A Case of Connective Tissue Nevi

— *Elastocollagenoma compared with Elastofibroma* —

Seung Min Lee, M.D., Won Hyung Kang, M.D.*, Seung Hun Lee, M.D.

Department of Dermatology, Yonsei University College of Medicine, Seoul, Korea
*Department of Dermatology, Aju University College of Medicine, Suweon, Korea**

Connective tissue nevi are developmental dysplasias of connective tissue with variable histologic changes, particularly as regard the amounts of collagen, elastic tissues, proteoglycans and glycoproteins. A number of clinical variants characterized by papules, plaques or nodules have been described.

We herein report a case of connective tissue nevi.

A 4-year-old girl had a palm sized hard nontender indurated plaque on her left thigh. The microscopic findings and electron microscopic findings are compatible with elastocollagenoma which lesion had increased amounts of both collagen and elastic fibers, and structually abnormal elastic fibers are intimately interwoven with collagen fibers in disarray.

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Key Words: Connective Tissue Nevi, Elastocollagenoma

Connective tissue nevi are circumscribed hamartomatous malformations of the dermal extracellular matrix i.e. collagen, elastic fibers or glycosaminoglycans[1,2].

Present at birth or appearing within the first few years of life, connective tissue nevi are variable in appearance. These elevated soft to firm tumors vary in size from 0.5cm to several centimeters in diameter and may be grouped, linear or irregularly distributed over the body surface[3].

Connective tissue nevi of the acquired type have been classified by Uitto et al[2] as eruptive collagenomas, isolated collagenomas, and isolated elastomas depending on the number of lesions and predominant dermal fibers present. The hereditary types of connective tissue nevi include familial cutaneous collagenoma[3], dermatofibrosis lenticularis disseminata in the Buschke-Ollendorff syndrome[4] and shagreen patches in tuberous sclerosis.

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Reprint request to: Seung Min Lee, M.D., Department of Dermatology, Yonsei University College of Medicine, Seoul, Korea

In another classification[6], connective tissue nevi in which the elastic tissue changes dominate have been referred to as the Lewandowsky type and those in which the collagen changes are conspicuous as the Lipschutz type. However the situation is not clear-cut and mixed forms occur in which the pattern of both collagen and elastic tissue is altered

We report herein a case of connective tissue nevi with results from electron microscopy.

REPORT OF A CASE

A 4-year-old girl visited our clinic complaining of a skin lesion on her left thigh which she had had for 4 months. There were no accompanying subjective symptoms. There was no history of previous treatment. Her past and family history were noncontributory.

Physical examination revealed a 12 × 15cm sized well defined circinate non tender skin colored indurated plaque on the left thigh(Fig. 1). Her right thigh's circumference was 27cm and left thigh's circumference was 33.5cm. The following laboratory tests were within normal range or negative: Complete blood count, urinalysis, liver function

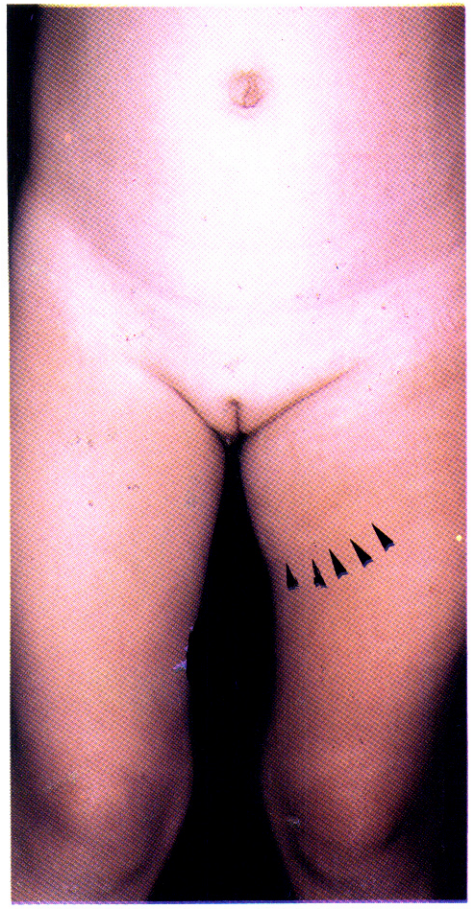

Fig. 1. A 12 × 15 cm sized, well defined, skin colored, indurated plaque on the left thigh.

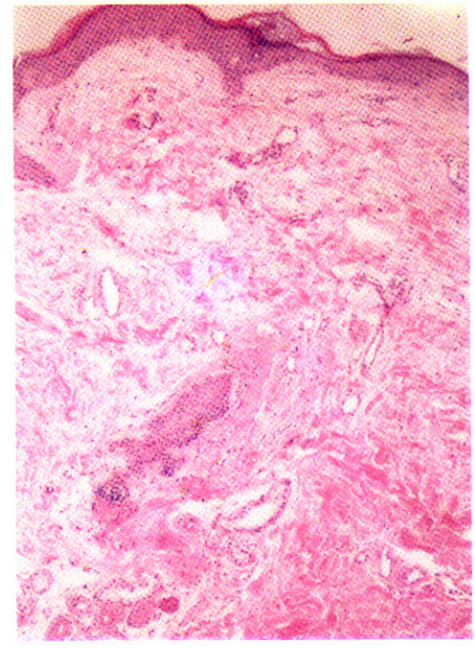

Fig. 2. Thickened dermal layer and collagen bundles which increase in amount(H & E stain, × 40).

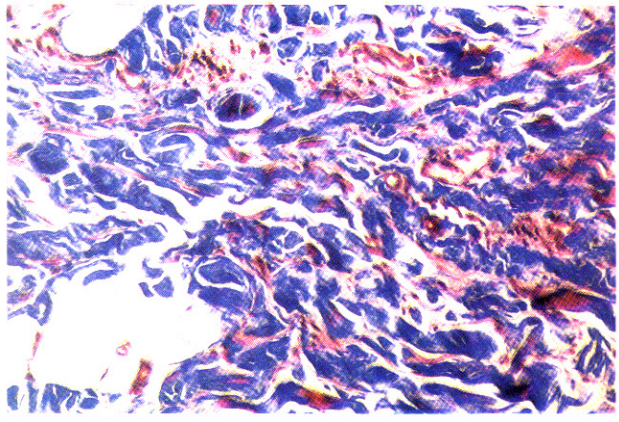

Fig. 3. Higher magnification of collagen bundles which increase in amount with shorter and irregularily arranged appearance (Masson's trichrome stain, × 100).

test, serum electrolyte, chest roentgenogram. Multiple bone series showed a normal range except for soft tissue swelling and irregular radiopaque density of the left thigh in pelvis AP. Magnetic resonnance image and RBC pooling scan of the both extremities showed no vascular lesion.

A skin biopsy specimen was obtained from the lesion on the left thigh. Hematoxylineosin stain showed a thickened dermis(Fig. 2). Masson's trichrome stain showed an increased amount of collagen bundles with a shorter and irregularly arranged appearance(Fig. 3). Elastic tissue stain revealed increased elastic tissue and thickening of elastic fibers and a few foci of agglomerated, coarsed irregular elastic fibers in the mid dermis(Fig. 4 a,b).

Electron microscopic findings showed compact bundles of normal collagen fibrils which had a uniform thickness in cross section and elastic fibers which had a shortened and thickened appearance(Fig. 5a). The fibroblasts showed markedly folded nucleus and cisternae of endoplasmic reticulum filled with amorphous material(Fig 5 b,c). She was not given any treatment.

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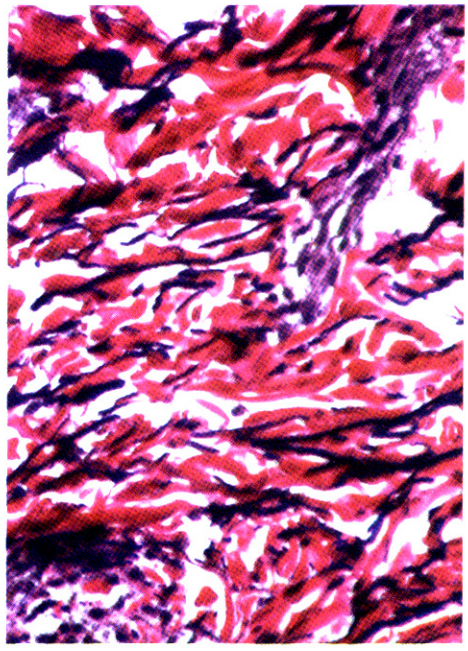

Fig. 4 a). The elastic fibers which increase in amount (Verhoeff van Gieson stain, × 100).

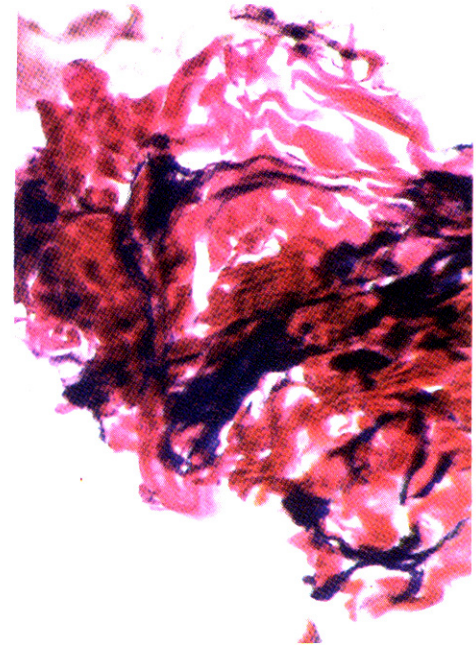

Fig. 4 b). Higher magnification of elastic fibers which are thickened, agglomerated and coarsed irregularily arranged (Verhoeff van Gieson stain, × 400).

DISCUSSION

Nevoid anomalies of the connective tissue of the skin are uncommon and are variable in appearance. The elevated soft to firm tumors may be grouped, linear or irregularly distributed over the body surface. The color may be flesh, brown, hypopigmented or yellow[7].

Nevi of connective tissue may be classified according to an excess or deficit in the number of cells per unit volume and amount or proportion of biosynthetic products, i.e., the various types of collagen, elastin, proteoglycans and glycoprotein[1,2,8]. Among them, elastocollagenoma lesion has increased amounts of both collagen and elastic fibers and structually abnormal elastic fibers are intimately interwoven with collagen fibers in disarray. Because elastic fibers cover thickened bundles of collagen like a sheath, staining procedure for elastic fibers may lead to a faulty conclusion that the dermis is made up only of elastic fibers[1]. In our case, we also observed that elastic and collgen fibers are mixed with bundles and elastic fibers are thickened, shortened and intimately interwoven with each other. Electron microscopy revealed a predominance of collagen fibers arranged in interwining bundles with characteristic periodicity. The striking feature was that the fibroblast had a markedly folded nucleus with a dilated rough endoplasmic reticulum and the fibroblast was surrounded by collagen fibers and amorphous elastic masses.

Dixon and Lee[9] in 1980 studied an ultrastructure of elastofibroma and proposed that the newly formed elastophilic material is produced by the fibroblasts. They observed the presence of numerous pinocytotic vesicles and conspicuous coated vesicles which suggested active cellular synthesis and secretion of fibroblasts. In our case, the elastotic mass was often seen in close proximity to the fibroblasts and numerous pinocytotic vesicles presented.

Elastofibroma, originally described in the lower subscapular area is connective tissue nevi of increased collagen and elastin which affects elderly person and is a slow growing mass has now been reported in many cases[9,10]. The morphogenesis of elastofibroma and the question of whether it represents a true neoplasm or a degenerative pseudotumor have been the subject of considerable debate. Some authors[9] suggested that abnormal elastic tissue

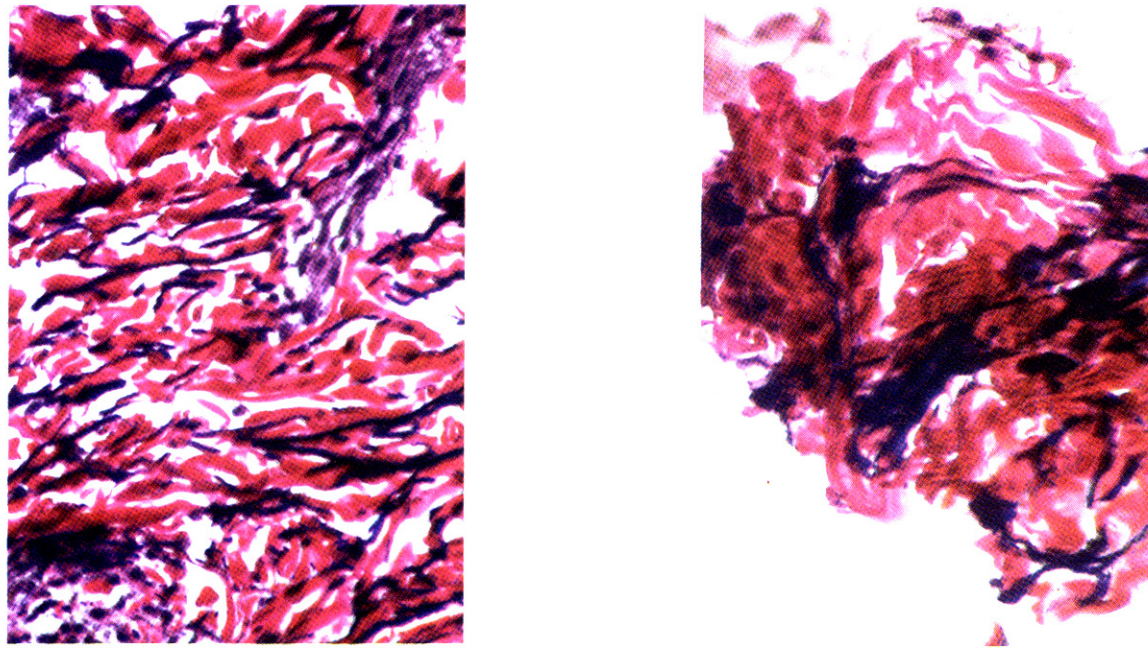

Fig. 5a). Compact bundles of normal collagen fibrils which had uniform thickness in corss section and fibers(arrowheads) which had shortened and thickened appearance(EM, × 9,780).

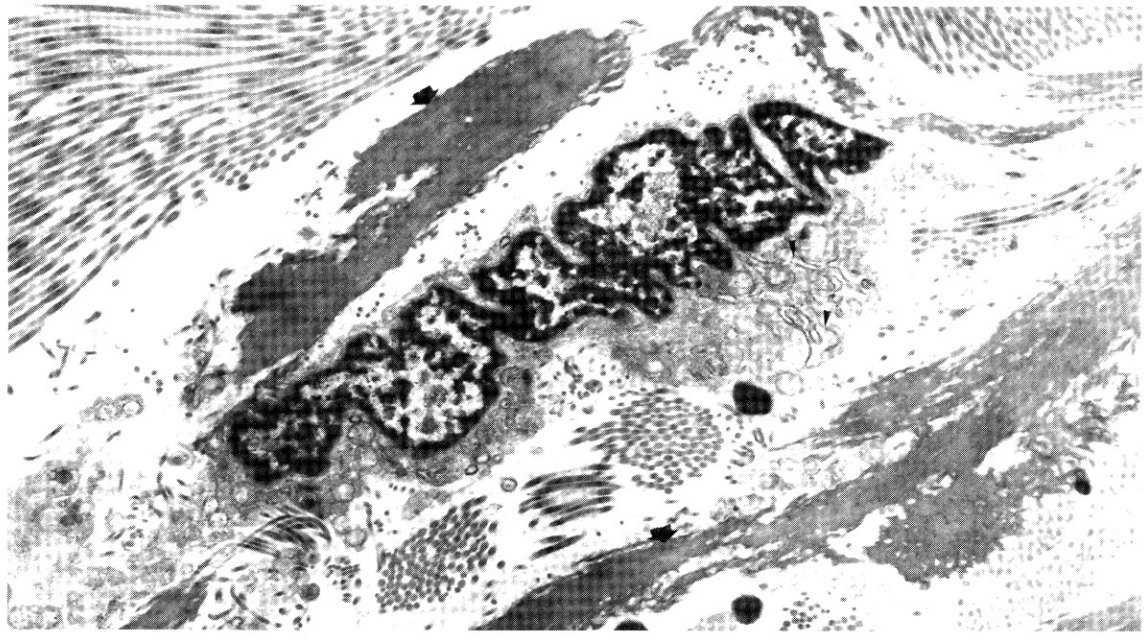

Fig. 5b). Fibroblast had markedly folded nucleus with dilated rough endoplasmic reticulum(small arrowheads) and fibroblast surrounded by collagen fibers and amorphous elastic masses(large arrows)(EM, × 14,000).

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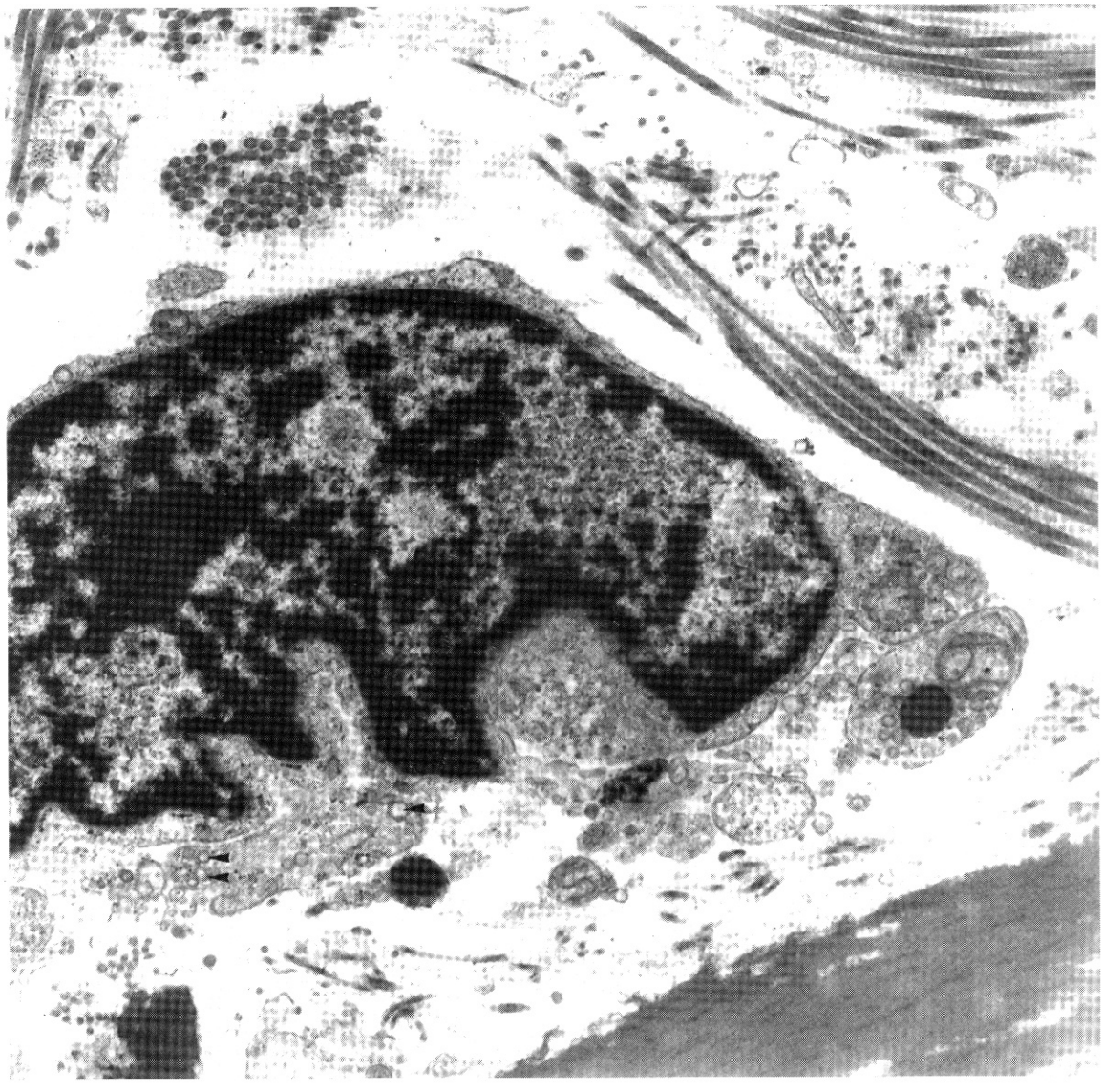

Fig. 5c). Higher magnification electron micrograph of fibroblast with numerous pinocytotic vesicles(arrowheads)(EM, × 28,000).

in elastofibroma is the result of excessive production of elastic material by fibroblasts rather than elastotic degeneration of collagen and another[10] suggested that repeated trauma or friction may play a role in this condition.

Elastocollagenoma is connective tissue nevi of increased collagen and elastin but is another type of lesion which shows a different predilection site, age and pathogenic mechanisms. Our patient is a 4-year-old girl who had the lesion on the left thigh. Uitto et al[11] suggested that excessive deposition of collagen in connective tissue nevi may have resulted from decreased local degradation of collagen and enhanced proliferative capacity of the lesional fibroblasts. We also noted good morphologic evidence for active fibroblast synthesis and secretion and observed elastotic material around the fibroblasts. The absence of family history supports the suggestion of the non-hereditary origin of this disorder.

We suggest that elastocollagenoma is a distinctive entity which includes connective tissue nevi and has characteristic histopathologic and electron microscopic findings.

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