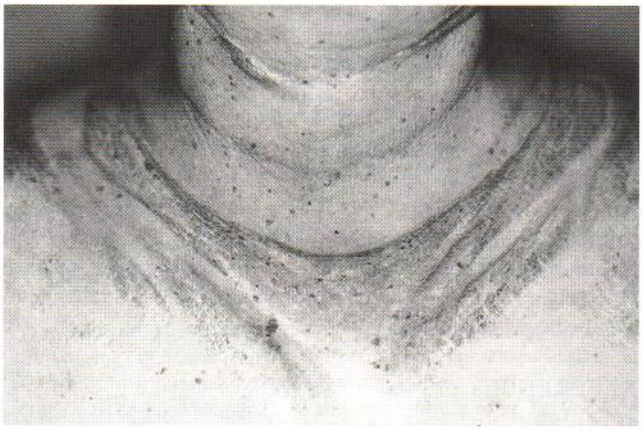

Fig. 1. Keloidlike sclerotic plaques on the chest were symmetrically arranged over the bones.

DISCUSSION

Nodular or keloidlike scleroderma is considered by Jablonska to be a rare variant of localized scleroderma (1). The terms "nodular scleroderma" and "keloidlike scleroderma" are now used as synonyms (2–4). In 1894, Unna described a keloidlike variant of scleroderma as the rarest type of scleroderma (5). Since then over 20 cases have been reported as nodular or keloidlike scleroderma. Previously reported cases appear to represent two categories: (a) systemic scleroderma accompanied by a generalized distribution of nodules (3, 4, 6, 7) and (b) scleroderma without documented systemic involvement with localized and generalized nodules or plaques (2, 8–11). For the latter category, Micalizzi et al. proposed the designation "nodular morphea" to differentiate it from nodular eruption with systemic sclerosis (11). Nodular scleroderma or keloidlike scleroderma, reported previously, was described as

nodules with a central depression, thickened red plaques with irregular margins (8) and handsized tuberosities (2).

Our patient had no clinical or laboratory evidence of systemic sclerosis, so this case was considered in the latter category of nodular scleroderma. Unlike the recently reported cases of "nodular scleroderma", our case did not present with nodules in sclerotic plaque, but keloidally elevated symmetrical plaques. We regard this case as strictly meeting the criteria described in the original case of Unna's "Keloidahnliche Sklerodermie", because of the keloidlike appearance and histopathology compatible with localized scleroderma. If "keloidlike morphea" and "nodular morphea" are considered to be in different categories, this is quite a rare case of "keloidlike morphea" in the strict definition.

REFERENCES

1. Jablonska S. Localized scleroderma: scleroderma and pseudoscleroderma. 2nd English edn. Warsaw: Polish Medical Publishers, 1975: 277–303.
2. Cabre BJ, Landes E. Ein Beitrag zur nodulären Sklerodermie (keloidähnliche Sklerodermie UNNA). Z Hautkrankheiten 1960; 28: 359–364.
3. James WD, Berger TG, Butler DF, Tuffanelli DL. Nodular (keloidal) scleroderma. J Am Acad Dermatol 1984; 11: 1111–1114.
4. Perez-Wilson J, Pujol RM, Alejo M, Geli C, de Moragas JM. Nodular (keloidal) scleroderma. Int J Dermatol 1992; 31: 422–423.
5. Unna PG. Die Histopathologie der Hautkrankheiten. Berlin: A. Hirschwald, 1894: 1119.
6. Krell JM, Solomon AR, Glavey CM, Lawley TJ. Nodular scleroderma. J Am Acad Dermatol 1995; 32: 343–345.
7. Burge SM, Mortimer PS, Ryan TJ. Pseudokeloidal scleroderma. Br J Dermatol 1985; 113(Suppl 29): 99.
8. Korting GW. Über keloidartige Sklerodermie nebst Bemerkungen über das etagenmäßig differente Verhalten von einigen sklerodermischen Krankheitszuständen. Arch Dermatol Syph 1954; 198: 306–318.
9. Akintewe TA, Alabi GO. Scleroderma presenting with multiple keloids. BMJ 1985; 291: 448–449.
10. Stefanato CM, Gorkiewicz-Petkow A, Jarzabek-Chorzelska M, Jablonska S, Chorzelski T. Morphoea with high titer of fibronectin antibodies. Int J Dermatol 1992; 31: 190–192.
11. Micalizzi C, Parodi A, Rebora A. Morphoea with nodular lesions. Br J Dermatol 1994; 131: 298–300. [letter]

Accepted July 19, 1996.

Masahide Kubo, MD, Takeshi Tamaki, MD, PhD, Manabu Fujimoto, MD, Kanako Kikuchi, MD, PhD and Kunihiko Tamaki, MD, PhD
Department of Dermatology, Faculty of Medicine, University of Tokyo 7–3-1 Hongo, Bunkyo-ku, Tokyo 113 Japan.