A Case of Isolated Plantar Collagenoma

Jong Woo Kim, M.D., Young Mook Yoon, M.D.,
Dong Seok Kim, M.D., Sang Won Kim, M.D.

Department of Dermatology, Catholic University of Taegu-Hyosung School of Medicine, Taegu, Korea

Isolated plantar collagenoma with a cerebriform appearance is a hamartomatous lesion consisting of proliferation of normal collagen tissue and has been reported with other systemic alterations described as Proteus syndrome.

We report herein a rare case of isolated collagenoma located on the plantar surface with macrodactyly of the 3rd and 4th toes occurring on the right foot, which is considered as an incomplete form of Proteus syndrome.

A 36-year-old male with normal intellectual and physical development visited with a pedunculated, soft, dome-shaped tumor and multiple, grouped, slightly elevated nodules on the right plantar region and hypertrophy of the 3rd and 4th toes of the right foot. The lesions appeared during early childhood and have been growing steadily since then. Total excision of the plantar mass with skin grafting and debulking of the 3rd toe and amputation of the 4th toe were done. (Ann Dermatol 9:(4):289~292, 1997).

Key Words : Isolated plantar collagenoma, Proteus syndrome

Collagenoma or connective tissue nevus of the collagen type is a hamartomatous lesion consisting of proliferation of normal collagen tissue, which is hereditary or acquired. The lesion consists of slightly elevated nodules that may be grouped or disseminated[1]. The hereditary lesions have been described in familial cutaneous collagenoma and tuberous sclerosis. Acquired isolated collagenomas have been described in various locations of the skin and sometimes in an unusual distribution pattern[2]. Isolated plantar collagenoma with a cerebriform appearance is rare and has been reported with other systemic alterations described as Proteus syndrome[1-4], but it has not been reported in the Korean literature.

We report herein a rare case of isolated collagenoma located on the plantar surface with macrodactyly of the 3rd and 4th toes occurring on the right foot, which is considered to be an incomplete form of Proteus syndrome.

Received June 2, 1997.
Accepted for publication September 25, 1997.
Reprint request to : Jong Woo Kim, M.D., Department of Dermatology, Catholic University of Taegu-Hyosung School of Medicine, Taegu, Korea

CASE REPORT

A 36-year-old male presented with a pedunculated, soft, dome-shaped tumor on the right plantar region and hypertrophy of the 3rd and 4th toes of the right foot. The lesions appeared during early childhood and have been growing steadily since then. He complained of intermittent pain on walking. The family history was insignificant. On physical examination, intellectual and physical development were normal. The skin findings consisted of a dome-shaped tumor located on the right plantar surface with elevated band-like nodular lesions from the tumor lesion to the heel(Fig. 1A). Macrodactyly of the 3rd and 4th toes was also observed. The fourth toe was about twice as large as the great toe with slight upward and medial deviation(Fig. 1B). Laboratory findings including a complete blood cell count, urinalysis, liver function test, serology and radiological examinations of the skull, thoracic and lumbar spines were within normal limits or negative. A chromosomal study revealed normal male karyotypic patterns. Radiographical findings of the right foot

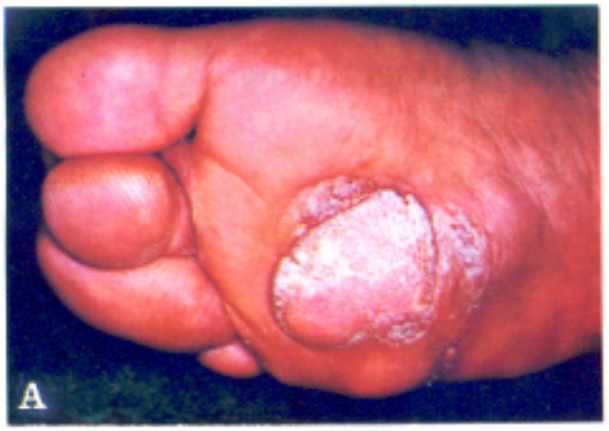

Fig. 1A. A 5.0 × 5.0 × 1.0 cm pedunculated, soft, dome-shaped non-tender tumor and nodules on the right plantar region and hypertrophy of the 3rd and 4th toes of the right foot.

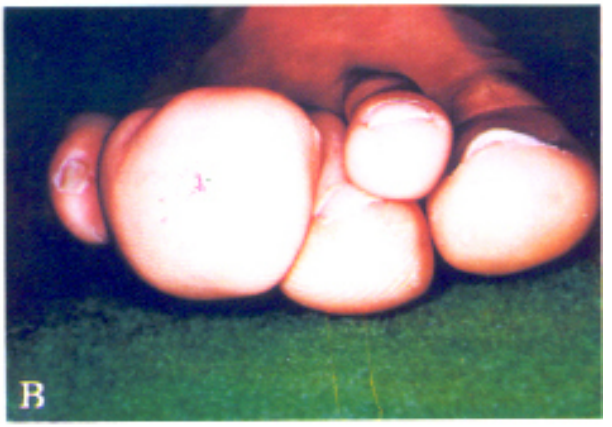

Fig. 1B. The fourth toe was about twice as large as the great toe with slight upward and medial deviation.

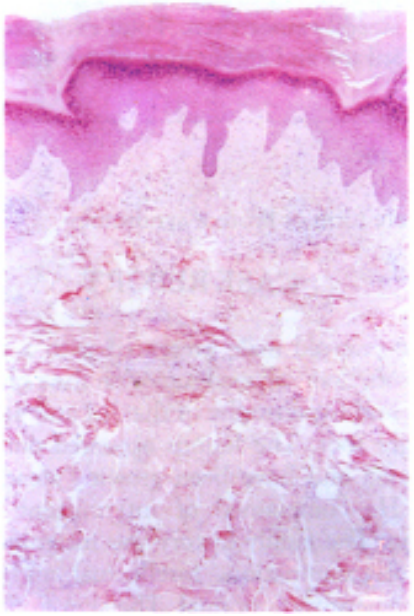

Fig. 2. Histopathological findings of the excised plantar mass showed epidermal hyperplasia and patchy infiltration of the inflammatory cells in the thickened dermis(H &E stain, ×100).

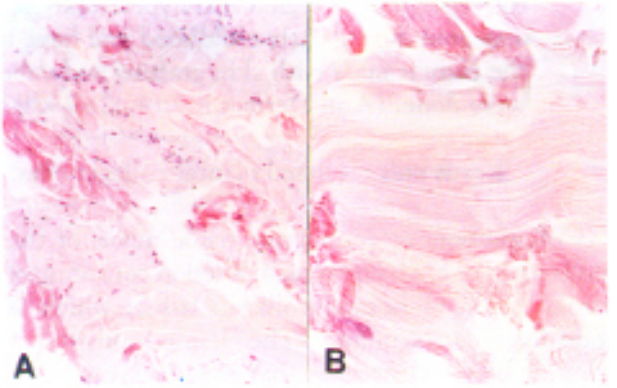

Fig. 3. Increased amounts of with a thickened appearance in the dermis(H&E stain, A: ×200, B: ×400).

showed hyperostosis of the 3rd and 4th toe, medial deviation of the distal and middle phalanx of the 4th toe, and increased soft tissue density. Histopathological findings of the excised plantar mass showed epidermal hyperplasia and patchy infiltration of lymphohistiocytes in the thickened dermis(Fig. 2). There were increased amounts of collagen bundles with thickened apperances(Fig. 3). A masson trichrome stain revealed a significant increase of disorganized collagen fibers(Fig. 4). Total excision of the plantar mass with skin grafting and debulking of the 3rd toe and amputation of the 4th toe were done. No relapse was noted after about 5 months' postoperative follow-up.

DISCUSSION

Connective tissue nevi of the skin are hamartomatous lesions that involve predominantly one of the extracellular matrix components - collagen, elastin, or proteoglycans[5]. Isolated plantar collagenoma with cerebriform appearances have been described as one of the major skin findings of Proteus syndrome[1,6,7].

The term Proteus syndrome was first introduced by

Fig. 4. Trichrome staining revealed an increased amount of collagen bundles(× 100).

Wiedemann et al[6] in 1983 to describe a disorder of mesodermal malformation with a wide range of birth and developmental defects and therefore, with an enormous morphologic variability. The syndrome was named after the Greek God Proteus, whose name meant "the polymorphous". Major clinical findings are hemihypertrophy, macrodactyly, subcutaneous tumors(with wide histologic variation), cerebriform skin hyperplasia of the soles and/or palms(collagenoma), exostoses, epidermal nevi, and scoliosis. This syndrome appears sporadically and although some manifestations are already present at birth, the majority appears during early childhood[8-11].

Hotamisligil reported a scoring system for diagnosis of Proteus syndrome[12]. A score of 13 or more points definitely establishes the diagnosis. Our patient had isolated plantar collagenoma with macrodactyly. The total scores were about 9 points. Therefore, our case may be thought to be an incomplete form of Proteus syndrome. Another criteria demand at least four of the seven characteristics[13].

As Uitto et al[5] have shown, collagenoma almost exclusively consists of type I collagen. The underlying defect seems to be a reduced production of collagenase in that location and therefore a decreased local degradation of collagen. Moreover, the mean population doubling time of the fibroblasts from these lesions is decreased ; thereby an enhanced proliferative capacity of the regional fibroblast may have contributed to the accumulation of collagen.

Isolated plantar collagenoma can be distinguished from neurofibromatosis by the absence of multiple cafe-au-lait macules, Lisch nodules, axillary freckling, and multiple neurofibroma[1,6]. The presence of hemihypertrophy and macrodactyly in neurofibromatosis is rare and usually occurs with plexiform neurofibromas.

Histologically, biopsy specimens reveal epidermal hyperplasia with a significant increase of disorganized collagen fibers, best seen with Masson trichrome stains, at the papillary and reticular dermis[14-15].

Surgical reconstruction is the primary mode of rehabilitation for children with Proteus syndrome[12]. In our case, total excision of the lesion with debulking of the 3rd toe and amputation of the 4th toe were done. There was no relapse of collagenoma after about 5 months' postoperative follow-up.

REFERENCES

1. Samlaska CP, Levin SW, James WD, et al: Proteus syndrome. Arch Dermatol 125:1109-1114, 1989.
2. Botella-Estrada R, Alegre V, Sanmartin O, et al: Isolated plantar cerebriform collagenoma. Arch Dermatol 127:1589-1590, 1991.
3. Calza AM, Balderrama F, Saurat JH: Isolated plantar cerebriform collagenoma. Pediatric Dermatol 11:84-85, 1994.
4. Uitto J, Santa Cruz DJ, Eisen AZ: Connective tissue nevi of the skin: clinical, genetic and histopathologic classification of hamartomas of the collagen, elastin, and proteoglycan type. J Am Acad Dermatol. 3;441-461, 1980.
5. Uitto J, Bauer EA, Santa Cruz DJ, et al: Decreased collagenase production by regional fibroblasts cultured from skin of a patient with connetive tissue nevi of the collagen type. J Invest Dermatol 78:136-140, 1982.
6. Wiedemann H-R, Burgio GR, Aldenhoff P: The Proteus syndrome. Eur J Pediatr 140:5-12, 1983.
7. Clark RD, Donnai D, Rogers J, Cooper J, Baraitser M: Proteus syndrome : an expanding phenotype.

Am J Med Genet 27:99-117, 1987.
8. Viljoen DL, Saxe N, Temple-Camp C: Cutaneous manifestations of the Proteus syndrome. Pediatr Dermatol 5:14-21, 1988.
9. Burgio GR, Wiedemann H-R: Further and new details on the Proteus syndrome. Eur J Paediatr 143:71-73, 1984.
10. Temtamy SA, Rogers JG: Macrodactyly, hemihypertrophy, and connective tissue nevi : report of a new syndrome and review of literature. J Pediatr 89:924-927, 1976.
11. Tibbles JAR, Cohen MM Jr: Proteus syndrome : the Elephant Man diagnosed. Br Med J 293:683-685, 1986.
12. Darmstadt GL, Lane AT: Proteus syndrome. Pediatric Dermatol 11(3):222-226, 1994.
13. Hotamisligil GS: Proteus syndrome and hamartoses with overgrowth. Dysmorphol Clin Genet 4:87-102, 1990.
14. Jeanfils S, Tennstedt D, Lachapelle JM: Cerebriform intradermal nevus. Dermatology 186:294-297, 1993.
15. Lee SM, Kang WH, Lee SH: A case of connective tissue nevi. Ann Dermatol 6:230-235, 1994.