Acrogeria (Gottron type): A Vascular Disorder?

Chie Hashimoto, Masatoshi Abe, Nozomi Onozawa, Yoko Yokoyama, Osamu Ishikawa

Department of Dermatology, Gunma University Graduate School of Medicine

We report a 27-year-old Japanese man with the peculiar clinical features of acrogeria. He had had perniosis since early childhood. Prominent atrophic skin changes over the hands, hallux valgus, shortened distal phalanges and atrophic scars on his auricles were noted. X-ray of the hands revealed acro-osteolytic changes of the distal phalanges, and arteriography demonstrated multiple occluded branches of the digital arteries. There were no histological changes of systemic sclerosis in his forearm skin, nor antinuclear antibodies or coagulation disorders. Western immunoblottings demonstrated decreased production of type III collagen by dermal fibroblasts both from an affected finger and from the unaffected upper arm. Although the pathogenesis of acrogeria is unknown, the present case suggests that peripheral circulatory disturbance, as well as a congenital abnormality in type III collagen synthesis, may partly account for the pathogenesis of Gottron-type acrogeria.

Acrogeria (Gottron type)の一例

橋本智恵子、安部正敏、和田 望、横山洋子、石川 治

群馬大学大学院医学系研究科皮膚病態学